MALIGNANT TUMOURS OF THE MAXILLA

A Thesis Submitted for Partial Fulfilment of Master Degree in Otorrhinolaryngology

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INTRODUCTION

Cancer was first mentioned as a cause of death in 1629 in the Bills of Mortality which were produced annually in England. Pott's studies on scrotal cancer were presented in 1775, but it was not until the 19th century that the first significant modern studies on cancer appeared. At the end of the 19th and the early 20th centuries, studies based predominantly on autopsy and biopsy material became increasingly frequent. Statistics became more readily available from such registrars as were present in the United Kingdom and by the beginning of the Connecticut Cancer Registry in 1935, the modern era of cancer epidemiology came into being.

Tumours arising within or involving the nasal cavity and related para-nasal sinuses were recognized in the time of Hippocrate who distinguished between hard and soft tumors, but believed that the treatment only shortened the patient's life.

While not among the leading causes of death from cancer, cancer of the maxilla is a particulary threatening experience for the patient, his family and friends. Cancers in this area are disabling and fatal if untreated, but even if treated a patient faces possible loss of sensory organs, disability from loss of physiological function, sometimes a drastic change in personal appearance and often death.

This study is concerned with the review of the tumours arising within or involving the maxilla. The anatomy, radiology, aetiology, pathology and clinical picture of the disease were discussed with stress on the recent studies and latest knowledge in the subject. Then the methods of diagnosis were discussed obviating the use of the most update means which could make diagnosis an easier and earlier mission. Lastly, the history of management of the disease was reviewed discussing the various modalities of treatment and the trials made by most sincere and devoted authors and researchers, aiming at the conclusion of the best available mangement keeping in mind the longer and happier life of the patient as a final goal.

ANATOMY OF THE MAXILLA

The maxillae are the largest bones of the face next to the mandible, and it is their growth which is responsible for the elongation of the face between the ages of 6 and 12 years. In the norma frontalis of the skull, the part above the mouth is formed almost entirely by the maxillae. The two maxillae unite in a midline intermaxillary suture to form the whole of the upper jaw. They form most of the roof of the mouth, the lower and lateral boundaries of the anterior nasal aperture, and the floor and lateral wall of the nasal cavity.

In addition, on each side the maxilla forms most of the orbital floor, the medial part of the lower orbital margin and part of the medial margin. The maxillae also share in the formation of the infratemporal and pterygo-palatine fossae, and the inferior orbital and pterygomaxillary fissures.

Each maxilla consists of a body, enclosing the maxillary sinus (antrum) or antrum of Highmore, and four processes: zygomatic, frontal, alveolar and palatine.

This chapter will not describe details of the gross anatomy of the maxilla but will be restricted to the anatomical variations which have some surgical importance.

The body is roughly pyramidal and has four sides or surfaces; anterior (facial), infratemporal (posterior), orbital (superior) and nasal (medial), which enclose a large cavity, the maxillary sinus (antrum) or antrum of Highmore.

The Maxillary Sinus (Antrum of Highmore):

It is the largest accessory air sinus in the skull. Embryologically, it is the first sinus to develop, and it does so by evagination from the middle meatus. It appears as a shallow groove on the medial surface of the maxilla about the fourth month of intra-uterine life and continues to expand until adulthood. (Warwick and Williams 1973).

At birth, it presents as a shallow slit with average measurements of 7x4x4 mm and average capacity of 1 cm^3. The sinus reaches full size after eruption of all the permenant teeth (Last 1978).

In the mautre skull, it is a large pyramidal cavity that occupies the whole of the body of the maxilla. There is considerable variation in the size of the maxillary sinus in different skulls and even in the same skull, the average measurements being: the vertical height opposite the first molar tooth, 3.5 cm; the transverse breadth 2.5 cm; the anteroposterior depth, 3.2 cm. The capacity of the sinus varies from 9.5 to 20 ml with an average of 14.75 ml (Logan Turner 1977).

The walls of the maxillary sinus are thin and correspond to the orbital, alveolar, anterior and infratemporal surfaces of the body of the maxilla.

Its apex is directed laterrally, extends into the zygomatic process and may invade the zygomatic bone itself.

The base faces medially and represents the lateral wall of the nasal cavity and presents the maxillary hiatus which is the opening of the maxillary sinus.

The posterior wall is pierced by the alveolar canals which transmit the posterior superior alveolar vessels and nerves to the molar teeth; these canals occasionally project as ridges into the maxillary sinus.

The floor is formed by the alvealer process of the maxilla, and its lower part is about 1.25 cm below the level of the floor of the nasal cavity in 65% of the population, level with floor of the nose in 15% and above the level of the nasal floor in 20%, but in infants the floor of the sinus is level with middle meatus.

The relation of the upper teeth to the maxillary sinus varies considerably according to the pneumatization of the alveolar process. In most skulls, radiating septa of varying sizes spring from the floor of the sinus in the intervals between the adjacent teeth. Frequently, the roots of the first

and second molar teeth, less commonly, the roots of the first and second premolars and third molar, and occasionally the root of the canine, project into the floor of the sinus which is sometimes perforated by one or more of these teeth. The roots are usually covered by a thin layer of compact bone; occasionally, the frangs and dental nerves and vessels are in contact with the mucous membrane of the antrum.

The infra-orbital canal usually projects into the sinus, a well-marked ridge extending from the roof to the anterior wall transmitting the infra-orbital vessels and nerves.

The bony wall of the antrum is 5-8 mm thick, but it may be reduced to be paper thin according to the pneumatization of the antrum. There may be a considerable amount of cancellous bone in the walls of the antrum, on the other hand, the cavity of the sinus may be extremely large and modeled into all the surrounding structures.

Occasionally, the sinus may be divided into two more or less separate cavities by septa. These 2 cavities may communicate or may be completely separate and open separately in the nose.

Mucous Membrane of the Maxillary Sinus:

It is in direct continuity with that of the nasal fossa. It is respiratory in type, composed of pseudostratified columnar ciliated epithelium resting

on a tunica propria of lose connective tissue. The corium is lax and cellular containing small groups of serous and mucous glands. Many of the ciliated cells are goblet cells and are mainly close to the ostia through which the cilia wash the mucus in spiral tracks.

The Blood and Nerve Supply:

The blood supply of the mucous membrane is by small arteries that pierce the bone, mostly from the facial, maxillary, infra-orbital and greater palatine arteries, and veins accompany these vessels to the (anterior) facial vein and to the petrygoid plexus.

The nerve supply of the mucous membrane is by the superior alveolar nerves (posterior, middle and anterior), the anterior palatine nerve and the infra-orbital nerve; all are branches of the maxillary division of the trigeminal nerve.

The posterior superior alveolar nerves, usually two, pierce the posterior wall of the maxilla and pass forwards in the bone above the apices of the molar teeth, which they supply.

The middle superior alveolar nerve leaves the infra-orbital nerve on the floor of the orbit and runs down in the lateral wall of the maxilla to supply to premolar teeth, and the overlying mucous membrane of the antrum.

The anterior superior alveolar nerve leaves the infra-orbital nerve in the infra-orbital canal in the roof of the antrum. It passes laterally, then curves medially below the infra-orbital foramen, in the anterior wall of the maxilla. Branches supply the mucous membrane of the anterior wall of the antrum.

The anterior palatine nerve, in its canal, gives off minute branches which perforate the maxilla to supply the posterior part of the medial wall of the antrum.

The infra-orbital nerve gives perforating branches that supply the roof of the antrum.

The lymph drainage is, for the most part, via the infra-orbital foramen or the ostium to the retropharyngeal and/or subdigastric nodes, in either case the lymphatics flow to the submandibular glands. The maxillary sinus is lined by a lymphatic capillary network which connects with the network of the nasal fossa.

A clinically important point is that the area of choice for antral punctures is the medial wall of the sinus as the bone separating the inferior meatus from the sinus is very thin at the angle between the lateral wall of the nose and the inferior turbinate.

Another clinically very important point about the embryological development of the midface was noted by Panje and Ceilley (1979). Their studies on 150 cases of midface skin cancer revealed that local epithelial cancer spread is markedly influenced by embryological fusion planes. Six fusion planes which arise from the junction of embryonic masses of ectoderm and mesoderm eventually form the adult midface. Tumour sprelad appears to be contained within the junctional boundaries of the fusion planes, while cancer development over an embryonic fusion plane site tends to mark deep invasion.

So, tumour originating just lateral to the lateral nasal process in the development of the face will tend to invade the lower eye lid and the upper lateral lip originally developing from this process. Likewise, skin neoplasms originating on the lower eye lid and upper lateral lip tend to spread toward the neck. The large and more aggressive lesions in this area tend to invade deeply along the infra-orbital nerve into the maxillary sinus and orbital apex.

RADIOLOGY

Radiological appearance of the maxilla includes both plain and tomographic radiological appearances.

I. PLAIN RADIOLOGICAL APPEARANCE:

Normal sinuses are radiotranslucent, whereas diseased sinsues show varying degrees of opacity. Radiographs also reveal the extent of development of the sinuses.

The maxillary sinuses are best evaluated in the Water's view, a frontal projection.

The maxillary sinuses tend to be quite symmetric in size and configuration, although minor degrees of asymmetry are common place. Unilateral hypoplasia is present in 1.7% of cases and bilateral hypoplasia in 7.2% of cases.

In the antero-posterior (Waters) view, the sinuses appear as pyramidal-shaped translucent areas below the orbits and lateral to the lower part of the nasal cavity. Inferiorly they extend into the alveolar process of the maxilla. Fig. 1.

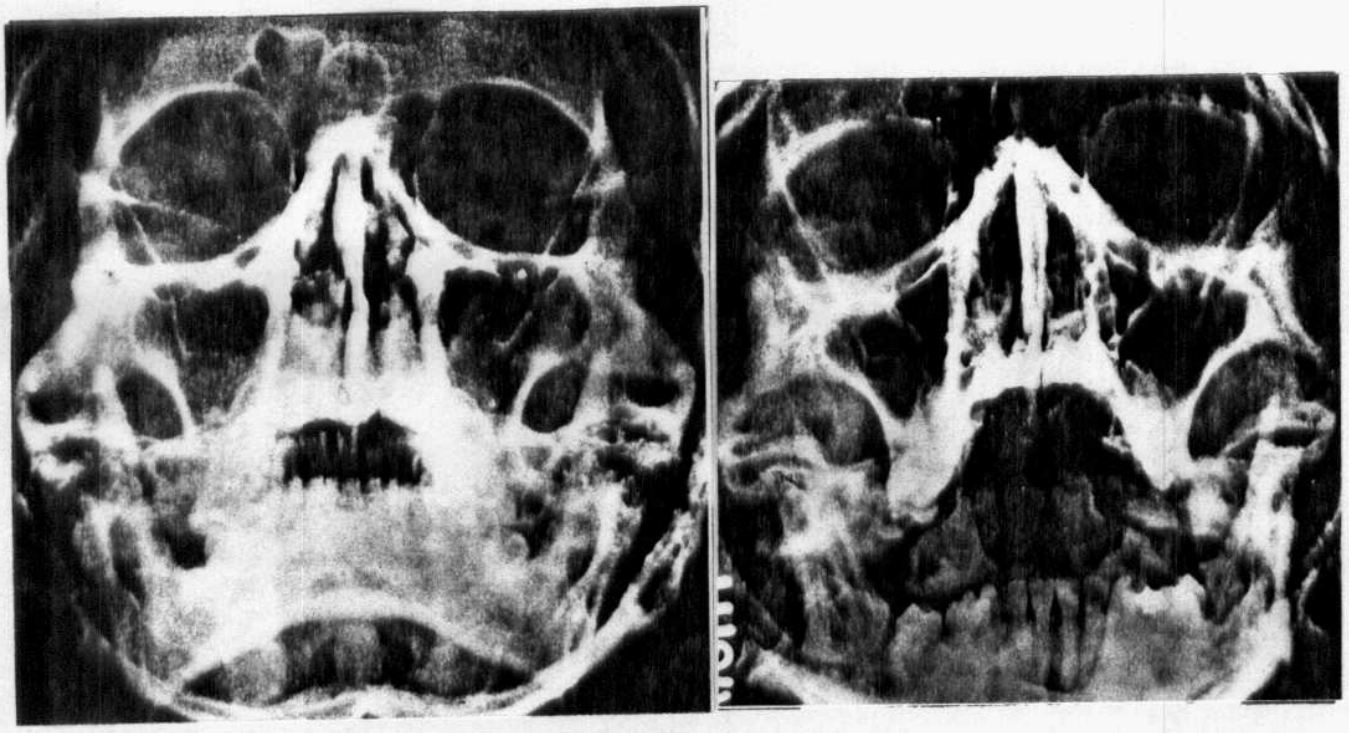

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Fig. (1):

Radiological appealrance of maxilla in waters view:

(A) Sample standard (closed mouth) radiograph.

(B) Open-mouth radiograph.

In the lateral view, the maxillary sinus is well seen; it lies below the orbit and its extent in relation to the roots of the teeth can be seen clearly. Fig. 2.

In the base view, the posterior maxillary sinus wall is represented as an s-shaped line. The pterygoid fossa is always viewed in the base view as a V-shaped space with its apex directed anteriorly. The medial and lateral pterygoid plates cast 3 shadows, a medial line for the medial plate and two lateral lines for the lateral plate.

The hard palate is best seen in the lateral view. Its nasal surface appears with a well-delineated cortical margin. The oral surface is slightly concave downward.

II- TOMOGRAPHIC APPEARANCE:

1. Conventional tomography:

Tomograms have the advantage of giving much more clear pictures especially of soft tissue. They are usually obtained in straight anteroposterior or lateral views in different levels at 0.5 cm intervals. The frontal processes of the maxillary bones, for example, form the slightly curved lateral margins of the nasal cavity. The contours of the maxillary sinus are not seen in their entirety on any single tomographic section.

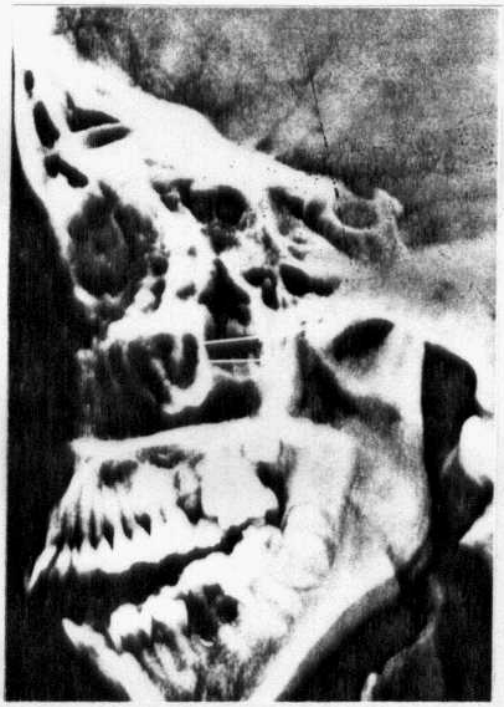

Fig. 2

Lateral view with white arrows indicating posterior maxillary sinus wall and anterior margin of pterygomaxillary fissure.

Anteriorly, the medial wall and roof are in the plane of focus. More posteriorly, the middle roof, part of the lateral wall and zygomatic recess are seen. Further posteriorly, the superomedial recess or ethmoidomaxillary plate (maxillo-ethmoidal bar) region is in focus. The inferior orbital fissure separates the roof of the maxillary sinus from the lateral wall of the orbit and is usually seen as two laterally divergent bony lines. The upper line is the lower margin of the greater sphenoid wing; the lower line is the maxillary sinus roof.

The maxillary sinus cavity diminishes in size as we go posteriorly. Its configuration changes from triangular anteriorly to a vertically oriented triangle posteriorly. Fig. 3.

2. **Computed tomography (CT):**

Computerized tomography (CT) has added the advantage of getting horizontal sections with excellent clarity with both soft tissue and bones. CT scans are obtained in either axial or coronal projections.

Axial projection: The most superior view obtained is that of the frontal sinus. The maxillary sinus roofs (orbital floors) usually appear quite asymmetric. The sinus apex appears as a round or ovoid lucency in the posteromedial portion of the orbital floor. The slightly flatter anterolateral floor is more easily seen in the AP view. Occasionally, portions of the infra-orbital canal can also be identified.

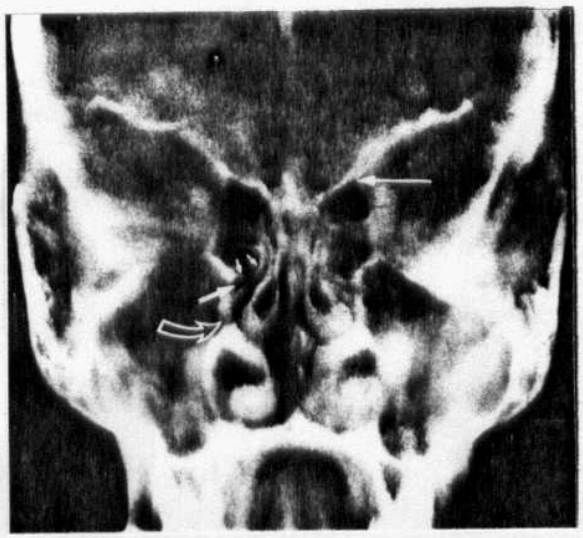

Fig. 3

Medium white arrows lie on ethmoid infundibulum, which opens into hiatus semilumaris. Small white arrows delimit ethmoid bulla. Open arrow points to membranous portion of medial wall of antrum. Long white arrows point to anterior ethmoid canal.

The antero-inferior orbital floor is often poorly seen because it is often obliquely sectioned. More caudal scans show a clear well-defined anterior maxillary sinus wall. The overlying facial soft tissues, postero-lateral wall, and medial wall all appear sharply defined.

Some thinning of the posterior wall near the pterygoid plates is often observed. The hard palate is often clearly visualized. (Fig. 4).

Coronal projection:

The anatomic orientation is similar to that described in conventional tomography Fig.5 (Bergeron and Osborn 1984).

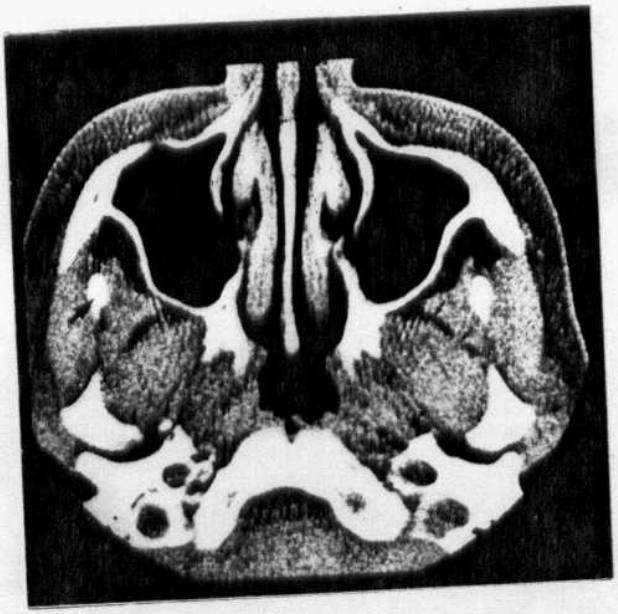

Fig.(4): Axial CT scan. Small arrows outline lateral pterygoid muscle. Large arrow points to coronoid process of mandible. Inferior turbinates, nasal septum, and antral walls are all clearly seen.

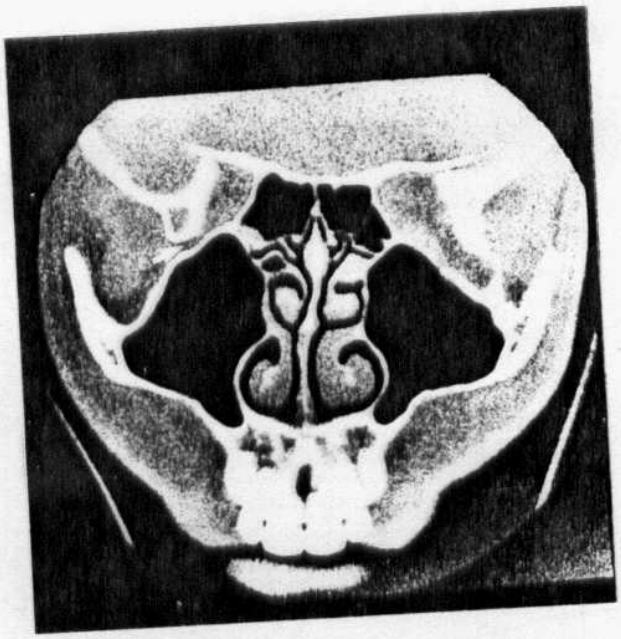

Fig.(5): Coronal CT scan. More posterior scan clearly shows antral walls and nasal cavity structures. Inferior orbital fissure is also seen (arrow).

AETIOLOGY

The maxilla, being a part of the aerodigestive tract is subject to a broad range of possibly carcinogenic agents. Several studies have been made and have shown positive relation between various factors and head and neck cancer. Hereby are the factors that have been shown to have possible relation to cancer of the maxilla.

I- OCCUPATIONAL AGENTS:

These represent a significant source of chemical carcinogens. Keane et al (1981) revised these factors and concluded these to be the following:

a- Bone seeking radioisotopes such as mesothorium have been implicated in upper respiratory tract cancer.

b- The ingestion of radium agents by radium dial painters and radium chemists has resulted in carcinomas of the paranasal sinuses, tongue and the lip.

c- The carcinogenic effect of mustard gas has been first reported in a study on Japanese workers in a factory manufacturing this gas in the period 1929-1945 where there was a dramatic increase in the incidence of respiratory tract cancer particularly the squamous undifferentiated type.

d- The polycylic aromatic hydrocarbons have been recognized as highly carcinogenic to the laboratory animals and the paranasal sinuses (Lloyd, J.W. 1971).

e- Nickel refining has been reported to increase the incidence of upper respiratory tract cancers among its workers. This effect is related to the earlier stage of refinery involving exposure to dust from relatively crude ore. (James Y. Suen & Eugene Myers 1981).

f- Chromates have been also blamed to increase the risk of respiratory tract cancer.

g- Wood and leather dust: Wood workers in the furniture industry of Oxfordshire, England, were shown to have a high incidence of adenocarcinoma of the nasal cavity and paranasal sinuses which was estimated to be 500-fold.

A similar increase was noted in the workers of the shoe industry but is confined to workers in dusty operations and foot wear repairers. Probably, the causative agents are chromium aldehyde compounds and aflatoxin (fungus metabolite).

e- Some other miscellaneous agents have been blamed to carcinogenicity in some occupations such as isopropyl alcohol, vinyl chloride, newspaper printing ink and certain textiles, but this lacks good evidence.

II- **NUTRITIONAL AGENTS:**

- Nitrosamines are carcinogenic and their precursors are present in our food and may be produced through cooking or storage techniques.

- Polycyclic aromatic hydrocarbons are present in air but also in food.

- Some food additives are suspected to be carcinogenic. Dyes used to color food and certain labeling products have been shown to contain aflatoxins.

III- **RADIO-ISOTOPES:**

Thorotrast was used in the past in radiographic studies of the maxillary sinus and when injected in the sinus, it decays to mesothorium which is capable of inducing neoplsia. Many authors reported this effect. Vianna et al (1982) studied the incidence of sinus cancer in New York state over a 30 year period from 1949-1978 and noted that a mean induction period of 15 years was usual.

Radium used for therapeutic reason was reported to cause high incidence of osteogenic sarcoma and paranasal sinus carcinomas. (James Y. Suen & Eugen Myers 1981).

IV- **SMOKING:**

Cigarette smoking plays a major nondebatable role in the development of cancer of the oral cavity, respiratory tract and to a lesser extent oesophagus and bladder.

A link between cancer of the hard palate and reverse smoking, that is burning of the lighted end of cigarette inside the mouth, has been noted in India, Venezuella and Panama (Keane et al 1981).

The use of snuffs which contain high amounts of chromium and nickel by the South African Bantu have been linked to an increased incidence of nasal cancer and antro-ethmoidal cancer (D.F.N Harrison, 1979).

McGuirt (1983) revielwed 533 cases of cancer head and neck and found that 73.7% of cases were tobacco users.

The carcinogenic element in tobacco has not been clearly identified. Polycyclic hydrocarbons and nitrosamines found in tobacco smoke have been blamed to be carcinogenic to laboratory animals. Also the microsomal enzyme system, of which is acylhydrocarbon hydroxylase, has been linked to carcinogenesis by some authors.

V- IMMUNOSUPPRESSION AND ONCOGENIC VIRUSES:

It is now well established that patients, who are chronically immunosuppressed are at greater risk for the development of de novo malignancies. Several large series have reported the incidence of malignancy ranging from 2-13% following transplantation. This incidence increases progressively with increased survival. Patients surviving 5, 8 and 10 years post-transplantation had incidences of 34%, 39% and 57% respectively for the development of de novo malignancies.

Skin cancers especially those involving the head and neck are increased by 4-21 fold being more in areas of high sun exposure. Non-Hodgkin's

lymphomas are increased by 45-100 fold over the incidence in the general population. Squamous cell carcinoma, as in the general population, forms 90% of malignancies.

Harris and Penn (1981) analysed 1081 neoplasms reported in 1023 patients amassed by the Denver Transplant tumour Registry and concluded that malignancies of the upper airway and related structures, including the maxilla, are increased in transplant patients. Most of this increase was in cancer lip. They also noted that patients were at younger age at the time of diagnosis than in the general population (26-63 with an average of 41.8 years compared to 55-65 years in the general population). Male involvement was 88% in immunosuppressed patients compared to 70% in the general population.

Harris and Penn suggested the possible causes of these malignancies to be disturbances of immunity, a direct oncogenic effect of the immunosuppressive agents especially cyclophosphamide and azathioprine, co-oncogenic effects of these drugs, activation of oncogenic viruses, genetic susceptibility or varying combinations of these factors.

Also immunosuppression increases the risk of acquiring Kaposi's sarcoma (idiopathic multiple haemorrhagic sarcoma).

Abemayar and Calcaterra (1983) reported the great increase of occurrence of Kaposi's sarcoma (KS) in patients who have undergone renal transplants and in patients receiving steroid and other immunosuppressive therapy.

Since June 1981 an epidemic of the rapidly fatal African or lymphadenopathic form of KS, simultaneous with an outbreak of community acquired immune deficiency syndrome (AIDS), have appeared among homosexual, bisexual men and women, intravenous drug abusers, Haitians residing USA and patients with haemophilia. Abemayor and Calcaterra found 37% of 590 cases with AIDS were having KS. In a series of 45 patients with AIDS they reported 18 cases with KS of the head and neck of which 4 cases were located in the hard palate. They also noted several interesting relationships between KS, the immune system and viruses. Inconclusive evidence has been presented implicating cytomegalovirus (CMV) in the pathogenesis of KS. The CMV viral particles have been observed in cell lines derived from African patients with KS. In alddition, elevated antibody titers to CMV have been noted in patients with KS. More recently, integration of CMV gene sequence into cellular DNA of KS tissues has been demonstrated.

The aetiological agent of AIDS is a retrovirus: "lymphadenopathy Associated Virus" or "Human T Lymphotropic Virus Type III", designated at present by the name "LAV/HTLV-III". (WHO Weekly Epidemiological Record No. 17, 1985).

Immunosuppressed patients have been shown to develop infections with potentially oncogenic viruses which include Epstein Barr, Herpes Hominis I and II and Polyoma viruses.

In the general population, increased evidence has accumulated for the role of these viruses in the development of such malignancies as Burkitt's lymphoma, nasopharyngeal carcinoma and carcinomas of the lip, skin and cervix.

The relationship of Burkitt's lymphoma and the Epstein Barr Virus (EBV) was first discovered in 1965.

The mechanism of how viruses induce cancer remains unclear. The chromosomes of these viruses become part of their host's chromosomes and participate in subsequent coding for DNA replication. This leads to the production of viral specific proteins such as the T and E antigens. These proteins may further directly stimulate the production of cellular DNA or interfere with a normal cytostructural component. They may mimic a normal mitogenic activity at the cell surface transforming the architecture of the normal cell to that of a tumour cell.

VI- CHRONIC SINUSITIS:

DFN-harrison (1979), Batsakis (1981) and James Y. Suen and Eugene Myors (1981) considered chronic sinusitis among the causes of cancer maxilla,

but this was considered by other authors to be of little significance. Batsakis explained that metaplasia occurring in chronic infection is the cause of cancer.

VII- TRAUMA AND OROANTRAL FISTULA:

Trauma was reported in some studies to have predisposed to malignant neoplsia especially vascular ones and osteogenic sarcoma (Batsakis 1981).

Long standing oroantral fistula was considered also by Batsakis (1981) to be a possible cause of cancer but this lacks good evidence at the present.

PATHOLOGY

INCIDENCE:

Malignant neoplasms of the nose andparanasal sinuses comprise 0.2-0.8% of all body malignancies and 3% of cancers involving the upper aerodigestive tract. Approximately, 80% of the cancers of the paranasal sinuses arise in the antrum.

Carcinoma of theparanasal sinuses represent less than 15% of all neoplasms of the upper respirtory tract. 80-90% are squamous cell carcinoma.

Incidence of carcinoma of the maxillary sinus is less than one case /200,000 people per annum (batsakis 1981).

The average age incidence of head and neck cancers in general is 59 years at the time of diagnosis, but patients with paranasal cancers have a less age average. (James Y. Suen & Eugene Myers 1981). Table (1) illustrates the age incidence in 773 patients with maxillary sinus carcinoma reviewed by Sakai et al (1983) during four periods between 1957 and 1979.

The male: female ratio in maxillary sinus carcinoma is 2:1. Table (2) illustrates sex distribution in the same series of patients reviewed by Sakai et al. This ratio was estimated as 4:1 throughout the 1940's to the 1960's and as 3:1 from 1964 to 1975 (McGuirt 1983).

Tarek K.(1977) reviewed 64 cases of malignant neoplasms affecting the nose and paranasal sinuses who were admitted to Kasr El Aini Hospital between 1972-1976 and found the commonest age affected to be between 30-49 years. Table (3) illuustrates age and sex distribution of these patients.

Table (1) Age Distribution in maxillary Sinus Carcinoma and Percentage to total.

Chronologic Period	No.of cases	-39	40-49	50-59	60-69	70-
1957-1966	282	38(13.5%)	58(20.6%)	85(30.1%)	77(27.3%)	24(8.5%)
1967-1971	191	14(7.3%)	28(14.7%)	68(35.6%)	57(29.8%)	24(12.6%)
1972-1975	166	14(8.4%)	25(15.1%)	41(24.7%)	61(36.7%)	25(15.1%)
1976-1979	134	11(8.2%)	30(22.4%)	36(26.9%)	38(28.4%)	19(14.2%)

Table (2): Sex distribution in Maxillary Sinus Carcinoma and percentage to total.

Chronologic period	No.of cases	Male	Female
1957-1966	282	181(64.2%)	101(35.8%)
1967-1971	191	109(57.1%)	82(42.9%)
1972-1975	166	89(53.6%)	77(46.4%)
1975-1979	134	85(63.4%)	49(36.5%)

Table (3): Age and sex distribution of malignant neoplasms of the nose and sinuses in Kasr El Aini Hospital.

Year	Age						Sex	
	-15y	15-29	30-49	50-59	60-	Total	Male	Female
1972	-	1	3	2	-	6	5	1
1973	1	1	6	2	5	15	10	5
1974	1	2	6	1	4	14	10	4
1975	1	1	5	2	2	11	7	4
1976	2	5	5	3	3	18	13	5
Total	5	10	25	10	14	64	45	19
Percent	7.18%	15.63%	30.06%	15.63%	21.87%	100%	2.77	1

PATHOLOGICAL FEATURES:

Squamous cell carcinoma comprises 80-90% of the malignant neoplasms of the maxillary sinus. Table (4) shows the histopathological distribution of malignnt tumours of the paranasal sinuses in 121 cases; of which 98 were located in the maxillary sinus, treated at Indiana University Medical Center during the period from 1960 and 1980. These cases were reviewed by Shidnia et al (1984).

Table (4): Histopathological distribution of 121 cases of cancer paranasal sinuses.

Histopathological type	No.of cases	Percentage
Epidermoid well, moderately,poorly differentiated	69	57.024%
Undifferentiated (including 5 transitioinal)	20	16.528%
Adenoid cystic	11	9.09%
Adenocarcinoma(including papillary)	7	5.785%
Lymphomas	6	4.958%
Plasmocytoma	4	3.305%
Osteosarcoma	1	0.826%
Malignant mixed tumour	1	0.826%
Rhabdomyosarcoma	1	0.826%
Benign papilloma	1	0.826%
Total	121	100%

In children, carcinomas and sarcomas of the paranasal sinuses occur with approximately equal frequency. Embryonal Rhabdomyosarcoma is the commonest sinus malignancy (Schromm 1979).

Epithelial Tumours:

I- **Squamous Cell Carcinoma:**

This represents 80-90% of all malignancies of the antrum in adults. Metaplasia from the normal respiratory epithelium to the squamous type, secondary to chronic irritation by carcinogenic agents, probably plays a role in the development of these tumours.

Gross picture:

A) **Carcinoma in situ:** This is an early stage of the disease when there is only thickening and induration of the affected epithelial surface.

B) **Infiltrating carcinoma:**

1) Polypoid (Warty) type: On this type, the tumour is formed of an irregular polypoid mass with a wide base. The mass is red or greyish in colour with its surface showing ulceration, necrosis and haemorrhage.

2) Malignant ulcer type: Here the tumour takes the form of an ulcer with a raised everted edge, a fixed indurated base and an irregular necrotic floor.

The polypoid type is commoner than the ulcerative type.

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Microscopic picture:

A) **Carcinoma in situ:** In this type, the cells show the characteristic features of malignancy (pleomorphism, hyperchromatism, frequent mitosis and loss of polarity), but no invasion of the basement membrane i.e. malignancy is confined to the surface epithelium.

B) **Infiltrating carcinoma:** Besides the malignant features in the epithelium, there is subepithelial invasion by infiltrating groups of epithelial cells of different sizes and shapes showing variable degrees of malignancy.

Spread:

Squamous cell carcinoma of the maxillary virus speeds mainly by direct infiltration of the contiguous structures. This is largely attributed to the intimate inter-relationships and unity between the nasal passages and the paranasal sinuses. The disease is limited to the maxillary sinus only in 25% of cases. Discernible bone destruction is found in 70-80% of cases. The tumour may extend through the innumerable foramina and fissures of the bones making up the sinus complex (Batsakis 1981). Invasion of the orbit occurs in about 45% of clases. Lymph node metastaxis is infrequent. Konno et al (1980) had 7.7% of patients with squamous cell carcinoma having cervical lymph node metastasis. The jugulodigastric and submaxillary are the most frequent sites involved.

Systemic metastasis is rare and was found to occur in 3.5% of patients treated by D.F.N. Harrison (1978), unless patient is terminal. 5-year survival is 20-30%.

II- **Mucous gland tumours:**

These malignant tumours arise in minor salivary glands and comprise 10-14% of all nasal and paranasal sinus malignncies. These tumours can be classified as follows:

1- Adenoid cystic carcinoma.

2- Adeno-carcinomas.

3- Mucoepidermoid carcinomas.

4- Malignant mixed (pleomorphic) tumours.

5- Acinic cell carcinomas.

6- Undifferentiated carcinomas.

Adenoid cystic carcinoma is the commonest type (42%) followed by adenocrcinomas. There is a lack of correlation of histologic types and clinical findings. Goepfert et al (1983) reviewed 66 patients with primary adenoid cystic carcinoma and primary adenocarcinoma of the paranasal sinuses and nasal cavity, treated at the University of Texas, M.D Anderson Hospital, Houston, between 1951 and 1980. They suggested a subclassification of these two types to satisfy a correlation between the histopathological findings and the course of the disease.

Adenoid cystic carcinoma:

This is composed of groups of small cells with defined borders and rare mitotic features. These cells are arranged in 3 patterns present in varying proportions: tubular, cribriform and solid. Male/female ratio is 11:7. Age between 20-80 years with a median of 50 years. Geopfert et al differentiated adenoid cystic carcinoma into low grade and high grade forms.

1- The low grade form is composed of a mixture of tubular and cribriform formations with less than 30% solid anaplstic areas, the cells are arranged in nests that give a "Swiss-Cheese" configuration.

2- The high grade form is composed of classic cribriform patterns mixed with more than 30% solid areas. The solid areas are composed of units of cells and few lamina. The cells are bland or atypical cells with high mitotic acivities.

Spread of both types of adenoid cystic carcinoma is characteristically perineural. Perivascular and bone invasions occur often without destruction of bony architecture and little osteolysis. Recurrence is common within 2-5 years after treatment.

Adenocarcinomas:

These can be subclassified into papillary (with or without mucus

production), mucoid (colloid), clear cell, ductal, terminal duct, poorly differentiated and undifferentiated, and neuroendocrine carcinoms. Goepfert et al also classified adenocarcinomas as low grade or high grade.

1- Low grade forms are composed of tubular and papillary structures and characterized by absence of solid masses,lack of severe cellular anaplasia and small uniform single layer glands. Cystic and papillary formations with psammoma bodies are present in varying degrees. Margins are pushing and not destructive.

2- High grade forms are arranged in a predominantly solid patterns with ill-defined glandular formations which are lined by striated epithelium with severe nuclear pleomorphism and increased mitotic rate. Margins are invasive and destructive.

Spread of adenocarcinomas is by intraneural and perineural invasion. It occurs in males more than females commonly in the 4th and 5th decades.

Spread:

Adenocystic and adenocarcinoma spread by blood stream to lungs, bone specially the spine and rarely to the kidneys and liver.

Prognosis:

Adenoid cystic carcinoma: 5-years survival 5-10%.

Adenocracinoma: 5-years survival 20%.

Malignant salivary gland tumours may arise primarily within the jaws as intra-osseous tumours. In this case they arise from metaplastic odontogenic cyst lining, ectopic salivary gland tissue or embryologically entrapped retromolar mucous glands (Batsakis 1981). The mucoepidermoid carcinoma has a proclivity among these tumours, and is an aggressive tumour with a rapid onset and frequent metastses. Lymph node metastases do occur.

Central intra-osseous epidermoid carcinoma may also arise primarily in the jaws and originate from epithelial remnants.

III- **Malignant melanoma:**

Malignant melanomas of the nose and paranasal sinuses are rare tumours and comprise about 1 percent of all malignancies of these sites. The disease occurs most often as a primary (Valerie and Lund 1982).

Sexes are equally affected with a peak age incidence in the fourth to sixth decades of life.

Gross picture is variable, ranging from large polypoid masses to firmer sessile lesions. The degree of pigmentation is also variable, some tumours are amelanotic. The tumour bleeds easily on touch and manifests local multicentericity known as satellites.

Histologically, this tumour presents as polygonal epithelial cells (melanocarcinoma), or spindle shaped cells arranged in whorling bundles (melano sarcoma). However, histological appearance is variable. To diagnose melanoma, Fontana stain for melanin should be positive, pigment should be bleached by permanganate oxalate.

There is early local recurrence, and extension with frequent metastases to lymph nodes and viscera.

Prognosis is usually poor, 30% of patients were alive for 5 years in the series of Valerie and Lund when there was absence of regional and systemic metastases and with vigorous surgery.

IV- **Olfactory neuroblastoma:**

This is a rare tumour making up 3% of all malignancies of the nasal cavity and paranasal sinuses. It affects the maxilla by direct spread from the lateral nasal wall. It occurs at any age being more between 15-35 years, with slight male predominance. It arises from the neuro-epithelial cells of the olfactory mucosa.

The tumour is classified according to the predominant cell type into neuroepithelioma, neuroblastoma and neurocytoma where true rosette, pseudorosette and neurofibrils predominate in that order. Characteristically, the tumour shows intercellular neurofibrillar matrix with clusters of small cells grouped around vascular space. There is tendency for local recurrence and a potential for local invasion and intracranial extensions as well as systemic metastases which occur in 20% of cases. 5-year survival and cure are 50% and 30% respectively.

Non-epithelial tumours:

All mesenchymal tissues can be the seat of malignant tumours which make up no more than 10 per cent of all malignancies of the nasal cavity and paranasal sinuses.

I- **Malignant lymphoma:**

Lymphomas represent 3-8% of all malignant tumours occurring in the nasomaxillary region. it is slightly predominant in children and adolescents. When considering the maxillary sinus alone, the incidence of malignant lymphoma drops to be an extremely rare condition with 46 cases in the literature up to 1975 (Batsakis 1981). Fierstein and Thawley added 8 caes in 1978. Sex ratio is 1.5:1 in favour of males.

The Rappaport classification of Non-Hodgkin's lymphoma is widely accepted. Table (5): shows the modified Rappaport (1977).

Table (5): The Rappaport Classification of Malignant Lymphomas (1977)

* Well differentiated lymphocytic.
* Poorly differentiated lymphocytic:nodular and diffuse.
* Histiocytic: nodular and diffuse.
* Undifferentiated.
* Burkitt's.
* Lymphoblastic.
* Mycosis fungoides.
* Immunoblastic.

With new immunologic and cytochemical tlechniques, the Rappaport classification could be modified to that shown in table (7).

Table (6): Histological classification of Non-Hodgkin Lymphomas.

Type	Marker
* Nodular (foilicular) pattern:	
* Lymphocytic, well differentiated	B-cell
* Lymphocytic, poorly differentiated	
* Mixed lymphocytic "histiocytic"	Heterogenous
* Histiocytic	
* Diffuse pattern:	
* Lymphocytic,well differentiated	B-cell
* Lymphocytic, intermediate differentiation	
* Lymphocytic, poorly differentiated.	
* Mixed lymphocytic "histiocytic".	Heterogenous
* Histiocytic	
* Undifferentiated pleomorphic (non-Burkitt)	
* Undifferentiated, burkitt type	B-cell
* Lymphoblastic (with and without convoluted)	T-cells
* Unclassified	

Microscopically, the tumour manifests a variety of histologic types in the various types of lymphomas. Thus, histiocytes, lymphocytes, lymphoblasts and reticulum cells are present with various proportions. The lymphoblast is twice the size of the lymphocyte with a larger nucleus while the reticulum cell is 2-4 times as large as the lymphocyte with round or oval shape, abundant cytoplasm and single, oval or round lobulated nucleus.

Grossly, the tumour presents as a grey, soft and friable mass with necrotic zones and occasional proximal sclerosis. Peripheral lymph nodes are sometimes involved. The prognosis of the disease is good especially when it is localized and with the administration of radiation therapy.

Burkitt's lymphoma:

This is now regarded as a separate entity of the disease. It was first described by Burkitt in 1958 in a larlge number of Central African children between 4-8 years of age. It proved later to be a worldwide disease but with less prevalence rate and is thought to be associated to Epstein Barr Virus (EBV). The disease can occur at any age especially in non-African cases.

The jaws are the most frequent sites involved. It occurs in 50% of African cases and in 15% of American cases (Batsakis 1981). The

The maxilla is involved three times more frequently than the mandible.

Grossly, the tumour tends to present as a nodular mass with encasement of the surroundings and secondary extension from capsules or surfaces.

Microscopically, the tumour presents a monotonous overgrowth of undifferentiated lymphoreticular cells. "Starry Sky" pattern is produced by uniform scattering of macrophages. Mitotic activity may be high and nuclei are prominent.

The tumour is the fastest growing neoplasm in man. The disease is multifocal, co-existing involvement of abdominal and/or pelvic viscera, retroperitoneal soft tissues, large bones, thyroid, and salivary glands, and central nervous system is frequent. Peripheral lymphadenopathy is rare.

The prognosis of untreated cases is rapidly fatal within 4-6 months of the onset.

II- **Malignant plasma cell tumours:**

These tumors are not uncommon occurrence in the maxilla. They are either multiple myeloma, solitary plasma cytoma of bone and extra-medullary plasmacytoma that may become a dissiminated disease, myelo-matosis.

The incidence of elxtramedullary tumours as compared to multiple myeloma is 1:40 (D.F.N Harrison, M.D. 1981).

Plasma cell is the secretory form of B-lymphocytes. Tumour cells produce an M(myeloma) protein.

Multiple myeloma:

It is not an uncommon disease of the jaws and represents 30% of all multiple myelomas (Batsakis 1983).

Grossly, the lesion appears as multiple, non-corticated small osteolytic lesions from 1 to 3 mm and not more than 1 cm in size.

Sex ratio is 1:1 and 5-year survival rate is 5.7%.

Solitary plasmacytoma of jaws:

This is a rare presentation of multiple myeloma. The peak age , incidence is in the 6th decade with male predominance. Dissimination always occurs. A paraprotein or M-band in serum or urine is usual & helps in follow-up.

Extramedullary plasmacytoma:

It is an uncommon tumour which presents outside the bone. 85% of all tumours occur within the head and neck primarily in the nose,

sinuses and nasopharynx. 10% of these tumours are multiple. It represents 4% of all non-epithelial tumours of this area.

Male female ratio is 4:1 and 3/4 of patients present between 40 and 70 years of age.

Gross appearance is non-specific and presents as fleshy, yellow gray to dark red, sessile, polypoid or pedunculated mass. Ulceration is not freuent and 25% of patients have cervical lymph node involvement.

Microscopically, the tumour consists of monocellular proliferation of plasma cells set in a very sparse matrix. Binucleate and trinucleate cell forms may occur.

The histologic behaviour of the disease is variable from localized, locally extensive, metastatizing and dissiminating.

III- **Chondrosarcomas:**

It is a malignant tumour of connective tissue origin arising from cartilage. 10% of chondrosarcomas arise in the maxillofacial area (Myers and Thawley 1979). They represent up to 4% of non-epithelial tumours of the nasal cavity, paranasal sinuses and nasopharynx. Sex ratio in maxillary lesions is 10:1 in favour of males with peak age incidence

in the 3rd to 5th decades. In the maxilla, the anterior alveolar region is the most common site.

Myers and Thawlay (1979) classified chondrosarcomas into 3 main types:

1- Primary chondrosarcomas, arising from undifferentiated perichondrial cells.

2- Secondary chondrosarcomas, arising from altered cells either in a central chondroma or cartilagenous exostosis.

3- Mesenchymal chondrosarcomas, arising from primitive mesenchymal cells.

Condrosarcoma is usually richly cellular with irregularity of cells and nuclei. Cells are plump with multiple nuclei showing pronounced hyperchromatism. Mononuclear giant cells are also present.

Chondrosarcomas can be graded histologically as such; a myxomatous change with cystic alterations in the tumour correlates well with a low or medium histological grades. An absence of cartilagenous lobulation and the presence of spindle cell form is characteristic of a high grade malignancy with poor prognosis.

Mesenchymal chondrosarcomas of the maxilla are rare occurrences. They are most common in the 2nd and 3rd decades of life. Sexes are equally affected.

Histologically, the tumour is richly cellular composed of undifferentiated mesenchymal cells in which islands of relatively well differentiated benign-appearing cartilage is found. The undifferentiated mesenchymal cells vary from uniform round and small cells to spindle shape cells. They tend to arrange themselves about vascular spaces.

Chondrosarcomas of the facial region metastasize infrequently and usually during the terminal stage of illness.

IV- **Rhabdomyosarcoma:**

This is an uncommon hihgly malignant neoplasm which is particularly rare in adults that can involve the maxilla and maxillary sinus primarily or by extension from the nose, orbit and the nasopharynx.

Generally, it is the most common soft tissue sarcoma in children having an incidence of 0.44/100,000 whites and 0.13/100,000 blacks, yet it is a rare adult disease. (Feldman 1982). This represents 5-15% of all childhood neoplasms. The paranasal sinus involvement is estimated by Suzuki et al (1984) to be 1.5% of the head and neck cases.

In Feldman's series of 83 patients with paranasal sinus rhabdomyosarcoma, age ranged from 10 months to 28 years with an average of 7 years.

The disease is thought to arise from embryonal mesenchyme and has been divided into 4 subgroups:(1) Pleomorphic, usually seen in adults (2) Alveolar, usually in adolescents 63) Embryonal, the most common variant in childhood and (1) Botryoid.

Recently, the International Society of Pediatric Oncology has adopted a classification into two types: adult (pleomorphic) with an apparent dedifferenttiation of mature skeletal muscle, and juvenile comprising components of embryonal, alveolar and botryoid elements with one element usually predominating. Another group is sometimes added, the mixed group comprising mixtures of cell types.

Grossly, all tumours, except the botryoid type, are soft, fleshy and ill-defined, pink to gray pink in colour and may be haemorrhagic or necrotic. They invade the surrounding tissue compressing it to a pseudocapsule.

In the embryonal variant, the predominant cell is long, thin and spindle shaped with a single central nucleus.

In the botryoid variant, multi-layered band of cells is located just below the mucous membrane. Below this layer, myxoid cells and dilated blood vessels exist.

In the alveolar variant, small round cells grow in cords or bands around central clear spaces resembling alveoli.

Pleomorphic variant, is uncommon in the head and neck. The tumour is locally very invasive with bone erosion with about 1/3 of cases involving the CNS while lymph node and distant metastses are infrequent being mainly to the eyes and bone.

Prior to 1960, the median survival at M.D Anderson Hospital and Cancer Instiute was 19.2 months with no survivors amongst patients with extensive or metastatic disease at the time of diagnosis. This is due to lack of anatomicl confines and presence of abundant lymphatics.

V- **Odontogenic tumours:**

Most of odontogenic tumours are benign. Ameloblastoma, though benign, is locally very aggressive so considered an intermediate tumour. Malignant odontogenic tumours are most often malignant changes in benign ones. There are squamous cell carcinoma arising in an ameloblastoma, ameloblastic fibrosarcoma and ameloblastic odontosarcoma. (Batsakis 1983).

Ameloblastoma:

Previously termed adamontinoma, the ameloblastoma occurs frequently in the mandible (80%) than in the maxilla (20%), representing

about 0.1 per cent of all sinus tumours. They are much commoner in African people. Almost half of the lesions occur in the molar region, one third in the antrum and the remainder at other sites including about 2% in the anterior maxilla (Batsakis 1983).

Ameloblastoma occurs in 2 variants: 1- Extra osseous or peripheral ameloblastoma is a soft tissue variant occurring in soft tissues overlying the tooth bearing region. 2- Intraosseous or central ameloblastoma involves the maxilla in 20% of cases. The maxillary ameloblastoma is much more locally aggressive and invades the nasal cavity, paranasal sinuses, orbit, pharyngeal tissues and the vital structures at the base of the skull.

The average age at diagnosis is 39 years but can occur at any age.

The tumour is benign and loclly aggressive. 4.5% of patients develop systemic metastasis.

The site of origin is doubtful but the epithelial debris in the periodontal membrane and the epithelial lining of follicular cysts are suggested.

Histological appearance is either follicular with islands of epithelium in fibrous stroma, or plexiform with strands and cords of epithelium intermingling with islands of mature fibrous stroma.

Metastases via lymphatic, blood or aerogenous routes may occur rarely in longstanding tumours with repeated surgical and radiation therapies.

Calcifying epithelial odontogenic tumour (Pindborg tumour):

It is an extremely rare tumour with intermediate malignancy invasive and locally recurrent. It is commoner in the mandible.

Microscopically, the tumour shows cellular pleomorphism, extracellular matrix and calcification. It appears as sheets of large polyhedral epithelial cells separated by scanty connective tissue stroma. The cells are binucleate or trinucleate with disintegrated cell membranes. Psammoma bodies may be present.

VI- **Malignant vascular tumours:**

These are exceedingly rare tumours in which the cells of origin and degree ofdifferentiation vary considerably.

1- **Hemangioendothelioma:**

This is a malignant tumour arising from endothelial cells. I.E Coles (1982) reviewed the literature from 1937-1980 and recorded only 14 caes involving the nose and paranasal sinuses, 7 of these involved the maxillary sinus. Two histological features exist; abnormally great number of atypical endothelial cells and formation of vascular tubes in a delicate framework of reticulin fibres.

2- **Hemangiopericytoma:**

This is a rare vascular tumour that arises from a special cell "pericyte" found outside the capillaries. It is particularly rare in the maxilla. R. Güdrün (1979) reviewed all casesfrom 1942-1978 and reported 14 cases in otolaryngology of which 6 involved the maxilla.

Microscopically, these tumours show a vascular architecture and composed mainly of elongated cells with sparse nuclear or cytoplasmic pleomorphism and low mitotic activity. Scattered mast cells are always present. A few collagen and reticular fibres are present and are quite regular in architecture. Grossly the tumour has no specific features, and is greyish red in colour with an irregular surface and firm elastic consistency.

The criteria for distinguishing benign from malignant tumours remain controversial but some authors consider all hemangiopericytomas

to be potentially malignant, although those involving the sinonasal tract are low-grade tumours.

Metastasis by lymphatic or hematogenous routes occur, in 5% of cases, to the lungs, bones, liver and local lymph nodes. Recurrence rate is 25%.

This tumour resembles very much hemangioendothelioma. They differ microscopically in that the endothelial cells are gouped inside the vessel walls in hemangioendothelioma and outside in hemangiopericytoma. Also, the endothelial cells in the latter are esentially normal while those in the former, occur in excess numbers, have voluminous cytoplasm with many processes and irregular border.

3- **Angiosarcoma:**

This refers now to a poorly differentiated malignant vascular tumours where no endothelial cell participation occurs. It is extremely rare in the maxilla, only 4 cases have been reported in the literature upto 1979 (Sharma and Nawalkha 1979).

The histological features are variable with predominance of conglomerates of atypical capillaries and marked tendency to anastomosis. Capillaries are lined by rounded or elongated anaplastic endothelioblasts.

Grossly, it is usually a solitary, firm, bulky tumour which infiltrates the surroundings, but bones and tendons are resistant to invasion. The growth is moderately rapid; haemorrhagic cysts, necrosis and mucoid degeneration are common. Distant metastasis occurs via blood stream and lymphatics.

4- **Angiofibroma:**

This is a highly vascular, locally invasive non-capsulated tumour found almost exclusively in the nasopharynx of adolescent males. It is considered an intermediate tumour and can involve the maxilla by direct spread from the primary tumour either forwards to occupy the maxillary antrum, or laterally through the pterygopalatine fossa then directly forwards into the antrum. The incidence of lateral extension lies between 20-53% (R.N. Patil 1982).

VII- **Osteosarcoma:**

This is a relatively rare tumour of the jaws representing 6% of all osteogenic sarcoms which themselves have an incidence rate of 1/100,000. The mandible is more affected than the maxilla. In the maxilla, the alvealar ridge is a frequent point of origin. Age of diagnosis is often within the third and fourth decades.

Irradiation, trauma and pre-existing bone disorders as fibrous dysplasia and Paget's disease have been blamed by some authors as

predisposing conditions. Viral aetiology has also been suggested by Vener et al (1984).

The microscopic appearance is considerably variable. The essential proliferating and infiltrating stroma is formed by spindle, oval or polyhedral cells with hyperchromatic nuclei. The sarcomatous connective tissue stroma is best seen in the periphery. The central position tends to be richer in osteoid matrix. Individual sarcomas may have abundant cartilage. Telangiectatic areas are not unusual. Late metasis occurs in about 37% of cases and is usually hematogenous to the lungs and brain.

Parosteal osteogenic sarcoma is a very rare variant that may occur in the maxilla.

The median survival for patients with the disease in the maxilla is 2.9 years being more with antral neoplasms (Batsakis 1981). The 5-year survival ranges from 19-37%.

VIII- **Fibrosarcoma:**

Fibrosarcomas are relatively uncommon tumours that affect both soft tissues and bone. They constitute 0.5% of all tumours and 5.5% of the malignant soft part sarcomas. The maxillary antrum is the second most frequent site of fibrosarcoma.

Histologically, fibrosarcomas of bone have features like those of its soft tissue counterpart. They are either differentiated or undifferentiated.

The differentiated type constitutes 75-80% of soft part fibrosarcomas. It infiltrates and recurs after removal but only rarely spreads by metastasis. It manifests as interwoven texture of differentiated cells and fibers. The fibroblasts appear relatively uniform in size and shape, do not display much hyperchromatism and lie surrounded by well developed collagen. They are arranged in bands and bundles. Mitoses are seldom seen.

The undifferentiated type constitutes 20-25% and in addition to locally aggressive behaviour, it also manifests a propensity to metastasize. It is richly cellular. Intercellular matrix, fibroblastic products are relatiely sparse. Mitoses are frequent. The pleomorphic giant cell sarcoma represents the epitome of the poorly differentiated type.

In gross appearance, the tumours are generally densely firm gray-white masses. When highly celluar or myxomatous foci are present, the tumor may manifest zones of softening and fluctuance.

Radiologically, the tumour has a non-specific feature of radiolucent defects of variable sizes.

Fibrosarcomas occur predominantly in males at any age group but most commonly in the fourth through the sixth decades.

Recurrence occurs in 30-60% of cases after treatment. According to Batsakis, the potential sources of osseous fibrosarcoma of the maxilla are the periosteal soft time, the periodontal membranes and the endosteum.

Tumours of the periosteal origin are more malignant, exihibit greater tendency to invade bone, more tendncy to local recurrence and more tendency to distant metastasis than the soft tissue type.

IX- **Fibrous histiocytoma:**

This is a rare tumour of the maxilla. Blitzer et al in their review of the literature through 1981 reported 15 caes involving the maxillary sinus and the maxilla out of 87 caes of the head and neck. In 1981 Mugliston and Shaw added 2 cases and in 1983 Sasaki et al added one case. It may involve the maxillary sinus or the maxilla itself.

It occurs most commonly in males in the 6th decade. When arising in the bone, it is highly lethal and is diagnosed by exclusion of other definable bone malignancies.

Histologically, the tumour presents multinucleated malignant giant cells as a constant finding. Fibrosis varies from prominent to

scanty. A storiform pattern of the fibrogenic cells is frequently present. Foamy (lipid containing) cells may be present and appear to be histiocytes or modified fibroblasts. Tumour necrosis, inflammation and numerous mitosis are frequent.

According to the predominating histologic backgrounds, the tumour is divided into 4 variants : storiform, inflammatory, myxoid and pleomorphic (Sasaki et al).

This tumour (especially when arising in bone) has a tendency for local invasion and metastasis via hematogenous and lymphatic routes to lymph nodes, bone, lungs and brain.

X - **Neurogenic tumours:**

Other than olfactory neuroblastoma, already discussed, the neurogenic tumours affecting the maxilla are:

Schwannoma:

Although benign, schwannoma malignant variants occur. Affection of the paranasal sinuses is quite rare with only sporadic cases reported in the literature.

Schwannomas arise from the Schwann cells of myelinated nerves. Malignant schwannomas probably arise from perineural fibroblasts.

In the nose and paranasal sinuses, these tumours arise from intranasal nerves, the ophthalmic and maxillary branches of the trigeminal nerve, and branches of the autonomic nervous system; the olfactory nerve contains no Schwann cells.

Two characteristic histologic forms exist: 1- The palisading (compact stroma) or Antoni A type and 2- The whorling (loose fiber) or Antoni B type.

Neurofibroma:

This tumour also arises from Schwann cells and is often benign but malignant variants occur especially when associated with Von Recklinghuman disease and these carry a poor prognosis with a 5-year survival of 30% compared to an overall 5-year survival of 65.7% (Shugar et al 1982).

Meningioma:

This is most often a benign tumour but malignant variants occur. It arises from the same cells that give origin to the arachnoid villi and endothelium. Malignant meningiomas represent less than 1 percent of all meningiomas and are diagnosed by unusual cellular activity, local infiltration and systemic metastasis.

Nasal, paranasal and nasopharyngeal involvement occurs as extracranial presentation of 3% of intracranial cases.

XI- **Fibro-osseous disease of the maxilla:**

Fibro-osseous lesions cover a group of conditions where normal bone is replaced by collagen, fibroblasts and varying amounts of osteoid tissue.

Ossifying fibroma:

This tumourarises from the fibrous connective tissue of the periodontium which contains mesenchymal blastic cells with the potential to form cementum, alveolar bone and fibrous tissue.

Sexes are equally affected with the greater occurrence in the 3rd to 4th decades of life.

Microscopically, ossifying fibroma presents spaced spicules of bone rimmed with osteoblasts and osteoclasts within a fibrous stroma. Most of the spicules are centrally composed of woven bone with evidence of a lamellar transformation. A prominent feature is increased denseness of the stroma with rounding of the fibreblasts near the bone spicules.

The tumour is of intermediate behaviour and is potentially malignant. It turns frankly malignant with exposure to radiation.

Fibrous dysplasia:

This is an uncommon fibro-osseous lesion of the jaws which occurs predominantly in the maxilla.

Smith (1965) described 3 types of fibrous dysplasia according to activity: 1- The first or active form is characterized by a richly cellular connective tissue matrix containing numerous fusiform or stellate cells. Mitoses and intercellular collagen exist. Well-defined islands of bone with a characteristic scroll edge occur in various "jig saw puzzle" sizes. It is commoner in young patients. 2- A quiescent or "potentially active" form of fibrous dysplasia is seen in adolescents where more mature matrix, no mitoses and more prominent bone exist. 3. Inactive form which is least common with degeneration of matrix.

Though locally aggressive, frank malignancy may develop with sudden growth in a quiescent area. This is more prone to occur if the disease is irradiated.

Central fibroma:

This term comprises multiple variants. The following are the variants concerned with the maxilla: odontogenic fibroma and fibromyxoma. Both tumours are rare, locally aggressive and recurrence is a prominent feature of the fibromyxoma. Odontogenic fibroma arises from one of the mesenchymal components of the tooth or tooth germ,

such as the periodontal membrane, dental papilla or dental follicle. Non-odontogenic fibroma arises from the endosteal mesenchyme of the jaw itself.

XII- Myxomas:

These are mesenchymal tumours which may arise from soft tissue or bone.

Myxoma of bone is a relatively uncommon tumour. Histologically, it has been considered by most authors to have an odontogenic origin. This is supported by its almost exclusive occurrence in the jaws, its striking resemblance to dental papillae and the odontogenic rests present within the tumour. This is further supported by ultrastructural findings (Batsakis 1981).

Myxoma of the jaws is most common between the ages of 10 and 29 years with no sex predilection. Invasion of the maxillary sinus and destruction of the antral walls often occur. The zygomatic process and alveolar bone are primarily affected.

Grossly, the lesion has a pseudocapsule. Glistening mucus covering the cut surface of the tumour is an important finding. The mass is soft, shiny, smooth, yellow or whitish gray and gelatinous. There may be clacified regions.

Microscopically, polyhedral or stellate cells embedded in a soft, mucinous matrix are characteristic of the tumour. Stellate cells dominate with long anastomosing processes. Nuclei areoval and often hyperchromatic. The stroma is loose with course delicate reticulin fibers. The clinical behaviour is not unlike soft tissue myxoma i.e. slow progressive growth and a stubborn tendency for resistance or recurrence as its boundaries are not well defined. Recurrence rate is 25%.

XIII- Other extremely rare tumours include:

Chordoma:

This is a dysodontogenic neoplasm of intermediate malignancy that arises in the residual or vestigial remnants of the embryonic notochord. Involvement of the maxilla is considerably rare and occurs either primarily or by ventral extension from the spheno-occipital type to the maxillary antrum. Chordoma occurs at any age but most frequent between the ages of 20-40 years and is dominant in males.

Grossly, chordoma is a lobulated, partially translucent and mucoid tumour. The mass is non-encapsulated or has a pseudocapsule and invades bone with facility.

Microscopically, four findings are fairly constant:

(1) Lobular arrangement of cells.

(2) tendency of the cells to grow in cords, irregular bands or in a pseudoacinar form.

(3) Production of an abundant intercellular mucinous matrix.

(4) The presence of large vaculated cells.

Incomplete fibrous trabeculae continuous with copressed peripheral connective tissue, traverse the intercellular matrix and further subdivide the lobules. Infiltrative or peripheral lobules have a less mucoid matrix, better formed cells and less intracellular vacuolation.

The slow, yet progressive growth with infiltrative destruction of bone and the almost inevitable recurrence account for the bad prognosis of chordoma.

Ewing's sarcoma:

It is a primary malignant tumour arising from undifferentiated mesenchymal cells. Ewing's sarcoma of the maxilla is extremely rare and sporadic cases are only encountered (Batsakis 1981).

The histological criteria are distinctive. The tumour is cellular, compact with sheets of round or slightly elongated cells. The sheets of cells may be separated by strands of fibrous tissue. The nucleus fills the cells and the nuclear membrane is distinct. Zones of necrosis and haemorrhage may exist. Occasionally, the cells are arranged in a

rosette-like pattern. Intracytoplasmic glycogen granules are usually present, a significant element in diagnosis.

Ewing's sarcoma manifests a rapid hematogenous spread primarily to the lungs and other bones.

Liposarcoma:

It is malignant tumour of the adipose tissue. Liposarcomas of the maxilla are very rare.

The gross appearance is variable depending upon the histological composition i.e. myxoid components, fibrous elements, vascularization, necrosis...etc. It manifests an apparent circumscription and may give the impression of encapsulation. Stellate nodules about the main mass are very common.

Microscopically, liposarcoma is subdivided into: (1) well differentiated type (2) Myxoid type, (3) round cell type and (4) pleomorphic type.

The better differentiated liposarcoma often presented a jelly-like moist appearance, the less differentiated varieties present with a soft "brain-like" consistency. Hemorrhage and necrosis are common.

Recurrence is frequent and the 5-year survival for the myxoid and well differentiated forms exceeds 70%. This is 20% with the pleomorphic and round cell types.

Malignant mesenchymoma:

This is a considerably rare occurrence in the maxilla. To be diagnosed, a neoplasm must be composed of two or more mesenchymal elements, other than fibrous connective tissue elements, not originally found together. Rhabdomyosarcomatous and vasoformative malignant elements are the most common.

Extraosseous chondrogenic and osteogenic tumours:

These extremely rare tumours arise without concomitant involvement of adjacent bone.

Both diseases are sporadic, Goldman and Perzik (1967) reported the only example of an extra-osseous chondrosarcoma of the maxilla.

Leiosmyosarcoma:

This is a quite extremely rare occurrence in the paranasal sinuses. Kawabe et al (1969) reviewed five published cases of leiomyosarcoma of the nose, paranasal sinuses and nasopharynx. They also added two cases of the maxillary sinus. Kakar et al (1979) added two other cases of which one has involved the maxilla.

This arises from smooth muscles which are lacking in the nasal sinuses. Its source could be aberrant undiffierentiated mesenchyme, walls of blood vessels or both.

Oncocytoma:

This is a low grade malignancy tumour of minor salivary glands. It arises from the oncocyte which is a special type of cell with swollen granular cytoplasm. Handler and Ward 61979) reported the only case confined entirely to the maxillary sinus while N.A. Mahmoud (1979) reviewed only 3 cases of nasal cavity, which extended to involve the maxilla, and reported a fourth case.

Microscopically, the tumour is composed of nests, cords, and acinar - like arrangement of cells with abundant eosinophilic cytoplasm. The small nuclei are uniform and mitotic figures are rare. No capsule is present and nests of cells infiltrate the loose fibrous stroma.

XIV- **Metastatic tumours in the maxilla:**

The possibility of a metastasis to the nose or paranasal sinuses is rare and estimated to be about 1% of malignancies of this area. These metastatic tumours are almost totally carcinomata. Metastatic bone sarcoma to the maxilla is almost nill, H.B. Singh et al (1975) reportd the only known case, since then, of metastatic osteogenic sarcoma to the maxilla.

The relative low incidence of metastasis to the jaw bones is explained by the absence of red marrow where the thin-walled vascular channels provide a suitable channel for the enlodgement and proliferation of neoplatic emboli.

Metastasis to the bones of the jaws are almost always from regions below the clavicle.

Clauson and Poulsen studied 115 cases from 1884 to 1965, of which 22 cases involved the maxilla, while McMillan and Edwards (1975) reviewed 106 cases from 1966 to 1973. The sites of the primary lesions have been most often the kidney, lungs and breast in this order of frequency. Much less sites are the gastro-intestinal and distal urogenital tracts.

Between 20-30% of the group offered their jaw metastases as the first indication of harboring a malignant neoplasm. The average age of patients at the time of diagnosis was 56 years with a range 16-79 years. Prognosis is generally poor.

XV- Invasion of the maxillae can occur by tumours of the surroundings. Of these, the following are worth mentioning:

Squamous cell carcinoma of the head and neck can invade facial bones by direct spread. Carter et al (1983) studied morphological patterns

of direct infiltration of skull bones in 100 patients with squamous cancers of the head and neck. The overall incidence of direct bone invasion was 28%. The maxilla was involved in 7% of cases. They described the pattern of bone invasion as follows: the tumour gains access to bone principally by spread through contiguous soft tissues; perineural spaces may provide another route. The bone cortex has an irregular pitted surface and infiltrating tumour cells tend to spread into these defects. The local accumulation of host osteoclasts takes place and resorb bone in front of the advancing tumour. Then osteoclast response declines and tumour cells take over the destructive process.

In an in-vitro study, Carter et al came to a result that osteolysins including prostaglandins and non-prostaglandin elements are derived from tumour cells and from host stroma. The cellular source of these osteolytic factors can not be determined, but this raises the possibility that some aspects of bone invasion may be controllable by using the antagonistic drugs.

Another study was made by Panje and Ceilley (1979) on 150 cases of mid-face skin cancers by the fresh tissue technique of microscopic controlled excision. They revealed that local epithelial cancer spread of mid-facial tumours to the maxilla is markedly influenced by embryological fusion planes. So, large and aggressive tumours originating on

the lower eyelid and upper lateral lip tend to spread towards the cheek and invade deeply along the infra-orbital nerve into the maxillary sinus and orbital apex.

Kaposi's sarcoma has got a special attention due to its relation to the community acquired immune deficiency syndrome (AIDS) which occurs now in epidemics.

In a series of 45 patients with AIDS studies by Abemeyor and Calcaterra (1983), 18 patients (40%) had initial disease in the head and neck where the hard palate was the commonest site involved.

Kaposi's sarcoma consists of localized blue to purple nodules. The neoplastic process is manifested as multiple vascular tumours composed of proliferating connective, tissue cell and capillary vessels. It is believed to start at the mid-dermis level and extend to the epidermis. Cell of origin is unknown. The distinctive microscopical features are spindle cells and vascular structures embedded in a network of reticular and collagen fibers.

Classification:

Classification of tumours of the maxilla, as for tumours at any site, aims at facilitation of the exchange of information between centres and helping surgeons in planning treatment discipline. It also gives

a good indication of the prognosis of the disease and helps assessment of treatment results.

Most of the classifications were basically concerned with tumours of the maxillary sinus. As yet, there is no generally accepted classification for tumours of the nose or the paranasal sinuses. All classifications proposed are not suitable for a universal adoption without modification as they suffer from both intrinsic inaccuracies and an apparent failure to relate "T" categories (extent of primary tumour) to clinical experience of the spread of these tumours.

The first classification was proposed by Sebileau 1906. He divided the maxilla and its intimate connections into 3 regions:

1- The suprastructure: included the ethmoids, the orbital position of the superior maxillae and the malar bones.

2- The mesostructure: included the maxillary sinuses and the nasal fossa.

3- The infrastructure: included the hard palate and upper alveolus.

The precise levels of the partitions of these subdivisions are not clear. Huet and Baclesse (1952) developed this system where they designated each site by a letter.

Ohngren of Stockholm (1933) has suggested a classification based on the observation that tumours situated in the posterior superior part of the maxillary sinus, defined as being above an imaginary plane extending from the medial canthus to the angle of the mandible, carry a worse prognosis than those situated antero-inferiorly. This is primarily related to the difficulties of resecting the disease from the important structures related to this part of the maxillary sinus, such as the orbit, cribriform plate and pterygoid region.

In his classification, Ohngren divided the facial skeleton as seen in a profile section into 2 parts by line drawn from the inner canthus to the mandibular angle. The 2 parts are termed postero-superior and antero-inferior. Then he further subdivided these 2 parts into medial and lateral portions by a vertical line passing through the center of the orbit as seen on frontal section. 4 divisions are thus obtained, supero-medial, superolateral, inferomedial and inferolateral.

Both Sebileau and Ohngren classifications were really concerned with topographical classification of tumours of the maxillo-ethmoidal region, although Ohngren extended his classification so as to include histological variations of the tumours as well as the malignancy of the tumour. His lateral dividing line was considered a plane of malignancy. Tumours arising posterosuperior to the plane were considered as most malignant.

Ohngren's observations were correct but his classification was overcomplicalted in clinical practice, though it is a useful but rather rough guide to operability and prognosis.

Sisson (1963) developed the TNM classification as follows:

T: Primary tumour:

T_1 (a) Anterior wall of the antrum without skin involvement.

(b) Inferior nasoantral wall.

(c) Anterior medial palate.

T_2 (a) Invasion of the lateral wall without muscle involvement.

(b) Invasion of the superior wall without orbital involvement.

T_3 (a) Pterygoid muscle involvement.

(b) Orbital invasion.

(c) Invasion of the anterior ethmoidal wall without involvement of the cribriform plate.

(d) Invasion of the anterior wall of the antrum with skin involvement.

T_4 (a) Invasion of the cribriform plate.

(b) Invasion of the pterygomaxillary fossa.

(c) Invasion of the nasal fossa or other antrum.

(d) Invasion of the sphenoethmoidal recess or sphenoidal sinus.

(e) Invasion of the posterior ethmoidal cells.

(f) Invasion of the pterygoid plate.

N: Regional lymph nodes:

N_0 : No palpable lymph nodes.

N_1 : Mobile homolateral lymph nodes.

N_2 : Mobile contra-lateral or bilateral lymph nodes.

N_3 : Fixed ipsilateral or bilateral lymph nodes.

M : Distant metastasis:

M_0 : No evidence of distant metastases.

M_1 : Distant metastases.

G. Martensson (1965), submitted to the 8th International Oto-rhinological Congress a draft of a classification of carcinoma of the paranasal sinuses according to the TNM system as follows:

T: primary tumour:

T_1 : Tumour localized only to the mucous membbrane or to the periosteum.

T_2 : Tumour causing destruction of bone.

T_3 : Tumour with minimal infiltration into other anatomical structures such as to the floor of the orbit or the oral cavity.

T_4: Tumour with deep infiltration into other anatomical structures such as to the base of the skull and pterygoid fossa or the cheek.

N and M: as that of Sisson.

S. Sakai and Y. Hamasaki (1967) modified the classification proposed by G. Martensson as follows:

T_1 : Tumour localized only in the paranasal sinuses.

T_2 : Tumour with minimal infiltration into the surrounding tissues. The bone destruction of the posterior wall and the invasion into the nasal cavity is generously taken into consideration.

T_3 : Tumour with deep infiltration into other anatomical structures.

T_4 : Tumour with inoperatively extensive infiltration.

A more realistic classification covering the whole upper jaw, was suggested by Lederman (1970) based on his experience of approximately 350 patients. This classification has considerable merit and divides the region and sites into 3 regions by two parallel lines drawn across the frontal section of the skull, the upper line passing through the floor of the antra. These 2 lines are supplemented by 2 vertical lines extending down from the medial orbital wall on each side of the nasal floor.

The 3 regions delineated are:

1- The infra-structure or buccal portion of the upper jaw.

2- The meso-structure or nasal portion of the upper jaw. In this part of the antrum, ethmoid and nasal cavities border on one another, and because they are separated by extremely thin bony lamellae, the tumours arising in this region spread readily and render the

determination of their precise point of origin difficult.

3- The supra-structure or cranial portion of the upper jaw. Fig.(6) illustrates these division.

Three degrees of T for the primary squamous carcinoma can be recognized:

T_1 : Tumour limited to one sinus or one tissue of one tissue of origin e.g. turbinate, septum or nasal vestibule.

T_2 : Spread limited horizontally to the same region or two adjacent or two vertically related regions.

T_3: (a) Tumour involving three regions with or without involvement of the orbit.

(b) tumour extending beyond the upper jaw e.g. nasopharynx, cranial cavity, pterygopalatine fossa, skin, or buccal cavity.

N and M: as that of sisson.

Early mucosal lesions confined to the sinus properly belong to T_1 but T_2 can cause confusion in that extension to the orbit or ethmoid would be classified along with the less serious inferior involvement of sinus floor or hard palate. Similarly, either would be equated with horizontal extension to the nose below the level of the middle turbinate. Such comparisons are not compatible with clinical experience but cause a lot of confusion.

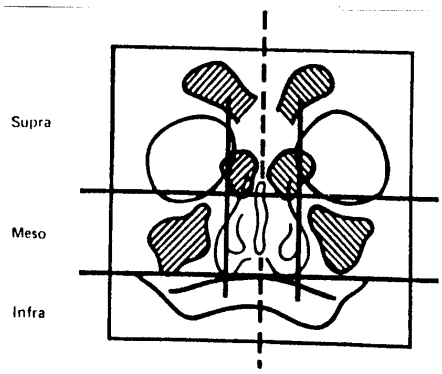

Fig.(6): Proposed TNM classification (Lederman) Regions and sites.

T_3 a is an uncommon clinical situation and could be well excluded leaving T_3 to cover all advanced cases.

Although not perfect nor entirely accurate, this classification has the merit of simplicity and being the only system covering the whole upper jaw.

Rubin (1972) suggested a classification of paranasal sinus carcinoma based on modifications of the early effort of Ohngren, Sisson et al and Dodd et al as follows:

T (tumour):

T_1 : Infrastructure site of neoplasm in the anterior and inferior compartments without bone destruction.

T_2 : Suprastructure location of neoplasm in the posterior and superior compartments without bone destruction: both compartments are involved.

T_3 : Radiographic evidence of bone destruction. Invasion of nares or cheek muscles, enlargement of the infra-orbital foramina or orbital destruction. No involvement of the oral cavity.

T_4 : Invasion of skin of the cheek; fistula may be present orbital and eye invasions, invasion of the oral cavity with fistula and extension to the ethmoid, sphenoid or frontal sinuses.

N (Lymph Nodes):

N_0 : No involvement of primary station lymph nodes (includes retro-pharyngeal, jugulodigastric and the submandibular and parotid lymph nodes).

N_1 : Palpable, movable involved nodes.

N_2 : contralateral or bilateral nodes.

N_3 : Fixed neck nodes.

N_4 : Nodes in the mediastinum or beyond primary station levels.

M (Metastsis):

M_0 : No metastsis.

M_1 : Distant metastsis.

More recently, two systems have been adopted by the American Joint Committee (AJC) in 1976 and the Japanese Joint Committee in 1977.

The AJC has adopted a system which utilizes Ohngren's line calling the region above, the suprastructure and that below, the infra-structure. Since Lederman's infrastructure is not really maxillary sinus, or at least very little of it, considerable confusion between these 2 systems evolves. Table (7) illustrates the classification adtopted by the AJC (1976).

Table : (7)

American Joint Committee 1976.

Definition of "T" Category of the

Maxillary Antrum

T_1 :	Tumour confined to the central mucosa of the infrastructure with no bone erosion or destruction.
T_2 :	Tumour confined to the suprastructure mucosa without bone destruction or to the infrastructure with destruction of medial or inferior bony wall only.
T_3 :	More extensive tumour invading skin of cheek, orbit, anterior ethmoidal sinuses or pterygoid muscle.
T_4 :	Massive tumour with invasion of the cribriform plate, posterior ethmoids, sphenoid, nasopharynx, pterygoid plates or base of the skull.

This system suffers some impracticability:

1- Since mucosal disease cannot be confined by the boundaries of supra- and infra-structures, it can be impossible to make sure by any means that any particular neoplasm is restricted as described in T_1. This also applies to mucosal involvement in T_2.

2- Involvement of pterygoid muscles is considered by some authors to be as serious as extension to the roof of the nose or nasopharynx. D.F.N. Harrison thinksit would be included within T_4.

The AJC has a little, modified this system in 1978 as follows in Table (8).

Table (8): American Joint Committee 1978
Modified system of classification of
the Maxillary Antrum Cancer.

Primary Tumour (T):

T_x : Tumour that cannot be assessed by rules.

T_0 : No evidence of primary tumour.

T_1 :
T_2 : as before
T_3 :
T_4 :

Nodal Involvement (N):

N_x : Nodes cannot be assessed.

N_0 : No clinically positive nodes.

N_1 : Single clinically positive homolateral node 3 cm or less in diameter.

N_2 : Single clinically positive homolateral node more than 3 cm but not more than 6 cm in diameter.

N_{2a}: Single clinically positive homolateral node more than 3 cm but not more 6 cm is diameter.

N_{2b}: Multiple clinically positive homolateral nodes, none more than 6 cm in diameter.

N_3 : Massive homolateral node(s), bilateral nodes, or contralateral node(s).

N_{3a}: Clinically positive homolateral node(s), one more than 6 cm in diameter.

N_{3b}: Bilateral clinically positive nodes (in this situation, each side of the neck should be staged separately; that is N_3b; right, N_{2a}; left, N_1)

N_{3c}: Contralateral clinically positive node(s) only.

Distant Metastases(M)

M_x : Not assessed.

M_0 : No (known) distant metastasis.

M_1 : Distant metastasis.

Stage Group:

Stage I: T_1 No Mo

Stage II: T_2 No Mo

Stage III: T_3 No Mo

T_1 or T_2 or T_3 with N_1 Mo

Stage IV: T_4 No Mo or T_4 N1 Mo

Any T with N_2 or N_3 with Mo

Any T with any N with M1

In 1977, the Japanese Joint Committee (JJC) proposed a TNM classification of maxillary cancer originally suggested by Sakai et al (1974) based on their study on 560 cases. This classification is shown in table (9). It differed a lot from his first system proposed in 1967. No boundaries, planes or subdivisions are employed, the maxillary sinus being defined as a simple anatomical entity.

Table (9): JJC proposal on TNM Classification of Maxillary Sinus Carcinoma.

T: Primary tumour:

T_1 : Tumour confined to the maxillary sinus with no evidence of bony involvement.

T_2 : Tumour causing destruction of bony wall, with the external periosteum remaining intact as the capsule and the surrounding tissue not involved but only compressed. Minimal infiltration into the ethmoid cells and the exophytic tumour in the middle nasal meatus is included in this category.

T_3 : Tumour infiltrlated deeply into the surrounding tissue by penetration of the external periosteum.

T_4 : Tumour extending to thebase of skull, the nasopharynx, the maxilla of the opposite side and/or the facial skin with ulceration. The extent to the base of the skull means not only the bony destruction

of the base of the skull but also deep infiltration to the orbit with markedly limited eye movement or visual impairment, extent to the temporal fossa with defect of the lalteral bony wall of the orbit or the zygomatic arch and/or infiltration to the pterygoid muscle with even slight defect of the pterygoid procless and marked trismus.

N: Regional lymph nodes:

N_0 : No palpable nodes.

N_1 : Movable homolateral nodes considered to contain growth.

N_2 : Movable contralateral or bilateral nodes considered to contain growth.

N_3 : Fixed nodes.

A separate table describing the clinical features of each T grade was also proposed by the JJC and will be shown in the chapter on clinical picture.

This system, though has given detailed description of its grades as not done before, has some intrinsic inaccuracies and vague description that may cause some confusion and may lead to variations in cure rates based on its grades. For example, clinical or radiological detection of bone destruction with an intact periosteum and compression without

invasion of all margins of the maxillary sinus (as mentioned in the classification) may be quite impossible. Imprecise descriptions as minimal infiltration of the ethmoid cells, fairly intact pterygoid process or minimal defect of bony wall are apt to variations in assessment and consequently in grading.

Harrison (1978) had a critical look at various classifications on basis of his study of 86 cases with squamous carcinoma of the maxillary sinus, he suggested the following T grouping that he thought will be more practical:

T_1 : Tumour limited to the antral mucosa with no evidence of bone erosion.

T_2 : Bony erosion without evidence of involvement of facial skin, orbit, pterygopalatine fossa or ethmoidal labyrinth.

T_3 : Involvemnt of the orbit, ethmoidal labyrinth or facial skin.

T_4 : Tumour extension to the nasopharynx, sphenoidal sinus, cribriform plate or pterygopalatine fossa.

This classification has the merit of omitting a lot of confusion and impracticability found in other systems, but still it is not perfect nor completely satisfactory as it suffers, like other classifications, from inaccuracies. In T_2, providing the surrounding structures are not grossly involved, bone erosion itself may not be significant and diagnosis

may not be possible until involvement has occurred in the sites excluded in T_2. In T_3, involvement of the orbit does not consider that involvement of the roof of the orbit could present very difficut technical problem that could make it nearer to T_4.

In addition, it is common for tumours within T_3 to have extended in other directions such as the ethmoids or pterygopalatine fossa and so differentiation between T_3 and T_4, unless the latter is massive, is not always possible. This can be misleading in treatment planning.

From the above discussions, one can conclude that accurate and satisfactory system of classification is not yet available. Further studies and much effort have to be done to realise this purpose as it will be a valuable achievement in planning standard modalities of treatment and getting accurate statistics of curability and survival by different treatment techniques.

CLINICAL PICTURE

The clinical picture of the malignant tumours of the maxilla and maxillary sinus depends on the primary site and the extent of the tumour. These malignancies, therefore, may present in a variety of ways and a useful concept is that the antrum looks like a box whose inner clinning first proliferates to fill it. Patients with maxillary cancer therefore rarely seek medical help until their neoplasmas are advanced. Besides, malignancies of the nose and paranasal sinuses often masquerades as a chronic inflammatory condition.

Most patients, therefore are not diagnosed until penetration of the surroundings has occurred. The presenting symptoms and signs, so, depend largely upon the direction of spread. Local spread will depend upon the primary site of origin.

Tumours arising in the infrastructure of the maxillary sinus may spread inferiorly into the alveolar process or the gingivobuccal sulcus producing pain and may be loosening of the teeth. Eventually mucosal ulceration of the palate and alveolus occurs. They may also spread into the soft tissues of the cheek below the zygoma causing pain or anaesthesia due to involvement of the maxillary divisions of the trigeminal

space, sphenoid sinuses, or base of the skull. Spread into the pterygoid space may involve the trigeminal nerve. Absent or decreased corneal reflex is usually due to involvement of the sphenopalatine ganglion.

It is unusual to find palpable lymph nodes when patients are first seen.

Lesions in the floor and anterior wall of the sinus drain into the digastric nodes or the nodes along the facial artery close to the submandibular salivary gland. The posterior and superior parts of the antrum drain into the retropharyngeal lymph glands which are inaccessible for examination. Later on both will drain to the upper deep cervical lymph glands.

Lymph nodes metastasise to the first stage lymph nodes (retropharyngeal), therefore cannot be diagnosed and only when the tumour has extended to skin, alveolar buccal sulcus or the pterygoid muscles do the second stage lymph nodes become involved.

Very rarely do patints with antral neoplasms present with demonstrable distant metastases.

The frequency with which the various manifestations may present in 648 patients reviewed by D.F.N Harrison between 1940 and 1964 is shown in table (10).

Table (10): Symptoms in 648 cases of carcinoma of the paranasal sinuses and nasal cavity from 1940-1964.

Nature or site	Percentage as initial symptom	Total percentage
Pain	17.4	40.9
Nasal obstruction	19.6	36.4
Purulent nasal secretion	15.7	30.6
Symptoms from the oral cavity	10.0	15.3
Dental symptoms	3.7	7.9
Swelling on cheek	11.3	33.6
Sanguineous nasal secretion	8.2	25.9
Ocular symptoms	4.6	18.2

Although early symptoms are rare, yet early tumour fortuitously situated close to naso-lacrimal duct or infra-orbital nerve may produce significant symptoms.

Chaudhry et al have divided the most important signs and symptoms of carcinoma of the maxillary antrum into five major groups.

Group I: Included oral signs and symptoms, these occur in 26% of cases and represented the first symptom in 15%. The first sign was localized or referred pain in the upper premolar or molar teeth. The next most common oral sign is loosening of teeth or alteration of their alignment. Other symptoms are swelling of the palate, the alveolar ridge or the gingivobuccal sulcus.

Group II: is comprised of nasal symptoms. In order of frequency they are: unilateral nasal stuffiness, unilateral nasal discharge and chronic epistaxis. Protrusion of the neoplasm into the nasal cavity is often seen.

Group III: is composed of ocular signs and symptoms. These occur in 23% of cases and represent the initial symptoms in 5%.

Group IV: includes facial symptoms.

Group V: includes all forms of neurological symptoms except those included in the other four groups. The majority of these are due to neoplastic involvement of the 7th and 8th nerves and the meninges. There may be unilateral facial paralysis and even hemiplegia and unilateral deafness.

In lymphomas of the maxillary sinus, nasal polyposis and palatal ulceration were the most frequent presentation according to Duncavage et al (1983).

In Burkitt's lymphoma, loosening of the deciduous teeth, expansion of the gingiva and displacement and distortion of the teeth are the earliest features.

In ameloblastoma, the presentation is usually with a rapidly growing swelling which starts usually in the cuspid and antral areas.

In rhabodomyosarcoma; nasal obstruction, mouth breathing, slow eating, bloody rhinorrhea are the most prominent presenting symptoms being more frequent in children.

Rubin (1972) had divided the insidious progression of cancer in the nose and paranasal sinuses into 4 clinical phases:

Phase I: The neoplasm is obscured by symptoms of a sinusitis and diagnosis requires drainage, access to the cavities and biopsy.

Phase II: Diagnosis relies mainly upon radiography while a dull ache may appear as the neoplasm erodes the wall of the nose and sinuses. Any area of bone destruction or asymmetrical sclerosis should be thoroughly investigated.

Phase III: It is characterized by fairly obvious signs of malignancy. Radiographic veiws are surely diagnostic. Hyposthesia, and deep pain herald nerve invasion.

Phase IV: There is gross and distant spread of the neoplasm with a major deformity. The diagnosis is obvious and prognosis is very bad.

The JJC, with the classification of maxillary carcinomas, has proposed a separate table describing the clinical features of each T grade. This is shown in table (11).

Rhadbomyosarcoma, being the commonest malignant tumour of the maxillary sinus in children has been classified clinically, by the Intergroup Rhabdomyosarcoma study (1977) into 4 groups based on the surgical resectibility of the tumour. This is shown in table (12).

Table (11): Main clinical features in each stage according to the direction of the tumour extension.

Direction of tumour extension	Grade of tumour extension by T category			
	T_1	T_2	T_3	T_4
Superomedial	Tumour without bony involvement	Mass in middle nasal meatus. Slight infiltration into ethmoid cells.Slight expothalmos with minimal defect of orbital bony wall.	Deep infiltration into ethmoid cells. Displaced eye ball	Markedly limited eye movements. Visual impairment. Defect of base of skull.
Superolateral	Confirmed by antrostomy	Defect of superolateral bony wall.	Defect of zygomatic bone	Defect of zygomatic arch. Defect of lateral wall of orbit-Extent to temporal fossa.
Inferomedial	---	Bulging of inferior nasal meatus	Bulging of floor of nasal cavity. Bulging of hard palate.	Extent beyond the midline.
Inferolateral	----	Bulging of upper gum. Detect of inferolateral bony wall.	Mass in upper gum	Wide infiltration to buccal mucosa.
Anterior	---	Swelling of cheek without adhesion. Bulging of canine fossa.	Swelling of cheek with adhesion. Defect of orbital rim.	Ulceration of facial skin.
Posterior	---	Rarefaction of posterior bony wall	Defect of posterior bony wall	Infiltration to nasopharynx. Infiltration to pterygoid muscles. Defect of pterygoid process. Defect of base of skull

Table (12): clinical staging of Rhabdomyosarcoma given by IRS.

Stage (group)	Extent of disease
I	Localized, completely resected (Lymph nodes not involved)
II	a. Grossly resected tumour with microscopic residual disease. No regional nodes involvement.
	b. Completely resected with regional node involvement and/or local extension.
	c. Disease and involved nodes grossly resected but with microscopic residuals.
III	Incomplete resection of biopsy with gross residual disease.
IV	Metastatic disease present at onset.

From all the above, a conclusion could be reached that most presentations are nonspecific, deceiving and confusion with benign diseases is quite common. A neoplasm should, therefore, be suspected when a patient has a unilateral nasal obstruction with bloody discharge, pain and swelling of the antrum. For early detection of cancer maxilla, it should be kept in mind that antral cancer is defnitely related and is often associated with diseases of the sinonasal tract which are usually benign. Also all polyps and masses of the nose and sinuses should be thoroughly investigated including biopsy.

DIAGNOSIS

Diagnosis of the tumours of the paranasal sinuses is a rather difficult thing and is usually late because it is very difficult to asses the extent of the disease, its spread bony destruction and even original site, even sometimes it is impossible to determine the extent of the disease during separation.

It is a fundamental concept that the accurate diagnosis of the extent of the tumour is essential in the proper management of the disease. Underestimations of the size and extent of the tumour leads to underexcision or failure to fully irradiate while over-estimation may mean unnecessary excision or expansion of the volume that requires irradiation.

The essential factor for early diagnosis of cancer maxilla is suspicion. The clues of early suspicion entail the patient's age, duration of symptoms and clinical examination. Once we suspect, it is imperative that full physical examination and a full program of investigations must be undertaken.

Thorough histories and pains taking, and methodical physical examinations support the diagnostic impression and provide direction for the treatment plan.

Good listening and interrogation of the patients complaint plus direction of the patient to tell symptoms that he may minimize or deny, may provide important clues.

Family history may reveal inherent weakness of immunity against cancer. A history of previous neoplastic disease and exposure to carcinogens including longstanding infection of the maxillary sinuses is important.

The nutritional status of the patient must be evaluated. A progressive loss of weight may result from distant metastasis to vital organs or from dysphagia caused by tumour mass.

A comprehensive physical examination should include evidences of weight loss and pain. The skin of the face must be examined for colour changes and ulcers. The face is evaluated for ptosis of eye lids, proptosis and asymmetry. The nasal fossa should be examined for crusting, ulceration, masses and infection. Palpation of the gingivo-buccal sulcus, palate and pterygoids is important evaluation of sinus tumours. Hypoesthesia of the maxillary division of the trigeminal nerve and displopia are important signs. The nasopharynx must also be carefully inspected. Examination of the neck for lymph nodes is essential.

The information obtained from the detailed history and physical examination is accomplished by the following investigations:

1- Radiographic imaging:
   a. Plain radiography.
   b. Polytomography.
   c. Computerized tomography.
   d. Intra-oral and occlusal films.
   e. Angiography.
   f. Radio-isotopic scanning.
2- Antral lavage.
3- Exploratory antrostomy (Mini-Caldwell Luc's operation).
4- endoscopy.
5- Biopsy.

These usually start with plain radiographic films in special projections. Opacity of the antrum necessitates proceeding to antral lavage and exploratory Coldwell Luc's antrostomy to confirm the presence of tumour. Afterwards, this proceeds to the other investigations.

I- RADIOGRAPHIC IMAGING:

At the time of presentation, the extent of the disease as assessed radiographically is usually greater than that assessed by clinical examination.

Radiological imaging of malignant disease of the paranasal sinuses

and the postnasal space consists of conventional radiographic imaging, that is plain x-ray and pluridirectional tomography (polytomography), and computerized tomographic (CT) imaging. Conventional radiography was the standard procedure before the advent of CT.

It must be emphasized that both CT and polytomography are not pathognomonic of sinus cancer as false negatives as high as 30% were encountered in cases studied by Sisson and Becker (1981).

(1) **Plain radiography:**

A- Malignant lesions involving the antro-ethmoidal sinus and nasal cavity:

The following positions are used:

1- Occipito-mental (Waters) projection (with mouth open) as a stereoscopic pair: This view is the best for the maxillary sinus as it is demonstrated unobstructed by the petrous portions of the temporal bones. Occasionally a canal-like lucency of the posterior superior alveolar canal is seen in the lateral maxillary sinus wall in this view. Fig. 7 shows a plain x-ray of the left maxillary sinus (Waters view) where there is a squamous cell carcinoma with bone destruction.

2- Occipito-frontal (Caldwell) projection with and without tube angulation: This is helpful for evaluation of the frontal and ethmoidal sinuses. It also reveals valuable information about the superior

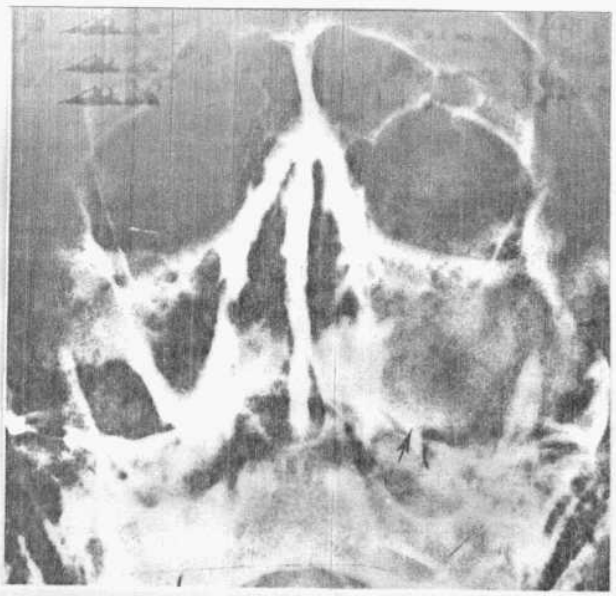

Fig.(7): Squamous cell carcinoma of the left maxillary sinus. There is soft tissue mass in the left antrum with complete destruction of lateral antral wall and portions of zygoma (arrows).

wall and the posterior medial apex of the maxillary sinus. Also the medial wall of the sinus is best seen in this view. The ethmoido-maxillary plate which is the boundary beween the ethmoid and maxillary bones is also best seen in this view. The infra-orbital canal appears in this view as a notch in the orbital floor.

Fig.(8) shows an x-ray of the right maxillary sinus (Caldwell view) where there is squamous cell carcinoma of right antrum with extension.

3- Lateral projection: It is also helpful in viewing the postnasal space, taken with the patient performing the Valsalva manoeuvre. It helps in evaluating the general size and contour of the maxillary sinus and related structures. The posterior wall of the sinus is particularly well shown in this ivew which allows also one of the best evaluations of the pterygomaxillary fissure.

fig.(9) shows a lateral view of the maxilla with osteogenic sarcoma.

4- Submento-vertical (SMV) projection: it helps as a stereoscopic pair. It is particularly useful for evaluation of the lateral wall of the maxillary sinus and related structures. The most medial portion can also be seen in this projection.

N.B.: The anterior sinus wall is not visualized well on any view. Both the lateral and SMV views show portions of this wall.

B- <u>Malignant lesions of the postnasal space:</u>

The following projections are in use:

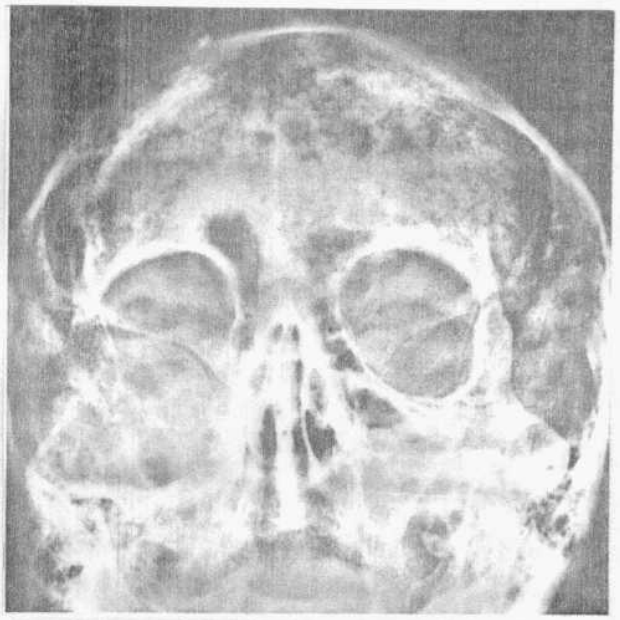

Fig.(8): Caldwell view of right maxillary sinus revealing squamous cell carcinoma with extension. There is extensive destruction of right orbital floor and lateral wall. body of right zygoma and right antrum are also destroyed.

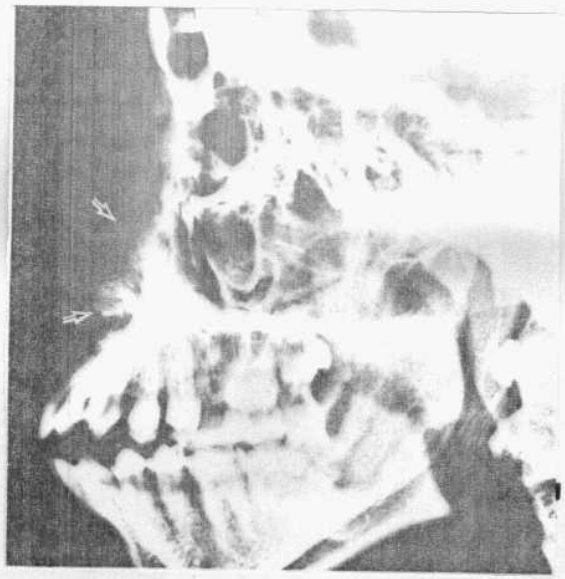

Fig. (9): Lateral skull view reveals "sunburst" periosteal reaction of maxilla characteristic of osteogenic sarcoma (arrows).

1- Lateral projections of the postnasal space to include the posterior nares and the antral wall.

2- SMV projections help as a stereoscopic pair. If the SMV projection reveals extension to the cranio-cervical junction, adjustment of the basal tilt will allow a more satisfactory view of this area.

(2) **Conventional pluridirectional tomography (Polytomography):**

For many years, polytomography has been in use for evaluation of otolaryngologic problems. The emphasis in polytomography has been on evaluation of the extent of bone destruction in an attempt to determine the extent of the lesion and indirectly get an information about its possible histologic nature.

A- Malignant lesions of the antro-ethmoidal sinuses and nasal cavity:

Sectioning is carried out at 0.25-0.5 cm intervals as is continued back to the level of the posterior ethmoids. If the posterior ethmoidal cells appear to be involved, sectioning is continued posteriorly to include the sphenoid sinus.

If the orbito-ethmoidal plate appears to be breached, or there is suspicion of early sprad to the maxillary antrum from the ethmoid sinuses, then linear tomography (25° angle of swing) is performed to demonstrate soft tissue more adequately.

fig. 10 shows a coronal tomogram that reveals a mesenchymoma involving the right antrum, nasal cavity and ethmoid sinus.

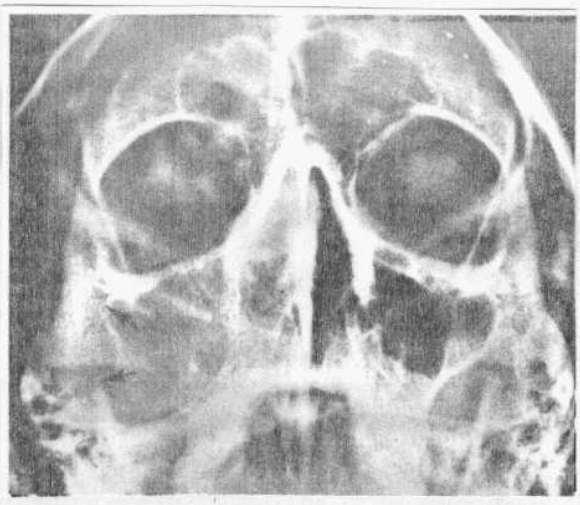

Fig. (10): Coronal tomogram reveals right antral, nasal cavity, and ethmoid mass that has destroyed inferior orbital margin medial and lateral antral walls. Mesenchymoma.

B- Malignant lesions of the postnasal space:

In these lesions, polytomography is undertaken if there is evidence of extension of the disease to the ethmoid sinuses, superiorly into the basi-sphenoid and floor of the sella or posteriorly along the skull base.

(3) **Computerized tomography (CT):**

This is a new non-invasive diagnostic tool developed in 1973 and considered the most important distribution to medical diagnostic techniques since roentegen discovered x-ray in 1895. In this technique the patient is scanned by a narrow beam of x-ray. The x-ray transmission is measured by sensitive detectors instead of the usual x-ray film. a computer receives the measurements and computes the relative attenuation of the x-ray beam at different points of a matrix representing the slice scanned. The computer processed information is then presented as images on a television screen by means of a cathode ray tube. The reconstruction diameter may be varied and the reconstructed image can be displayed over a full range of CT numbers or "window settings".

CT scans can be obtained at 5 mm or less contiguous sections in 2 planes; the high resolution axial plane and the coronal plane. Gantry tilt can be utilized so that the resultant images in the coronal plane become comparable with conventional tomography.

The transaxial view is particulary valuable in evaluation of posterior wall of the maxillary antrum and the pterygopalatine fossa. The coronal section gives better visualisation of the inferior orbital fissure with one limitation in patients with neck stiffness.

CT has the advantages of adding the horizontal plane of the area imaged, the amount of total x-ray received by the patient is less than that delivered in conventional radiography.

The most important feature of CT scan is its capability of imaging the soft tissue and extension of the disease from the paranasal sinuses into the orbit and cranial cavity, this is attributed to the sensitivity of CT scanners in discriminating small differences in tissue densities as low as 1% compared to 10% in conventional radiography. The excellent air-tissue contrast gives best visualization of the tumour mass and its outlines.Fig. 11 shows a coronal CT scan with invasion of the orbital structures.

CT provides better staging of sinus malignancy resulting in larger volumes of tumour being treated by radiography. If cranial invasion is suspected, intravenous contrast medium is given to view intracrnaial extensions. 70 ml of 420 conray are injected immediately prior to coronal CT sections. Fig. 12 shows a coronal CT scan with contrast enhancement which reveals intracranial extension.

Valerie et al (1983) studied 22 patients with paransal sinus and nasal cancer with orbital and intracranial extensions in whom craniofacial resection has been made.These patients had CT scanning before the operation.During the operation, assessment of the extension of the disease was carefully observed

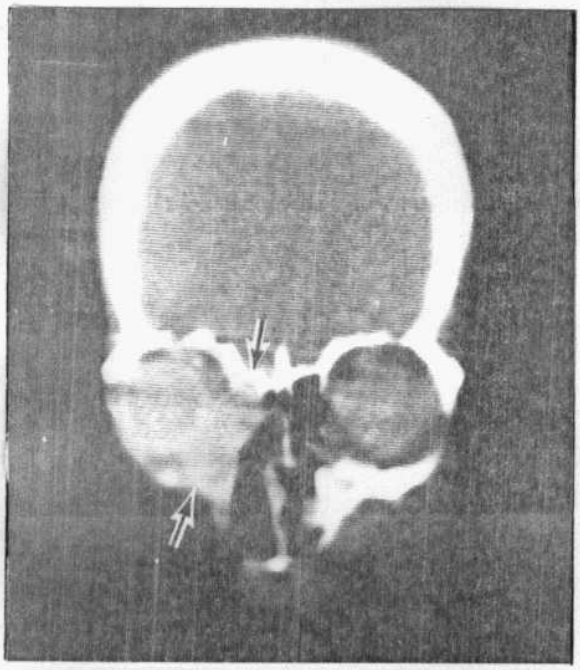

Fig.(11): Coronal CT scan reveals destructive lesion of right orbital floor, maxilla, and ethmoid. Mass has involved orbital stuctures, pushing globe laterally. Squamous cell carcinoma.

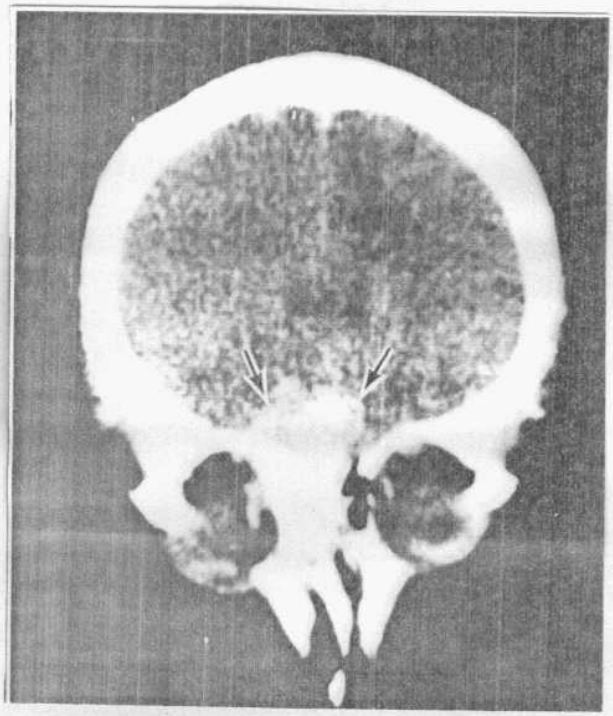

Fig.(12): Coronal CT scan with contrast enhancement reveals densely stained mass in right frontoethmoid area that extends intracranially. Meningioma extending into paranasal sinuses.

and specimens were referred for histological examinations. There was 78% complete correlation between the 3 methods of assessment.

CT is also, of great potential value in the planning of treatment of patients with malignant tumours of the nose and paranasal sinuses. It can give invaluable information as to the size and spread into the pterygopalatine fossa and/or the orbit, thus giving information about the operability of a lesion. Fig. 13 shows an axial CT scan of a juvenile angiofibroma of the nasopharynx with extension to the right antrum and pterygopalatine space.

When radiotherapy is appropriate, treatment can be planned more accurately and effectively. The advent of the treatment planner computer, which can superimpose planned radio-therapeutic isodose curves on the anatomical display of the tumour and its surroundings, is particularly important in this area of tightly packed anatomy of diverse histology.

Should radiotherapy be followed by surgery, the original extent of the lesions is accurately known to the surgeon and his "en bloc" excision can be planned accordingly.

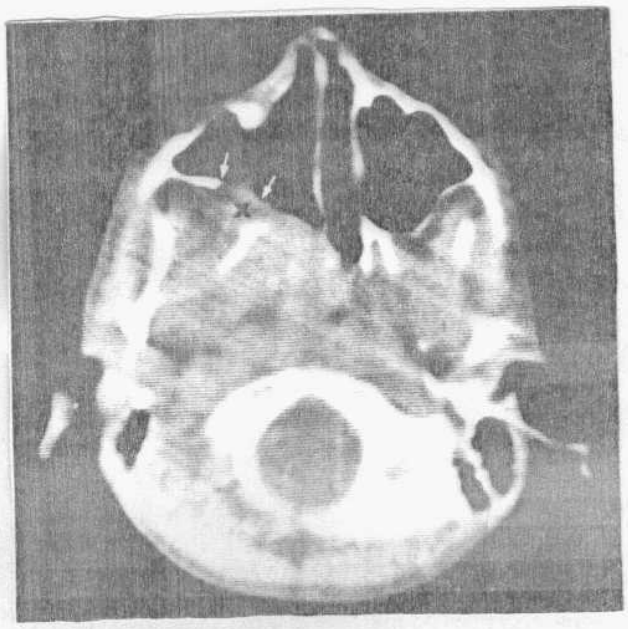

Fig. (13): Axial CT scan reveals anterior displacement and thinning of right posterior antral wall (arrow) with identing of pterygopalatine space (X) and destruction of medial pterygoid process. There is a large nasopharyngeal mass. Juvenile angiofibroma.

The use of the scanner in follow up is somewhat limited as surgery distorts anatomy but trismus resulting from invasion of the muscles of mastication can be seen in the scan due to the loss of contrast normally afforded by the abundant fat that lies between the muscles.

CT, thus, has several advantages over polytomography. Brian Eddleston and Richard Johnson (1983) made a study on 22 cases of paranasal sinuses and postnasal tumours to compare the results obtained by CT and polytomography and came to the following results:

In 21 out of 22 patients, the extent of the disease as assessed by the two methods was almost similar.

Certain features were better appreciated on CT. These were:

1- The presence and extent of associated bone destruction, while some authors (Thawley et al 1978) considered bone destruction to be equally demonstrated by the two methods.

2- The extent of orbital disease, this agrees with the findings of D.W Proops and P.D. Phelps (1982).

3- Extension of disease into the infratemporal fossa.

4- Intracranial extension of the disease, this also agrees with findings of D.W. Props and P.D. Phelps. Valerie et al (1983) think that the true extent of intracranial disease can only be assessed by CT.

Both methods are deficient in determining the nature of the lesion, so inflammation of soft tissue may resemble carcinoma while radiation necrosis resemble intracranial metastasis. Biopsy and subsequent histology are required for confirmation.

Also there are difficulties in the interpretation of bone changes on both methods especially if the bone is demineralized but in CT the use of contiguous thin sections (5 mm or less) can reduce this problem.

Although CT defines the extent of the disease more accurately, it does not alter the assessment of the planned radiotherapy according to polytomographic assessment. It makes difference when surgical treatment is to be undertaken.

CT, thus, has established itself as an extremely important component of any preoperative assessment of malignant disease.

Fig.(14) shows a coronal tomogram of a case of meningioma extending into paranasal sinuses to be compared with coronal CT of the same case shown in Fig. 12.

(4) **Intra-oral and occlusal films:**

The occlusal films demonstrate the antral floor and inferior portion of the nasal cavity. The mucosal changes are seen in intra-oral

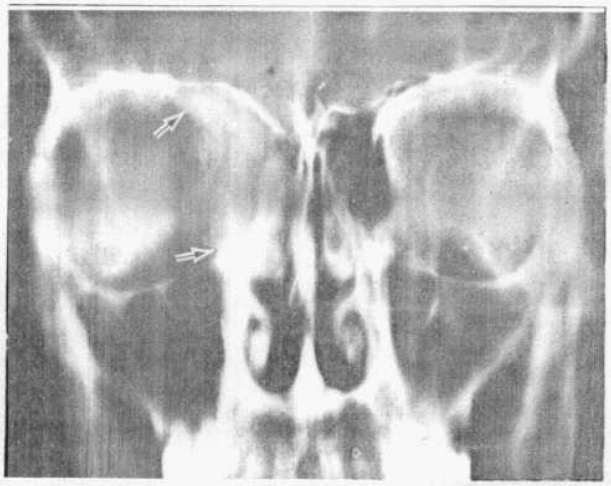

Fig. (14): Coronal tomogram reveals soft tissue mass in right frontal and ethmoid region that has broken through into orbit.

films but with limitations. In dentulous patients, periapical and bite wing x-ray films demonstrate the teeth and the surrounding alveolar process. Alveolar bone loss due to periodontal tumour invasion is quantitated. The antrum is often seen too.

(5) **Angiography:**

While most malignancies of the maxilla are avascular, preoperative angiography is used when the lesion is suspected to be vascular, is possibly invading or bound to a major vascular structure or is likely to be an angiofibroma of the nasopharynx extending into the nose or sinuses.

Fig. 15 shows an angiogram of a metastatic hypernephroma of the antrum.

(6) **Radio-isotopic scanning:**

This procedure is based on the principle that neoplastic cells may incorporate isotope-labelled compounds in different manner than do normal cells. it appears to have a limited value in the diagnosis and staging of head and neck cancer excluding thryoid.

King and Johnson (1981) concluded, on a base of their study of 198 cases of epidermoid carcinoma, that radio-isotope scans were of little value in evaluating preoperative metastatic disease.

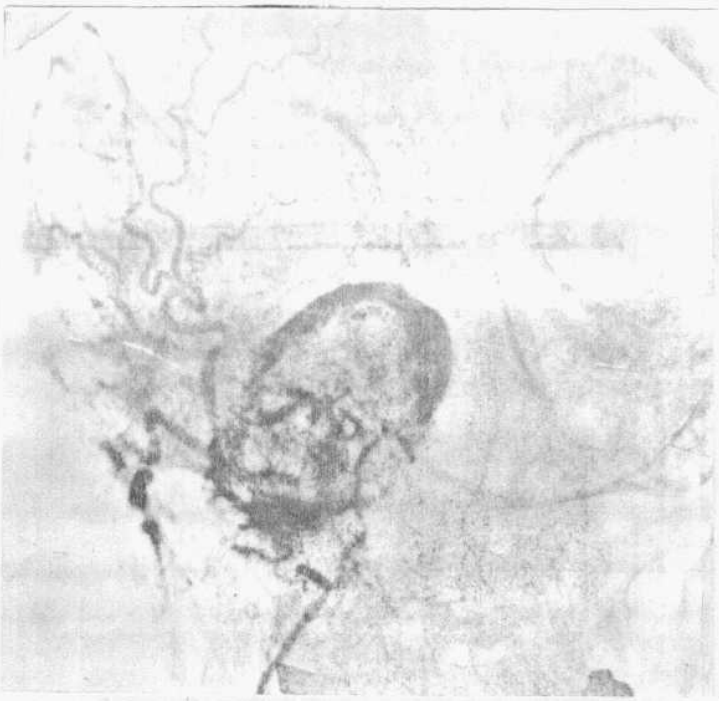

Fig.(15) : Frontal angiogram reveals vascular stain in right antrum. Metastatic hypernephroma.

On the other hand, Cummings et al (1981) studied 46 cases of head and neck cancer, including two with cancer antrum, one with cancer ethmoid, one with cancer hard palate, one with cancer alveolar ridge and one with cancer pterygomaxillary space, who were examined for the uptake of cobalt 57 tagged bleomycin by tumour cells and metastatic cervical lymph nodes. They concluded that this procedure increases the acumen in staging of head and neck cancer. Malignant tumours more than 2 cm in size appear to demonstrate active uptake of the imaging agent. Small tumour size and excess background radio-activity contributed to 17% false negatives while false positives were 10%.

II- **ANTRAL LAVAGE:**

This is an early investigation done when a malignant lesion is suspected to involve the maxillary sinus which looks opaque on plain x-ray films.

It may produce a blood stained fluid with fragments of the tumour. The obtained fluid is referred for cytological examination using Papanicolou stin.

It is of utmost importance to keep in mind that negative results obltained from a clear antral lavage are not indicative of absence of malignancy and the patient should be further investigated.

III- **EXPLORATORY (MINI) CALDWELL LUC OPERATION:**

This is done under local analesthesia when there is a suspicion of antral malignancy while radiographs are equivocal and no abnormal tissue is present in the nasal cavity. The incision is made in the inferior portion of the canine fossa to permit removal of the biopsy site at the time of definitive surgery. The anterior wall of the sinus is removed to visualize the interior and if a preoperative irrigation is to be considered, a nasoantral window assists in drainage of the necrotic antral contents.

Exploratory antrostomy is the only certain test to determine if and where the cancer is present in the sinus. It is the only sure way the extent may be staged.

IV- **ENDOSCOPY:**

Antroscopy is of great value in the study of cancer involving the maxillary sinus because it provides information that is unattainable in any other way, it gives proof of diagnosis and often assists in treatment. The regular use of antroscopy has proved that this technique is an ideal means for the early diagnosis of malignancy in the maxillary sinus.

This technique was first carried out by Hirschmann in 1901 when he introduced a cystoscope into the antrum. Since then, endoscopy

of the antrum has been brought to its present perfection by many investigators.

In fiberoptic endoscopy of the maxillary antrum, endoscopes with different viewing angles are introduced into the antrum through the lumen of a trocar by which the sinus is punctured via the inferior meatus or the canine fossa. Fine forceps attached to the optical instrument can be used for biopsies. The endoscopes can also be connected with a cold light fountain as well as with a camera and electronic flash for endophotogrpahy.

Papangelou (1983) described a new simple and inexpensive method for the endoscopy of the maxillary sinus. He used the common otoscope and a trocar specially designed to fit into the lumen of the otoscope. They are introduced into the maxillary sinus through the canine fossa. After the removal of the troca, the otoscope remains in the sinus and endoscopy of all the sinus can be performed very easily and quickly by mere moving of the axis of the otoscope. The anterior wall could be visualized by a small mirror of posterior rhinoscopy. This way needs no special experience needed for fiberoptic endoscopies plus it allows easy having of a biopsy.

V- **BIOPSY:**

Investigation of any patient with a neoplasm of the maxilla must include biopsy. If the tumour is accessible as when it presents in the nose, palate or under the cheek, satisfactory biopsy material can be taken under local anaesthesia with ease, otherwise exploratory Caldwell Luc operation is performed. In many instances, biopsy needs be taken under general anaesthesia.

Generous amounts of tissue should be taken, especially from the interior of the mass because many of these tumours are inflamed and necrotic on their periphery. Care should be taken not to crush the tissue taken as this may create artifacts. In taking a biopsy from a lymphoma, the use of puch biopsy forceps should be avoided as lymphoma cells are extremely susceptible to maceration. The specimen biopsy itself is taken with sharp dissector or a snare. The specimen should be prepared promptly and placed in preservative then immediately delivered to the laboratory.

Other investigations may be needed. Chest radiographs are recommended as a routine for detection of hidden primaries and metastasis. Scans of liver and spleen, bone scans and abdominal CT scans are needed for suspicious metastasis. Cerebrospinal fluid cytology should be done if there is neurologic symptomatology or radiographic evidence of intracranial involvement. Bone marrow aspirations, trephine biopsies and

also lymphangiograms in suspicious cases of malignant lymphomas are sometimes needed.

It should be stressed that even after all these investigattions, diagnostic errors are as much as almost one third of cases.

Robin and Powell (1981) studied 282 cases of carcinoma of the nasal cavity and paranasal sinuses and found the diagnostic errors to be up to 34.8% (3.2% overestimations and 31.6% underestimations). The most frequent errors were found in the ethmoids, pterygoplatine and infratemporal fossae, the anterior wall of the antrum and face, the orbit, the lower nasal cavity and other sites in that order of frequency. The only sure diagnosis is achieved by surgery.

Radiographs of maxillary sinus squamous cell carcinoma are not pathognomonic and always show soft tissue opacity of the sinus and nasal cavity with possible extensions to the postnasal space. Erosion of antral walls is frequently seen.

Radiographs of pleomorphic adenomas of the hard palate can show formation of a recess in the alveolar bone due to pressure resorption. The interface between the normal hard palate and the region of osseous involvement is sharply defined by cortical bone and the

edges of the bony defect may be bevelled upward. It may extend into the maxillary antrum.

There is nothing characteristic about the x-ray appearance of malignant lymphomas, with usual picture of osteolytic destruction. While in Burkitt's lymphoma, one of the earliest radiographic signs is a loss of break in the lamina dura around erupted or developing teeth. Small discrete radiolucencies may also be seen at this stage. These later Coallesce to produce larger defects in the jaw. The earliest sign may be blurring of the shaddow of the antrum. Sun-ray appearance is not uncommon.

In malignant plasma cell tumours, two different x-ray characteristics have been described:

1- A sharply demarcated, osteolytic, multicystic lesion located int he medulla.

2- A destructive, trabeculated lesion with occasional cortical expansion.

In myxoma of the maxilla, the x-ray appearance is that of a multilocular cyst with a dense radiographic margin. Initially, the cortex is intact but thinned and expanded. Progressive growth of the tumour produces perforation and destruction of the cortex with invasion of the soft tissue. The antrum as well as the alveolar bone are often invaded

giving a dense cloudy appearance to the sinus on a water's view.

The radiographic appearance of fibrous dysplasia of the maxilla is variable and characteristic. There may be diffuse uniform sclerosis with enlargement of the jaw. Another picture is expansion of the cortex with multiloculations and thinning. A third picture is a unilocular form. Large lesions usually obliterate the maxillary sinus with involvement of the infraorbital margin and molar bones.

In osteosarcomas, the radiographic appearance is variable depending upon the extent of bone destruction. If the neoplasm comprises the cortex, the sun-ray appearance is present. This occurs in about 25% of cases (Vener et al 1984). If the lesion is primarily lytic, no specific radiographic appearance exists. A symmetrically widened periodontal membrane space about one or more teeth is a peculiar finding on a periapical dental view. Low aggressiveness is suggested by localized destruction, well defined margins, solid periosteal response and cortical expansion, while high aggressiveness is suggested by moth-eaten destruction, ill-defined margins, cortical expansion, soft tissue mass and interrupted periosteal response.

Differential Diagnosis:

A variety of conditions have to be considered in the differential diagnosis of tumours of the maxilla.

Maxillary sinusitis seldom produces a tumefaction, and its clinical evolution and marked inflammatory elements often facilitates its diagnosis. Inverted papillomas may occur in the maxillary sinus, primarily or secondary to nasal cavity manifestations. On biopsy, an unexperienced pathologist may make a diagnosis of papillary carcinoma (Harlan J. Spjut 1977).

Osteoma of the maxilla is a relatively common benign tumour of slow growth. It may be either a peripheral or a central (intraosseous) lesion. It presents as a painless mass between the ages of 15-40 years. Grossly, the tumour is attached to the surface by a pedicle or a sessile stalk. Three microscopic variants occur; osteoma eburneum, osteoma spongiosum and osteoma durum having different ratios of bone to stroma. Chondroma of the maxilla should be diagnosed with reservation as the zone between benign tumours overt chondrosarcomas may still be incompltely defined (Batsakis 1981).

Chondromas of the maxilla are slowly growing and locally invasive lesions which are difficult to remove with tendency for recurrence. Microscopically, most of the cells possess a single nucleus which tends to be small. There is well differentiated hyaline cartilage. Myxochondromatous, fibrochondromatous and osteoid or even bony areas exist. There appears to be equal sex affection of the tumour with a peak age incidence in the 3rd decade.

Radiographically, they are transparent and may present spotted calcifications.

Odontogenic tumours affecting the maxilla include the following types:

a- Adenomeloblastoma: This is a hamartoma, benign, slow growing lesion with peak incidence in the second decade of life and modest female predominance. it is a localized painless swelling exclusively in the anterior tooth-bearing area. Grossly, encapsulation, calcification and cystic alteration are seen. Microscopically, central epithelial proliferation and rosettes are seen.

Radiographs show well demarcated unilocular radiolucency with possible peripheral condensing osteitis and small opacities in the central radiolucencies.

b- Ameloblastic fibroma: This is a completely benign hamartoma. It presents in the maxilla as a painless asymptomatic swelling of the premolar-molar area (Batsakis 1981). It is encapsulated having islands, cords and buds of epithelial tissue in mesenchymal connective tissue stroma. Radiographically, it resembles ameloblastoma and differentiated by its stroma which strongly resembles embryonal connective tissue of the primitive dental pulp.

c- Odontomas: The majority of these tumours are hamartomas were induction has resulted in the development of enamel and dentin. Odontomas in order of frequency are: 1) Compound and complex odontomas. 2) Ameloblastic odontomas and 3) Ameloblastic fibro-odontoma (batsakis 1981).

Microscopically, they contain tissue elements seen in adult tooth structures. In complex odontomas, these elements are randomly distributed whereas in compound odontomas, the relative positions of the dental tissues are normal but the resulting teeth are dwarfed and multiple.

Odontomas generally occur between the first and third decades of life with slight female preponderance.

Radiographs reveal mass of tooth-like structures surrounded by narrow radiolucent bands. Odontomas, but compound type, show cyst-like destruction.

d) Cementomas: These are a group of lesions some of which are benign, viz benign (true) cementoma and cementifying fibroma, and others are dysplasias, viz periapical cemental dysplasia and familial gigantiform cementoma (Batsakis 1981). They are derived from the periodontal membrane and are very rare occurrence in the maxilla.

the presence of cementum is a common feature. Amounts of fibrous connective tissue vary in different types.

Radiographic features are of a radiolucentto mottled mass at the apex of tooth roots in the periapical type, radiolucent defect with central mottled radio-opaque mass at molar site in the cementifying fibroma, radio-opaque mass attached to tooth root in cementoblastoma and ill-defined radiolucent to radio-opaque multiple lesions in gigantiform type.

e- Squamous odontogenic tumours: It is a very rare benign lesion which presents as a painless mass, loose teeth or asymptomatic radio-lucency of the alveolar bone. Microscopically, the tumour appears as benign squamous islands in a dense collagen connective tissue stroma.

Cysts of the upper jaw are either odontogenic or fissural cysts and are a very rare occurrence in the maxilla. Odontogenic cysts comprise the periodontal, dentigerous, gingival, odontogenic keratocyts, keratinizing and calcifying cysts.

Fissural cysts comprise lateral, medial and median cysts. These arise from odontogenic origin or from developmental faults of the region.

Accurate history, precise localization and high quality radiographs are necessary for diagnosis since epithelial lining of various cysts are closely similar.

Non-secreting cysts and retension cysts arising from the mucosa of the maxillary sinus are usually the result of chronic inflammation.

Mucoceles may cause facial deformity and bone erosion. They commonly arise from frontal and ethmoid sinuses.

Benign myxomas of the maxilla are rare mesenchymal tumours. Odontogenic myxomas originate from either dental tissue, nondental mesenchyme or represent dysplastic bone lesions. Microscopically, myxoma shows stellate cell within an intercellular substance. Fragments of calcification occur. Radiographs are non-specific and consist of multi-loculated radiolucency that may perforate the cortex. Myxoma is usually asymptomatic until it compresses adjacent structure (Batsakis 1981).

Mucous and salivary gland benign tumours arising from the mucosa of the maxillary sinus are very slowly growing. They are well circumscribed, well-encapsulated and in their expansion destroy, but seldom infiltrate, the surrounding tissues. They have varied histologic appearances. (Regato and Spjut 1977).

Ectopic meningioma of the maxillary sinus is extremely rare occurrence. From 1931 through 1983, only 6 cases have been reported (J.J. Manni 1983). Their origin is still uncertain. They have no characteristic symptomatology consisting of nasal swelling and obstruction and are readily diagnosed by histological examination.

Neurilemmoma of the maxillary sinus is a rare benign tumour which arises from the neuroectodermal cells of the Schwann sheath. Histolgoically, it consists of elongated cells with palisading nuclei. Areas of myxomatous degeneration, hyalinization and foci of recent and old haemorrhages may occur. Grossly, the tumour is well encapsulated with nodular surface. The tumour is slowly growing and may remain silent until large size where pressure effect gives clinical symptoms (Puterman et al, 1981).

Paget's disease is an idiopathic inflammatory disease of bone infrequently affecting the jaws more frequently the maxilla (Batsakis 1981). dull pain, growth deformity and gradual enlarlgement of the jaws with spreading of the teeth are the commonest complaints. Retroclination of the incisor teeth and palatoversion of the posterior teeth are constant and most striking features. The affected area is characteristically warmer than the unaffected areas as a result of extensive arteriovenous communications (Batsakis 1981).

The initial radiographic finding is osteoporosis, later "cotton wool" appearance is seen. In the fully developed stage of Paget's disease, there is coexistance of osteoblastic and osteoclastic activity.

Giant cell granuloma of the maxilla is a rarity and has to be differentiated from true giant cell tumours. It occurs in two forms: (1) peripheral; involving the gingiva or alveolar mucosa. It rarely exceeds 2 cm in size and may be sessile or pedunculated showing no radiographic abnormality and (2) central; occurring as an endosteal lesion within the jaw bones. It may be asymptomatic or produce local deformity of the jaw. Radiographs show radiolucent areas of varying sizes and occasionally "soap bubble" appearance is present.

Microscopically, this lesion shows proliferation of fibroblastic or mesenchymal connective tissue. It is richy vascular with variable number of multinucleated giant cells (Batsakis 1981).

Plasma cell granuloma of the maxilla represents unusual abundance of plasma cells normally present in the upper aerodigestive tract. it should be differentiated from extramedullary plasmacytoma. Microscopically in the granuloma, plasma cells show no abnormality and inflammatory cells and Russel bodies are present (Batsakis 1983).

Wegener's granulomatosis and midline (non healing) granuloma must be differentiated from lymphomas, histiocytosis, lympho-epithelioma and renal clear cell metastatic carcinoma of the maxillary antrum. Wegener's granulomatosis is characterized by three criteria (1) necrotizing granulomas with vasculitis (2) systemic vasculitis and (3) focal necrotizing glomerulitis. There is always some form of respiratory tract involvement. The nasal and paranasal findings are characteristic. A history of chronic sinusitis with purulent rhinorrhoea particularly involving the maxillary sinus (Batsakis 1981). The nasal and paranasal midline granuloma can be locally ulcerative, destructive and diffuse.

Hemangiomas of the jaws are considerably rare. No more than 60 cases have been reported (Batsakis 1981). They mimic very much other tumours of the jaws. Symptomatology consists of teeth spacing, disturbed occlusion, hemorrhage around the teeth necks and severe bleeding after extraction. Radiographs are diagnostic. The principal change is an area of rarefaction. "Honey comb" or "soap bubble" is the most common feature.

Cherubism is an autosomal dominant disorder affecting the jaws espeically the maxilla. Marked involvement results in a narrow V-shaped palate vault. Premature loss of deciduous teeth and a failure of eruption of the permanent teeth are common findings. A "raised-to-heaven"

look is imparted to the patient by the diffuse enlargement of the lower half of the face and retraction of the lower eyelids. The lesion manifests during the 2nd to 3rd year of life and there is gradual improvement after the age of 10 years. There is proliferation of fibrous tissue and variable amounts of giant cells within the jaws as constant fidings (Batsakis 1981).

Torus, exostosis and enostosis are benign bony growths projecting outwards from the surface of the jaw (as for torus and exostosis) or inwards (as for enostosis). The lesion is formed of a bony nodule of variable sizes but exostosis may be pedunculated or flat. The lesion may be lobulated and the overlying mucosa may be ulcerated in torus. Exostosis appears radiographically as a circumscribed radio-opaque mass.

MANAGEMENT

Lizare, in 1826, was the first to report the successful resection of the upper jaw and sinus. In 1888, Parsons reported the destruction of a tumor in the sinuses by the use of cautery. With the introduction of radiotherapy, surgeons from 1910 until about 1925 began to combine irradiation with electrosurgery. From 1925 to 1940, there were a number of reports on the use of electrodessication and radium implantation combined with surgery. From 1945 until 1955, there was a reawakening of interest in radical surgery, which usually included a total maxillectomy and extenteration of the orbit. Orthovoltage irradiation was given in cases where disease was left behind (Sisson and Becker 1981).

At the present time there is no standard treatment endorsed by all surgeons. More recent reports stress the importance of individualizing each case and of developing a treatment after consideration of the histology, location, extent (Stage) of the tumour, and various patient factors.

A variety of therapeutic measures are available for the management of malignancies in the maxilla. These include surgical excision, radiation therapy, cryotherapy, laser excision, chemotherapy, immunotherapy and others. The choice of treatment modality depends upon

many factors such as (1) the site and extent of the primary lesion; (2) the likelihood of complete surgical excision; (3) the possibility of preservation of function and cosmosis; (4) the presence of bone and muscle involvement; (5) the presence of nodal and metastatic lesions; (6) the gross characteristics of the tumour; i.e., exophytic-superficial vs endophytic-invasive; (7) the physical status; (8) the social status and occupation of the patient and (9) the experience and skill of the surgeon and the radiation therapist (C.C Wang 1981).

At the present time, cryotherapy and laser excision are used in experienced hands primarily for accessible tumour with limited treatment success. Both chemotherapy and immunotherapy are used primarily for palliation or as adjuvant therapy (C.C. Wang 1981).

Surgery and radiotherapy are more frequently used in the treatment of maxillary cancer and each has its own merits, indications and limitations. Radiation therapy has the advantage of being able to control the disease in situ, thus avoiding sacrifice of a useful and necessary anatomic part as well as preserving function. On the other hand, surgery is preferred and can be carried out expediently and effectively without functional and cosmetic mutilation when lesion is localized and occupying less strategic locations.

Failure of surgical management of paranasal sinus cancer is often due to inability to excise microscopic tumour extensions outside the sinus bony walls. Regional lymph node and distant metastases are uncommon, the latter being 3.5% or less (D.F.N Harrison 1979). It is extension of tumour to orbital apex, posterior ethmoidal cells, pterygoid region and nasopharynx which results in disease left in situ. Consequently, it is such extensions which must be determined initially and classified accordingly.

The mechanisms of radiotherapeutic failures are different. The tumour core greater than 150 to 180 μm often contains hypoxic cells that are insensitive to radiation therapy in contrast to the better oxygenated, well nourished marginal tumour cells. Local failure from radiation therapy, therefore, is central rather than marginal in nature.

Hereby, I will try to give a detailed account of the different surgical and radiotherapeutic procedures as well as the chemotherapeutics in common use for maxillary cancer. Later on I will give an account of different treatment disciplines and combinations.

SURGICAL PROCEDURES

Paleopathological evidence suggests that neoplastic disease existed in the upper jaw in earliest times, being found in pre-Christian Nubian skulls.

Probably the first successful total maxillectomy - for osteo-sarcoma - was carried out by Joseph Gonsol of Lyons in 1827. Moure (1902) of Bordeaux described his lateral rhinotomy in 1902 and these two operations have provided the mainstay of sinus cancer surgery to date (D.F.N Harrison 1979).

Lateral Rhinotomy - Initial Exposure:

A lateral rhinotomy is a procedure used to provide the necessary exposure so that the boundaries of the tumour can be defined (Figs. 16 A, B, C). There has been a failure to appreciate the value of this approach to the nasal passage despite the excellent exposure, of both ethmoid and maxillary sinuses, that can be obtained with little or no postoperative deformity (D.F.N Harrison 1978).

This procedure is the initial step prior to most subtotal and radical maxillectomies with or without orbital exenteration. The oribital contents and globe are a part of the surgical field and the cornea must be protected by suturing the eye lids together.

This approach when combined with a maxillectomy is begun with an orbital incision running along the lower lid about 3 mm from the edge of the tarsal plate, otherwise it starts at a point halfway between the medial canthus and the dorsum of the nose. For cosmetic purposes, this incision should run inferiorly along the nasofacial groove, follows

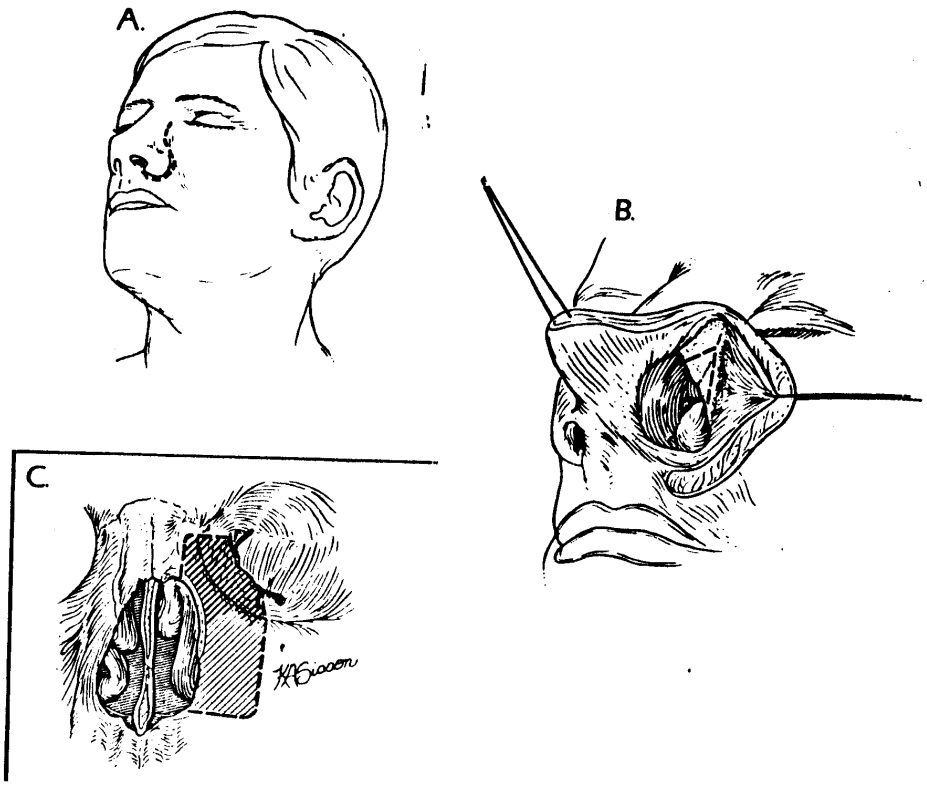

Fig. (16): (A) Lateral rhinotomy incision.
(B) Nasal bone removal.
(C) Antral bone removal.

the ala to the columella, and is continued into the philtrum without splitting the upper lip, unless there is no other to obtain adequate exposure (Sisson and Becker 1981). All layers are divided, retraction of the alar region away from the incision then allowing a clear view of the vestibule and front of the nasal cavity.

A flap is developed by periosteal elevation exposing the anterior surface of the maxilla, the inferior aspect of the orbit, the frontal process of the maxilla, and a portion of the nasal bone. The lateral rhinotomy is completed by lateral osteotomy with, in some cases, an anterior-superior division of the nasal septum. The nose is retracted towards the side opposite the lesion. It is often necessory to remove the cartilage of the nasal septum to facilitate evaluation of the tumour boundaries. Bone is removed from the lateral wall of the pyriform aperture and a section of the nasal bone is resected for better exposure of the nasal cavity.

The periosteum is elevated from the medial and inferior walls of the orbit. If one is able to free the periosteum easily without encountering tumour, orbital exenteration may not be necessary. Frozen section control is important. If there is periosteal involvement without gross invasion of the orbital contents, the eye can still be saved.

Bone removal can be extended to include both lacrimal bone and the lamina papyracea of the ethmoid. By this means the ethmoidal

labyrinth can be completely removed together with all the turbinals. (D.F.N. Harrison 1978). If the lesion is primarily located in the ethmoid and the extent of the disease in the antrum has not been surveyed, then an antrostomy is performed through the canine fossa to explore the maxillary sinus and to decide which portions of the maxilla are involved by the cancer.

Bleeding is minimal unless the nasal passages contain an unusually vascular or malignant tumour. The excellent visualization obtained by this approach enables the surgeon to control even the most troublesome haemorrhage. Skin grafting of bare septal cartilage or bony nasal floor is unnecessary since re-epithelialization occurs rapidly.

Radical Maxillectomy:

For total maxillectomy to be an effective oncological operation, the disease must be confined within the bony walls of the maxillary sinus or, in the case of primary bony tumours to the maxilla itself. Unfortunately, this is rarely the case and most operations for carcinoma of the maxillary sinus will require not only a total maxillectomy, but also orbital clearance together with exenteration of the ethmoidal labyrinth. So, radical maxillectomy alone is suitable only for most T_1 and T_2 antral lesions. Hypotensive anaesthesia is a common practice for this operation to minimize bleeding which allows better visualization of the anatomical limits of the excission (D.F.N. Harrison 1978).

The Weber-Fergusson incision is commonly used to expose the facial surface of the maxilla. By following the techniques of lateral rhinotomy, exposure is achieved and the limits of the disease defined. Transverse extension of the incision to be 2 mm below and parallel to the lash margin of the lower lid is preferrable to extending the incision up to the eye brows as it minimizes post-operative oedema of the lower lids (D.F.N Harrison 1979). If the orbital contents are to be preserved, then the eye lids are sewn together, while if the contents are to be removed then a circumferential incision is made through the conjunctiva, thus preserving both eye lids which facilitate the fitting of an ocular prosthesis. Fig. 17.A.

The remainder of the incision is intra-oral and follows the alveolar buccal sulcus, around the maxillary tuberosity and across the palate at the junction of the soft palate with the posterior end of the hard palate.

The final incision is slightly lateral to the midline to join the original incision in the region of the upper first incisor tooth which will have to be removed if present. Diathermy may be used for the intra-oral incision to minimize bleeding (Harrison 1978), Fig. 17.B.

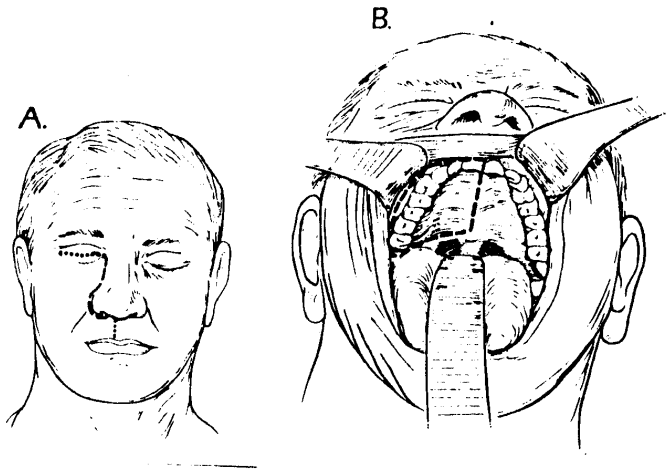

Fig. (17): (A) Extension of lateral rhinotomy incision for radical maxillectomy.
(B) Incision used to spare soft palate.

After the incisions, the flap of the facial skin and periosteum is raised further laterally, exposing the entire anterior surface of the maxilla. Exposure of the anterolateral portion of the zygomatic process of the frontal bone, the frontal process of the maxilla, and the medial and inferior walls of the orbit is required. Involvement of the periosteum, muscle or skin overlying these areas should be carefully checked. If they are involved, they are removed en-bloc with the bone.

If orbital exenteration is indicated, the incision in the lower lid is extended around the medial and lateral canthi and then just above the tarsal plate in the upper lid. The palpebral skin is elevated superiorly exposing the orbicularis oculi muscle. The periosteum along the superior orbital rim is incised and the oribital periosteum reflected off the roof and the medial and lateral aspects of the orbit. The orbital contents are then retracted inferiorly to permit ligation and transection of the ethmoid and ophthalmic vessels. The orbital contents are removed in-continuity with the roof of the maxillary sinus. If the eye is not to be included in the resection, the periosteum is elevated from the orbital floor and lamina papyracea and the ethmoidal arteries are ligated and transected as they penetrate their corresponding foramina (Sisson and Becker 1981). Elevation of orbital contents allows identification of the inferior orbital fissure and a curved forceps may be introduced under the malar bone and passed into the orbit to allow division of the bone by a Gigli Saw. Fig. 18 and 19.

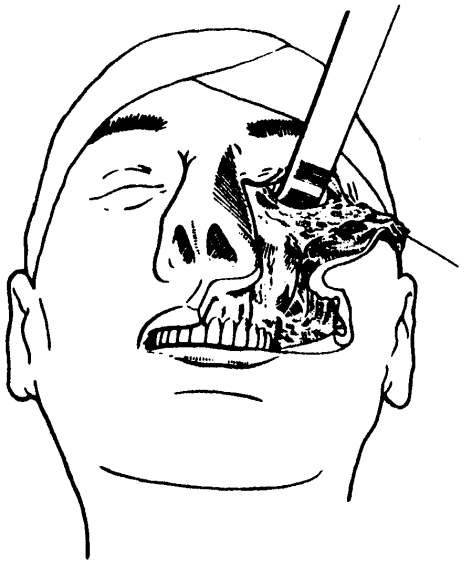

Fig.(18): Diagram shows elevation of the facial skin, buccinator and skin of the lower eyelid leaving the orbicularis oculi muscle in situ.

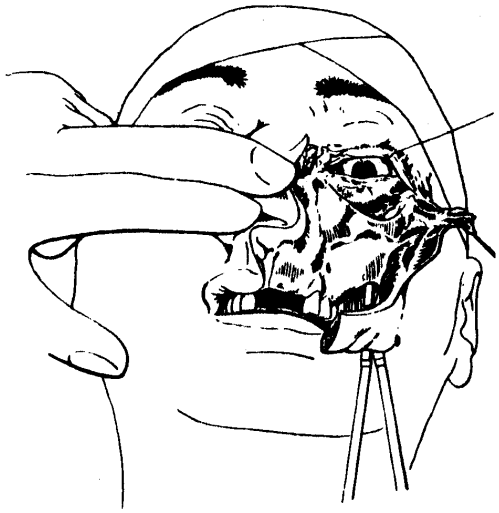

Fig.(19): Diagram shows a curved forceps introduced under the malar bone and passed into the orbit to allow division of the bone.

After the incision in the palate is made, the mucoperiosteum is elevated. if the floor of the maxilla is to be removed, the musculature of the soft palate is then separated from the hard palate at their junction.

Follwoing the elevation of the tissue from the underlying bony framework, bone cuts are performed without cutting through tumour tissue. The extent of the bone resection is modified individually according to the extent of the tumour. The first bone cut is an osteotomy just below the frontoethmoid suture or on a level passing just inferior to the anterior and posterior ethmoid foramina (Fig. 20.A). anteriorly, this cut passes through the frontal process of the maxilla.

When the orbital contents are to be saved, care must be taken to avoid damaging the optic nerve. Therefore, the posterior extent of the horizontal ethmoid osteotomy stops at the level of the posterior ethmoidal artery and a vertical cut is made carefully to provide a fracture line at the posterior aspect of the labyrinth (Sisson and Becker 1981).

Laterally, a Stryker or a Gigli saw is used to transect the frontal and temporal (zygomatic arch) projections of the zygoma after division of the masseter at its attachment.

The ethmoidal labyrinth is removed, wether involved in the disease or not, prior to separating the nasal bone from the frontal process

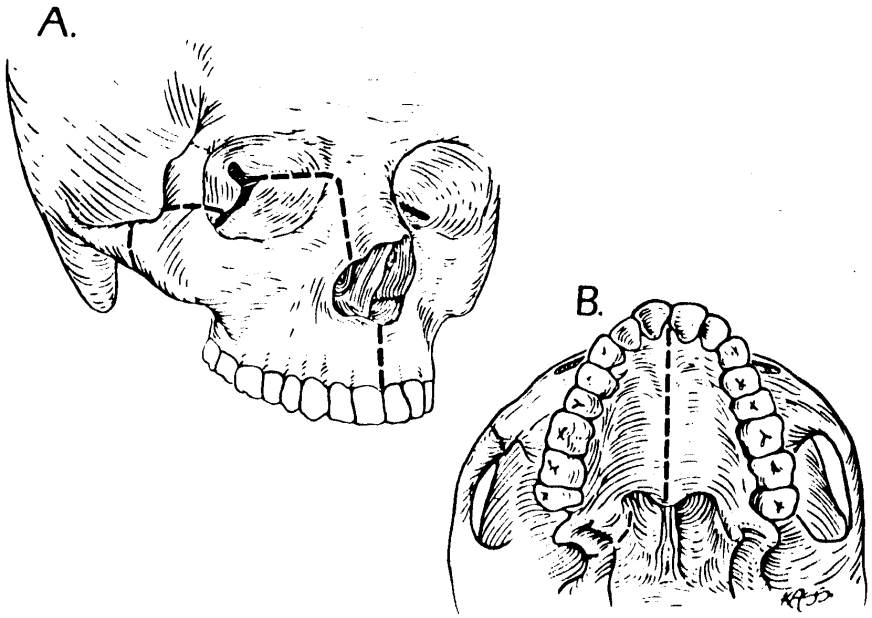

Fig.(20): (A and B) Bone cuts for radical maxillectomy and orbital removal.

of the maxilla, but care should be taken not to damage the cribriform plate (D.F.N Harrison 1978). The bony orbital floor lying between the inferior orbital fissure and the defect now left in the medial orbital wall is divided with an osteotome. The upper part of the maxilla is now free.

The hard palate may be transected with a Gigli saw but better with a wide osteotome for the alveolar margin, placed at the site of the upper central incisor, and a chiesel for the hard palate. The straight edge of the latter must be sited just lateral to the midline to avoid splitting the nasal septum superiorly (D.F.N. Harrison 1978) (Fig. 21).

The last bone cut is at the base of the pterygoid process, this is carried out after examining all other areas of transected bone with an osteotome to be sure of proper detatchment (Fig. 20.B). Removal of the medial and lateral pterygoid plates facilitates the fitting of a comfortable prosthesis and minimize postoperative trismus. Bleeding from the maxillary artery is rarely a problem but it should be secured and ligated. Any remaining soft-tissue attachments are cut and the specimen removed fromthe field. The remaining bony walls of the ethmoid, anterior wall of sphenoidal sinus and, where the eye has been removed, orbital floor are removed together with the pterygoid plates (Fig. 22 and 23).

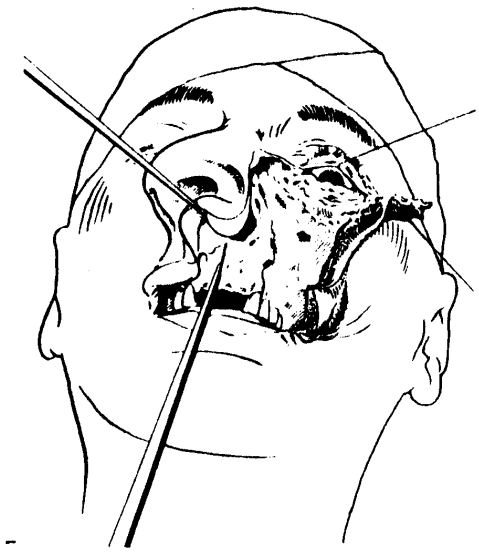

Fig. (21): Diagram shows division of the alveolar margin by a wide osteotome at the site of the upper central incisor.

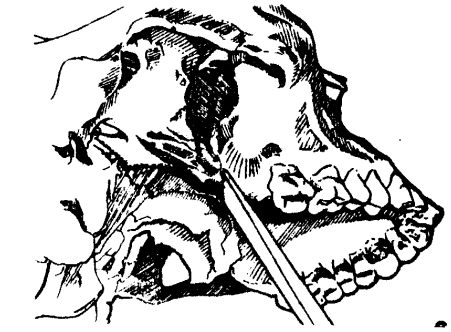

Fig. (22): Diagram shows separation of the pterygoid plate with an osteotome.

Fig. (23): Diagram shows removal of the maxilla.

Total maxillectomy is inevitably followed by sinking of the eyes (D.F.N Harrison 1979). The lower part of the fascia bulbi (Tenon's capsule) is thickened and connected to both medial and lateral cheek ligaments to form sling upon which the eyeball rests. The suspensory ligament (of Lockwood) will prevent dropping of the eye following removal of the orbital floor providing the periosteum and its medial and lateral attachments are undamaged. However, re-inforcement can be provided by a sling from the temporalis muscle fixed medially to the exposed orbicularis oculi muscle.

Prior to skin closure the inner raw surface of facial skin may be grafted with split-skin although this is not always necessary. when the eye has been removed, the lash margins and tarsal plates are cut off from the eye lids which are then approximated by sutures.

It is always desirable to introduce a prosthesis into the palatal defect at the end of the operation since this prevents contracture, maintains facial contour and enables the patient to eat a soft diet immediately (D.F.N Harrison 1978).

Larson et al (1982) studied the question of preservation of the orbital contents which are removed as an absolute indication when there is tumour invasion of the periorbita, posterior ethmoid cells or orbital apex. They had a retrospective study on 43 patients who had

primary cancer of the maxillary sinus observed between January 1973 and December 1978 in the M.D. Anderson Hospital and Tumour Institute, 32 of them had suamous cell carcinoma. The orbital contents were preserved in 27 patients but the orbitl floor was removed and the bed skin-grafted in 14 patients.

The criteria for preservation of the orbital contents were applied in general by the operating surgeon. These included attention to the condition of the periorbita, notice of invasion of the infra-orbital nerve, orbital apex or both, posterior ethmoid involvement and the morphologic condition and aggressiveness of the tumour.

Results in the 27 patients, whose orbital contents were preserved, are shown in table 12:

Table 12 : Results of Eye Function in Preserved Eye

Eye Function	No. of Patients
No problems	12
Minor problems	14
Removal secondary to irradiation	1
Total	27

Minor problems were diplopia, exposure keratitis, hypotropia and ectropion, most of which responded to local conservative management.

These results confirm that there are oncologically sound indication for preservation of the orbital contents in the treatment of primary cancer of the paranasal sinuses in selected cases. Reconstruction of the orbital floor with a skin graft even when combined with radiation therapy gave a functional eye in the majority of cases. The occurrence of local, regional and distant metastases seemed to be affected more by aggressiveness of the tumour rather than the preservation of the orbital contents.

Partial maxillectomy:

Anything less than complete removal of the maxilla is a partial maxillectomy. Consequently, this may cover procedures varying from simple alveolectomy to removal of palate, alveolus and anterior wall of maxilla. Since these operations are performed perorally there is no external deformity and the immediate fitting of a prosthesis minimizes post-operative disability (D.F.N. Harrison, 1979).

This operation is possible when diseas is located in the inferior portion of the maxilla and involves only the hard palate or alveolar process. The resection can be further modified to accomodate lesions

of the nose invading the ethmoid complex and the common wall between the nasal cavity and the maxilla. The alveolar margin is also included in the bony removal in order to produce an easily visible cavity.

Through a lateral rhinotomy incision, the ethmoid complex and medial aspect of the maxilla including the lateral nasal wall are removed as a block. This may be combined with a craniofacial resection to resect the cribriform plate and ethmoid roof. (sisson and Becker 1981).

The extensive cavity left after radical maxillectomy should be easily available for postoperative inspection. This is achieved by removal of bone and soft tissue surrounding the opening into the cavity, and by wide opening of the sinuses surrounding the cavity.

Medical Maxillectomy:

Sessions and Humphreys (1977) described a surgical procedure that they termed medial maxillectomy and in 1983 they added some technical modifications. This procedure gains access to relatively inaccessible areas of the middle part of the skull and floor of the frontal fossa involved in various tumours. It aims at visual exposure of the posterior aspect of the specimen involved for resection. Fig. 24 shows diagramatic scheme of bone removal in the medial maxillectomy. This procedure could be combined with frontal cranial resection utilizing a cranial-facial approach to en bloc ethmoidectomy.

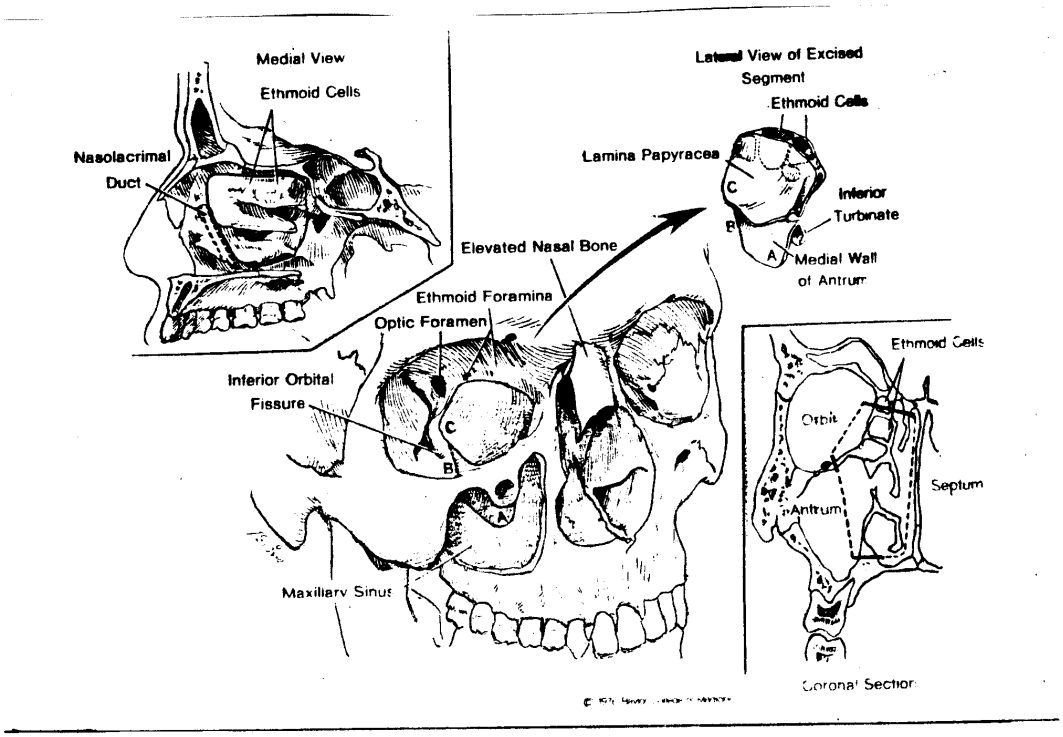

Fig. (24): Center diagram shows intended specimen for removal left inset. Sagittal view of skull shows lines of resection Frontal view shown lines of resection.

Specific modifications:

1- In certain circumstances, specifically anteriorly located lesions, it is unnecessary to extend the rhinotomy incision through the upper lip originally performed in the original techniue. Adequate visualization can be attained by the canine fossa incision plus single retraction of the facial tissue after rhinotomy.

2- In the original technique, orbital content retraction is performed with dissection of the lacrimal sac from the sheaths of the periorbitum, then the orbital contents are retracted and the lacrimal duct and sac are stretched. This sometimes resulted in injury to that structure. This is modified to complete transection of the lacrimal sac early in the operation prior to undertaking any of the bone cuts. This facilitates lateral retraction of the orbital contents. Fig. 25.

3- On completion of the medial maxillectomy, restoration of the integirty of the lacrimal draimage system is performed by placing the tip of 14-gauge polyethylene tube in the sac. A purse string of absorbable suture material is then placed in the sac tissue and tightened securely about the tube as in appendicectomy. The distal end of the tube is passed through the previous site of the lacrimal fossa into the nasal cavity.

In order to further stabilize this system, a small (4-0 or 5-0) monofilament suture is placed from the contralateral nasal cavity through

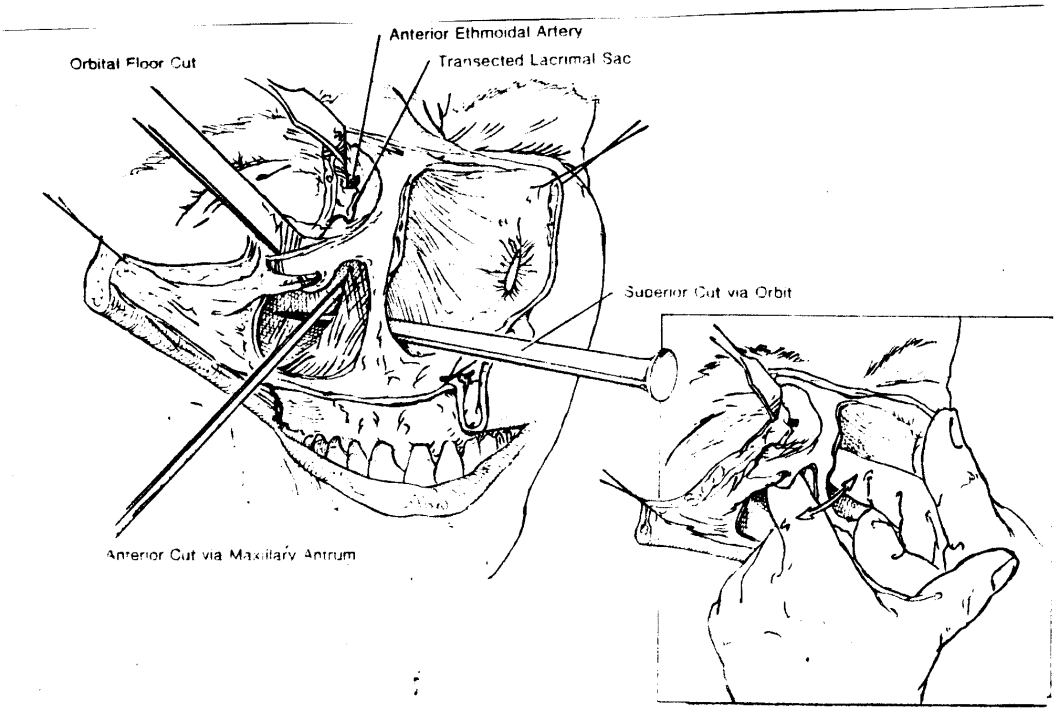

Fig. (25): Diagram shows transected lacrimal sac with lateral retraction of orbital contents. This is done prior to performing bone cuts, indicated by osteotomes.

the cartilagenous septum, looped around the tube and passed back through the cartilagenous septum and tied securely against the septum. Tube is left in place 3-4 weeks to allow mucosal lining of the newly formed tract.

4- Extensive removal of the lamina papyracea which is a part of medial maxillectomy can lead to misplacement of medial palpebral ligament producing an unsightly difference in the vertical position of the ocular commisure. A modification has been made where the medial orbital tissues are fixed to the correct spot. This fixation is accomplished after completion of the medial maxillectomy at which time a small hole is drilled in the maxillary bone shelf and a suture of non-absrobable matlerial is placed into the tough tissue of the medial palpebral ligament. The suture is then fastened to the prepared bone, Fig. 26.

This technique though safe, effective with minimal morbidity, is not reliable for the extirpation of maxillary-ethmoid squamous cell carcinoma or other tumours with propensity for diffuse involvement of adjacent structures. In such circumstances, conventional radical maxillectomy is employed. Also, in non-squamous malignant neoplasm, the medial maxillectomy is inadequate to deal with palatal, pterygoid or most orbital bone involvement.

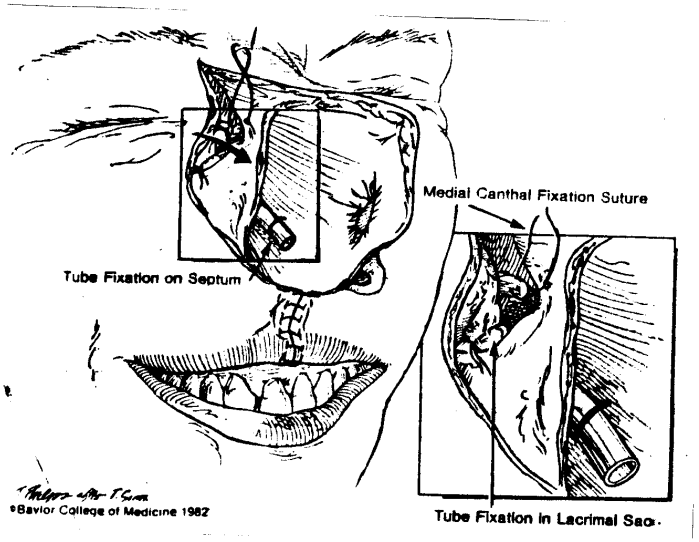

Fig. (26): Diagram shows technique for restoring lacrimal drainage and technique for reapproximating medial palpebral ligament. Inset, Detail of medial canthus fixation suture and pursestring fixation suture securing polythylene tube to lacrimal sac.

Cosmetically, the postoperative appearance is satisfactory especially when lip incision is not needed.

Craniofacial Resection:

The technique of cranio-facial resection for tumours of the paranasal sinuses was first described in 1954 by Smith et al and was subsequently developed most noteably by Ketcham et al (1963, 1973) and by Terz et al (1980).

It represents an attempt at more thorough surgical extirpation of antro-ethmoidal malignancy, beyond that achieved by the combination of maxillectomy and radiotherapy traditionally employed, by utilizing a combined approach which provides greater access to the diseased area (Valerie et al 1983).

Technique:

This surgical approach combines an extended lateral rhinotomy incision with a mid-line window craniotomy cut in the frontal bones. Through the craniotomy, the frontal lobes are retracted extradurally around the ethmoid labyrinth, exposure is enhanced by releasing cerebro-spinal fluid through a previously placed subarachnoid catheter. Resectability is determined at this point. The whole of ethmoid labyrinth on both sides with the cribriform plate is removed en bloc through the

lateral rhinotomy incision (Fig. 27 A and B). If the tumour extends through the cribriform plate, the lesion may still be resected by leaving the overlying dura attached to the specimen. Contraindications to further surgery are the following:

1- Superior extension of the tumour through the dura and into the frontal lobes.

2- Posterior tumour extension beyond the cribriform plate or the roof of the ethmoid complex to cause excessive traction on the optic nerve.

3- Involvement of both optic nerves.

4- lateral extension outside the boundaries of the fovea ethmoidalis and into the region of the superior orbital fissure.

This combined approach offers excellent visualization and access. Depending upon the extent of the tumour, additional resection of the overlying skin, nasal bones, frontal sinuses, orbital walls and contents, lateral wall of the nose, nasal septum, maxilla, dura and brain is possible.

Bone-cutting instruments are used to excise the block of involved bone and a margin of normal tissue from its attachments. The placement of these cuts varies with the extent of the tumour. A partial or total maxillectomy with or without an orbital exenteration is performed in conjunction with removal of the ethmoid complex.

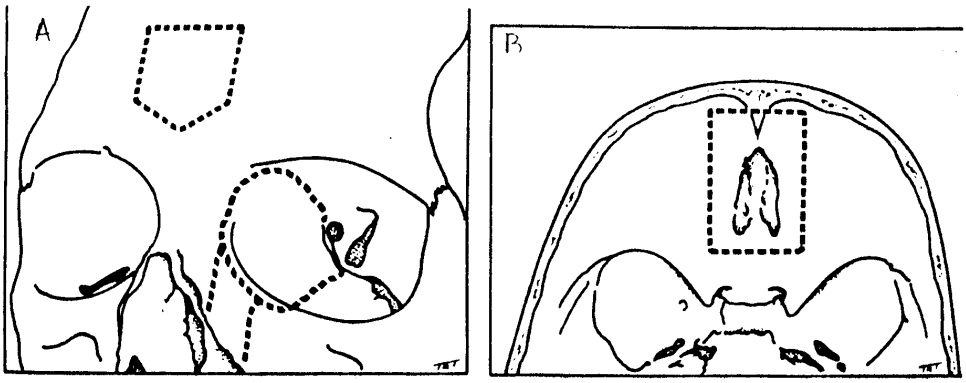

Fig.(27): A. Line drawing to show elxtended lateral rhinotomy and midline craniotomy window in the frontal bone. B. Diagram of the floor of the anterior fossa showing line of the osteotomies around the cribriform plate.

Repair is made in three layers. The superior layer is made by fascia lata to mend dural defects. A middle supporting layer of iliac-crest bone, septal cartilage, or posterior lamina of frontal bone provides rigidity to prevent dehiscence of anterior cranial contents into the nasal fossa. The inferior layer forms part of the lining of the common nasal and maxillectomy cavity and may consist of a rotated septal pedicle flap in selected cases, a free dermal graft, or a split thickness skin graft (Sisson and Becker 1981).

The facial cavity is lined with split-thickness skin grafts. The eyelid incision is closed but the lateral rhinotomy segment is left open in selected cases for observation. Once the cavity is healed, usually within 2 months, this defect is repaired. Closure of the incision in the philtrum and lower nasal alar region is completed at the time of the primary surgery. The craniotomy is closed conventionally. Sisson and Becker have been performing this procedure on selected cases during 18 cases and found it valuable in the management of advanced antro-ethmoidal cancer.

Sublabial Approach:

Anand and Conley (1983) explained the usefulness of this approach to the nasopharynx, paranasal sinuses, and the nasal cavity, and also in extensions in one or more of the adjacent areas.

It has been found useful in excising nasopharyngeal angiofibroma, certain melanomas, inverting papillomas of the nasal cavity and the paranasal sinuses, and malignant tumours of the maxillary and ethmoid sinuses which do not require a radical sinus excision.

Surgical technique:

1- Caldwell-Luc incision is carried out with the incision depth at the level of the bone and the lateral extent up to the maxillary tuberosity on both sides.

2- The periosteum is raised with the soft tissues as for a Caldwell-Luc on both sides and the infra-orbital nerve is identified and preserved.

3- At this stage a routine rhintoplasty incision is made in the limen nasi and columella, and the dorsum of the nose is skeletonized. The lateral extent of the incision is carried out to the lateral border of the pyriform aperture, and is connected medially to the transfixion incision along the floor of the nasal cavity.

4- The incision along the nasal cavity is then connected to the Caldwell-Luc incision, the lateral border of the pyriform aperture is identified, and dissection is carried out lateral to it along the ascending process of the maxilla to the root of the nasal bones.

5- Now the skin of the middle third of the face can be degloved from the skull all the way superiorly to the frontonasal suture line, infraorbital rim and the zygomatic process laterally.

6- The maxillary sinus is entered in the conventional manner anteriorly.

7- The pterygomaxillary space is entered by removing the posterior wall of the maxillary sinus.

In performing the medial maxillectomy, the three turbinates are removed underdirect vision. The ascending process of the maxilla need not be removed completely in all cases as deformity of the nose will occur following extensive removal.

8- The nasopharynx, postnasal septum and the pterygomaxillary space are well visualized through the nasal cavity and the maxillary sinus.

9- After completion of the procedure, the skin of the middle third of the face is redraped into its normal position and the incision sites are closed.

10- packing of the sinus and nasal cavity is meticulously carried out.

11- After completion of the procedure, the dorsum of the nose is taped to prevent collection of blood over the dorsum.

12- The Caldwell-Luc incision is closed and a posterior pack is used to obtain a dry nasopharynx and is removed on the third postoperative day.

Advantages:

1- Excellent exposure and haemostasis under direct vision.

2- Extensions and modifications can be devised by the surgeon to suit his needs.

3- The procedure is excellent in young patients, adolescents and children who develop no ugly facial scar.

4- It does not need a skin graft; healing is by secondary intention.

5- The functions of the palate are preserved as it is not incised in the procedure.

Complications:

1- Deformity of the nose may occur following extensive removal of the ascending process of the maxilla.

2- Improper packing of the nasal cavity and the maxillary sinus may delay the healing process and lead to epistaxis.

3- Early postoperative removal and poor follow-up may produce synechiae intranasally and extensive crusting. The crusting should be removed periodically using water irrigation.

4- Significant post-operative atrophic rhinitis may develop if proper medical treatment is not rendered after surgery. (Anand and Conley 1983).

Reconstruction of the Defect:

The raw surfaces of the entire cavity are lined with a split-thickness or dermal skin graft taken from the anterior thigh. The graft

should be between 0.016 and 0.020 inches thick. If the eye is retained, a fascial sling is employed on the under-surface of the orbit before it, too, is grafted. The skin graft is sewn in place with an absorbable suture and covered with a layer of Gelfoam. The bony cavity is packed with a continuous strip of gauze impregnated with antibiotic ointment (Fig. 28). This provides haemostasis and holds the skin graft in place. A soft silastic sponge is cut to fit the lower half of the cavity and is then inserted into the palatal defect (Sisson and Becker 1981).

If skin from the cheek was resected with the specimen, the defect may be reconstructed with a forehead pedicle flap (Fig. 29), or a sternomastoid myocutaneous flap (Fig. 30) and a split-thickness graft used to line the inner surface of either flap. Heavy preoperative radiation to the head and neck may require advancement of a tubed pedicle flap from a distant site (fig. 31). In institutions where the services of a maxillofacial prosthodontist are readily available a temporary surgical obturator is constructed and placed in the cavity immediately. This enables the patient to take liquids and soft foods immediately postoperatively until a permanent prosthesis can be constructed by the prosthodontist. The Silastic sponge works as well as a temporary surgical obturator and is less costly (Sisson and Becker 1981). Finally, the nasofacial and palpebral incisions are approximated with a 4-0 chromic or other absorbable suture for the subcutaneous tissue and with a 5-0 and 6-0 nylon suture for the skin. The packing is carefully removed 7 to 10 days postoperatively.

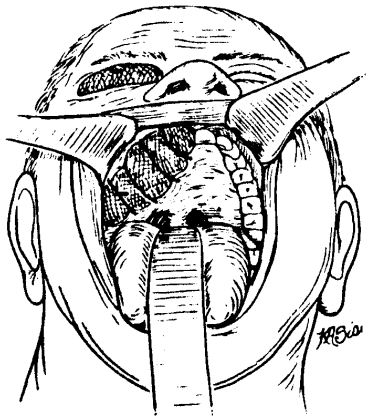

Fig. 28. Gauze packing sewn in place.

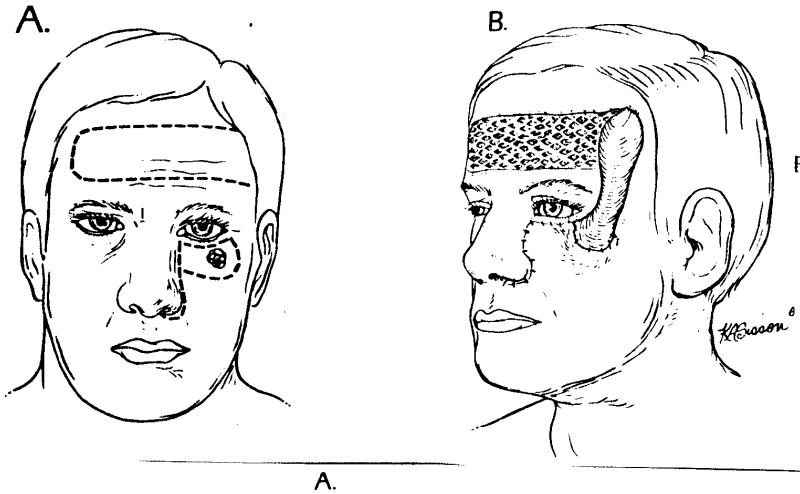

Fig.29 A and B Forehead pedicle flap.

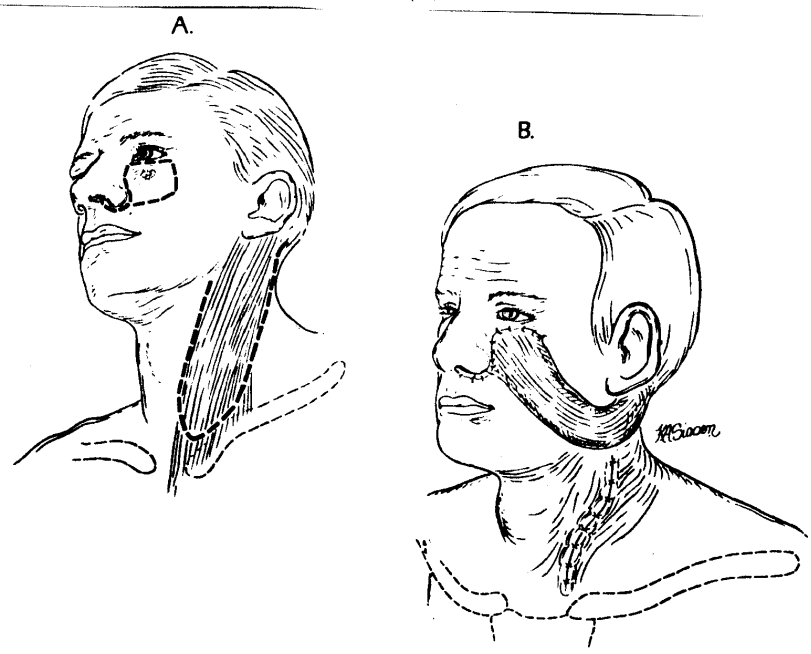

Fig.30 A and B Sternomastoid myocutaneous pedicle flap.

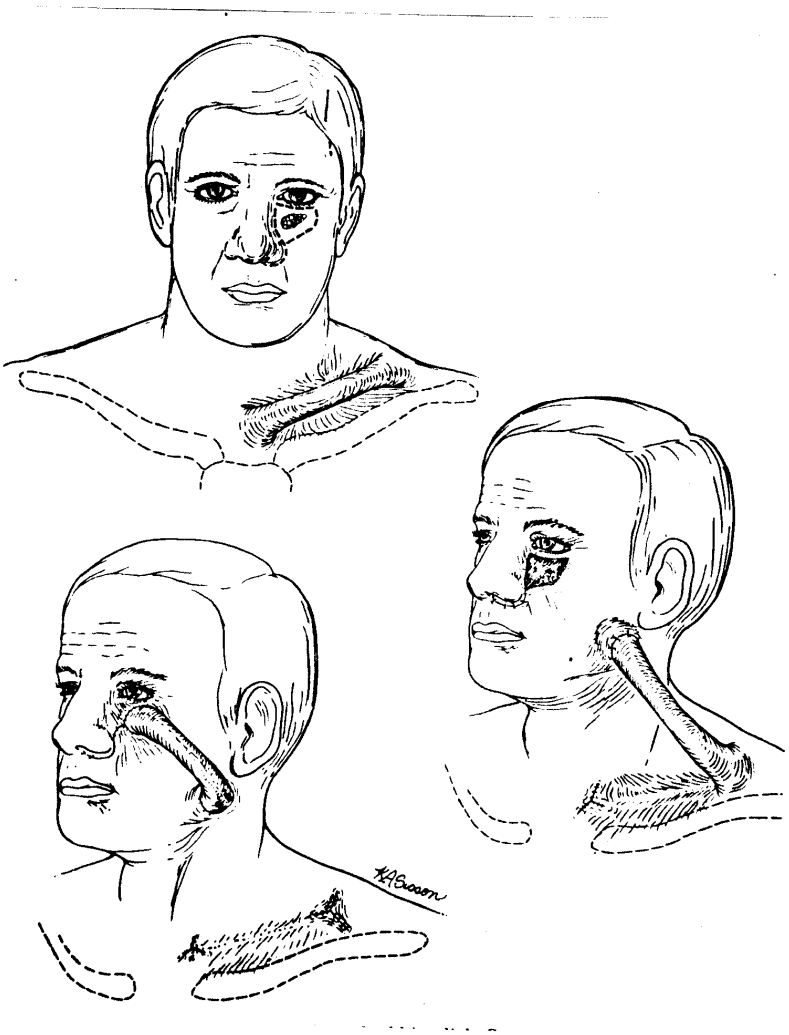

Fig.(31): Tubed bipedicle flap.

Weisberger and Hanke (1983) described the technique using local cheek and/or cervical skin as a rotation advancement flap, previously used for partial-thickness defects, for reconstruction of full thickness defects. Flap can be based anteriorly or posteriorly.

In the design of the anteriorly based flap, the incision begins from the superior-medial margin of the defect and extends along the ipsilateral lower eyelid, paralleling the ciliary margin at a distance of 4-5 mm. From this point, it extends along a curvilinear line from the lateral canthus to the preauricular crease at a point just above the tragus (Fig. 32 A). This latter portion should curve above a line drawn from the medial canthus to the tragus to minimize the risk of ectropion by suspending the lower eyelid superiorly. The incision then extends in the preauricular crease inferiorly and then curves posteriorly beneath the earlobe. At this junction, it can be extended into the postauricular region and converted into a bilobed flap or extended inferiorly on the posterior triangle of the neck following a vertical line just behind the external jugular vein until the incision crosses the clavicle. 2-3 cm beneath the clavicle, the incision is cut back anteriorly on the upper part of the chest wall to facilitate rotation and advancement.

The incision for the posteriorly based cervical facial flaps begins at the inferior medial aspect of the defect and extends along the melolabial fold and crosses the angle of the mandible at a point

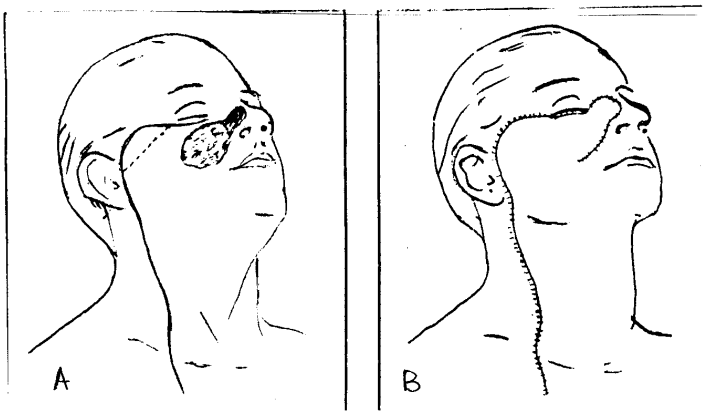

Fig. (32): A. Incision for anteriorly based cervicofacial flap.
B. Flap rotated and advanced into defect.

2 cm anterior to the facial artery notch. From this point, the incision curves posteriorly across the submandibular triangle and is terminated over the sternocleidomastoid muscle approximately 4 cm beneath the angle of the mandible.

With both flaps, a superior medial rotation advancement is performed with the line of closure falling in skin creases of the lower eyelid, along the alar-facial groove, and along the melolabial fold (Fig.32B). The anteriorly based flap places the remainder of the incision in the preauricular fold which is cometically superior to one crossing the mandible in a perpendicular manner, such as is required with the posteriorly based flap. Elevation is performed over the face in a plane immediately superficial to the parotid-masseteric fascia.

Weisberger and Hanke believe that the use of this technique for reconstruction of defects of the anterior part of the cheek and nasal alar groove offers some notable advantges:(1) the scar created by the line of closure tends to fall in natural junctures of the face such as the nasal-alar groove, the melolabial fold, the preauricular crease and the lower eye lid, (2) there is an excellent color match of the skin, (3) it offers excellent exposure for concomitant dissection of the parotid and cervical lymphatic compartment and (4) there is little donor site deformity and the primary closure is accomplished without the need to skin graft the donor site.

Problems encountered were: (1) Certain loss of contour of the face, (2) Some contracture of the flap, this could be decreased by another flap, contracture can lead to ectropion of the lower lid, deviation of the cartilagenous portion of the nose and a superior pull on the upper lip, (3) Loss of orbital floor due to concomitant maxillectomy causes additional deformity in the region of the eye. These effects can be improved by revision surgery.

Complications of Surgery:

1- **Haemorrhage:**

This may be immediate as is usually accompanied by marked oedema of the cheek and obvious bleeding. In the operating room, the bleeding sites are secured and a fresh pack is inserted. Bleeding is usually from the internal maxillary artery. Sisson and Becker advise that self-locking clips should be placed on the terminal branches of this vessel at the time of surgery. Delayed haemorrhage at 2 to 3 weeks after surgery is infrequent.

2- **Infection:**

This may occur due to the contamination by secretions from the nose, oropharynx, and paranasal sinuses. All these patients should be placed on parenteral antibiotics according to culture sensitivity. In postoperative craniofacial resection, the sequealae might be meningitis and/or brain abscess. Daily packing changes with aggressive debridement

of infected necrotic bone are required.

In the series of Sisson and Becker, patients receiving postoperative irradiation were less likely to have infection than those receiving preoperative irradiation.

3- **Cerebrospinal fluid leak:**

The three layered repair of the defect in the skull base in craniofacial resection introduced by Sisson and Becker lowered the occurrence of this leak. When it happens, packing and antibiotics can control readily the leakage.

4- **Loss of function and creation of cosmetic deformity:**

This relates to the amount of tissues removed. Anterior and posterior alveolar ridge defects include a substantial loss of the alveolar position of the maxilla and any associated teeth. These are readily repaired with prosthesis. Prosthodontic management of the hard palate defect can be relatively simple when the defect is localized to the hard palate (Laney 1978). Support for the prosthesis can be provided by natural teeth, the residual alveolar ridge in edentulous areas, and the residual palate. There is a recent trend for immediate reconstruction of the defects as this provides less postoperative problems both for the treating team and the patient especially as regards nutrition.

5- Complications related to radical neck dissection performed in some cases include exsanguinating haemorrhage due to any significant loss of skin-flap coverage or mucosal lining that exposes the carotid artery or its major branches, tissue necrosis and sepsis being the usual precipitating factors.

RADIOLOGICAL PROCEDURES:

It is a well known fact in radiobiology that an approximate expotential relationship exists between the dose of ionizing radiation administered to a cell population and the surviving fraction of these cells. Experimental studies have demonstrated that relatively low doses will inactivate a vast number of cells in a tumor, i.e., D_{37} (dose to reduce survival cell population to the original 37 percent) ranges from 100 to 250 rads in most biologic systems. Small microscopic subclinical tumour can be controlled with a dose of 4500 to 5000 rads in 5 weeks in better than 90% of the cases. However, a large tumour requires higher dose such as 6000 to 7000 rads in 6 to 7 weeks for inactivation or eradication of the entire cell population. For such advanced stage tumours, the radiation therapy is further handicapped by excessive cell tumour cell population as well as by the presence of large number of central hypoxic cells (Wang 1981).

The major strength of radiation therapy is to eradicate the actively growing, well-oxygenated cells in the periphery of a tumour.

During the past two decades, two conceptual approaches to combined radiation therapy and surgery have emerged; preoperative and postoperative radiation therapy. It is still debatable which of these two approaches is superior.

Preoperative Radiation Therapy:

Conventional preoperative radiation therapy:

This aims at the prevention of marginal recurrences, control of subclinical disease in the primary site or in the nodes, and technical convertion of inoperable tumours into operable ones.

It has the advantage of being performed with the tumour cells in their maximum oxygenation unlike postoperative irradiation. When combined with surgery, preoperative irradiation has been found to decrease both local recurrence and the incidence of distnt metastases. The disadvantages are that the exact tumour extent is obscured at the time of surgery, the surgery is delayed, and postoperative complications increase. The dosage employed in this conventional preoperative radiotherapy program is subcancerocidal consisting of 4500 rads in one month. This is followed in one month by radical surgery. This program is applicable to medium-sized or advnaced tumours with poor radiotherapeutic or surgical cure rates (Wang 1981).

Cancerocidal preoperative radiotherapy (Sequential postradiation resection):

The radiation dosage used in this program is cancerocidal, i.e, 6000 to 6500 rads in 6 to 7 weeks. The treatment protocol must be progressively reduced after 5000rads is achieved. It is followed by limited surgical resection of the primary left after irradiation. This approach has the advantage of avoiding excessive functional and cosmetic mutilation by surgery.

Postoperative Radiation Therapy:

This aims at the eradication of residual disease at the resection margins and subclinical disease in the neck nodes or implanted in the wound. The procedure is usually carried out 3 to 4 weeks after surgery when the wound is well healed. The dose is 5500 rads in 6 weeks following radical surgery and 6500 rads in 7 weeks following debulking procedures.

Modalities of Radiation Therapy:

Iradiation may be delivered to diseased tissue with internal or external media.

External radiation sources:

External radiation sources are more widely used and may categorized as seen in table 13.

Table (13): External Radiation sources:

	Source (Usual peak voltage)	Approximate meV equivalent (Range)
Ortho voltage	250-400KV roentgen ray	0.1-03
Supervoltage or	1000 KV roentgen ray	0.5
megavoltage	CS-137 teletherapy	0.6
	Radium teletherapy	1.1
	Co-60 teletherapy	1.2
	Linear accelerator	1.6 +
	(6, 9, 12, 18 meV)	
Ultra-high-Voltage	20 meV and above	
therapy (seldom		
used)		

The tools of radiation therapy used in the management of carcinoma of the head and neck are primarily megavoltage radiations, with energies at or above 1 million volts, and radio -active isotopes (Wang 1981).

Before approximtely 1950, studies of the effects of external radiation were based mainly on the use of orthovoltage. Low and medium-energy radiations are absorbed in direct proportion to the density of the tissues being irradiated. Thus bone, which is 1.8 times the density of soft tissue,

would absorb more of the delivered dose than would soft tissue, and it is more susceptible to secondary and scatter radiation because of its mineral content.

Megavoltage radiation energies possess certain inherent physical advantages such as a skin sparing effect; an increase in the depth dose, which is reflected by the fact that often a full course can be given without causing radiation dermatitis, a sharp beam which confines the irradiation to the lesion with minimum damage to adjacent normal tissues, and a bone sparing effect (Wang 1981).

The delivery of external radiation is planned for entry through bilateral specific skin ports or fields in precalculated fractions. Protective shielding devices incorporating 2 mm thickness of lead have been used for protection of normal tissues adjacent to diseased tissues that are being treated with orthovoltage therapy. These shielding devices are not effective with supervoltage radiation, because a minimum thickness of 5 cm would be necessary when cobalt-60 is used. However, the sparing effect and precision possible in directing supervoltage radiation somewhat offset the penetrating power (Laney 1978).

Electromagnetic radiations:

The present means used for radiotherapy by roentgen or gamma rays are the telecobalt "bomb" and the linear accelerator. The former

is not actually an x-ray machine but is used as a megavoltage x-ray one. The source of ionizing rays, in this case gamma rays having energies of 1.17 and 1.33 MeV, is several thousand curies of radio-active cobalt-60 housed in a shielded container, the bomb, with collimating devices and electrical circuits.

The linear accelerator, known as a linac or clinac, provides a compact source of x-rays in the range of 4 to 10 million volts. It has a metal tube that permits it to conduct an electromagnetic wave or microwave frequency with high efficiency. This tube acts as a wave guide where an elelctron is injected in vacuum with sufficient velocity, the electron is caught up by the wave and carried along by it, gaining energy and momentum from the wave. Either x-rays or electron beam can be produced.

Linear accelerators have the advantage of high output (i.e., 200 to 1000 rads per minute at isocenter) and compactness.

Electron beam:

Energetic electrons can be generated by a linear accelerator or a betatron. Varying energies are available including 6,9,12,15 and 18 MeV or above. The characteristics of electron beams are rapid dose build-up and sharp fall-off beyond the specified energy applied; thus protecting structures beyond the target.

Radioactive isotopes:

Radio-active isotopes are used interestially as a temporary or permanent implant. Radium 226, in the form of needless, was the isotope used most frequently in the past few decades. After-loading devices using angiocatheters with iridium 192 have been developed and extensively used in lieu of ractium needless in clinical practice for more safety. Such implants are removed after 2 to 3 days after deliverying a prescribed dosage to the tumour. Radon gas, a gaseous product of radium was used extensively in the past in the form of permenant interstitial implanted seeds, but has been largely replaced by gold 198 or iodine 125 grains or others.

Heavy charged particles:

Proton and alpha particles generated from a cyclotron have been used for radiation therapy in the past few years. They can produce sharp beam margins deep in the body. When they are slowed down in the tissues, they release their maximum ionizations shortly before stoppage, known as Bragg Peak phenomenon. By employing various thickness of absorber and also by using a rotating wedge disc of the absorber, it is possible to place the position of the Bragg Peak in a predetermined site and also to spread the Bragg Peak within the desired width of volume of irradiation and thus form a so-called modulator beam (Wang 1981). Although the relative biological effectiveness (RBE) is similar to x-rays, this technique may improve dose distribution and localization. This technique has been used

by radiation therapists at the Massachusetts General Hospital with the 160 MeV photons from the Harvard-MIT cyclotron and it is too soon to evaluate the results.

High LET radiation:

In order to overcome the problems of hypoxic central tumour cells hampering radiotherapy, currently radiations with high LET have been employed for clinical trials. LET is a term designated as energy transfer per unit length of ionizing track. Cobalt 60 and 200 KV x-ray have low LET, while fast neutrons and heavy particles have high LET. Experimentally, the higher the LET the lesser the oxygen enhancement ratio (OER). Fast neutrons generated by the cyclotron have an intermediate OER of around 1.5 to 1.8 and yet maintain adequate penetration similar to the cobalt 60 beam. Although fast neutrons have the advantages over x-rays of low OER and lack of repair of sublethal or potentially lethal damage, there is no improvement in survival rates, although local tumour control is definitely superior (Wang 1981).

Other particulate radiations such as pimesons, helium, carbon, and neon ions have been explored for clinical radiation therapy. No clinical data are available to indicate their usefullness (Wang 1981).

Radioactive sutures:

Goode et al (1979) used I-125 seeds in vicryl suture as a radioactive suture in 24 cases of advanced malignancy of the head and neck including 3 cases of cancer maxilla aiming at surgical excision of as much tumour as possible then implantation of any residual tumour with this radioactive suture which remains as a permenant implant (half life of 60 days).

The I-125 seeds are a low energy gamma-ray emitting radioisotope with half-life of 60 days. Each seed produces between 0.4 and 0.5 millicuries (mCi) of radiation. At the end of a year, only 11/2% of the initial radioactivity remains. The seeds are slid into the center of 15 cm length of No. 1 polyglactin 910 (Vicryl) suture after removing the central core.

I-125 has the advantage over the radioactive seeds, e.g radon-222, gold-198 and iridium-192, in having higher therapeutic effectiveness and low complication rate, and also in being a low energy Gamma source, so there is minimal radiation hazard to the patient's contacts. These can be more than one implant according to sites of involvement.

The number of seeds needed is estiamted using a computer program. The sutures are sewn into the tumour area so that the seeds lie equidistant from each other, about 7 mm apart. The suture ends are held in position with small vascular clip to minimize later slippage. An x-ray istaken a

day or two after surgery to check on the position of the radio-opaque seeds. The polyglactin suture is absorbed in 60-90 days.

The average I-125 implant produces a continuous low dose of radiation of 10,000 to 16,000 rads to the tumour volume over the effective life of implant (about 84 days).

Complications occurred in 20.7% of cases. They are in the form of skin sloughs, fatal carotid blow out secondary to a neck fistula and injury to the superior laryngeal nerve. Goode et al came to the following conclusions:

1. Sutures are relatively easy to insert.
2. Sutures can be placed adjacent to vital structures without complications.
3. Excision and I-125 implantation technique work best when accompanied by external radiotherapy given immediately before or after implantation.
4. The technique is indicated only for localized tumours which cannot be completely removed surgically.
5. The techniue appears to be more effective for slower growing tumours since the dose is low.

Goode et al felt that the use of the above mentioned technique for any unresectable tumour or close margins tumour may produce improved

local control when used as the primary treatment of certain advanced head and neck malignancies. It is also useful in palliation as it produces a definite decrease in tumour size for at least 2 months with low morbidity.

More time and more cases are required for accurate evaluation of this technique in treatment and/or palliation of advanced head and neck malignancies.

CO_2 laser:

Rontal and Rontal (1983) described the use of CO_2 laser for the treatment of recurrent carcinoma at the base of the skull. Conventionally, recurrent carcinomata at this site were treated by radiotherapy and surgery in the form of massive resections of the orbit, floor of the anterior cranial fossa and the sinus complex. These techniues were less than satisfactory.

Cases to be treated with CO_2 laser had to fulfill certain criteria: (1) These patients must be radiation failures. (2) CT scanning must ascertain absence of intracranial extensions as this procedure is not useful for intracranial spread because of their inability to ascertain the extent of the disease or guard against iatrogenic injury to the surrounding brain. (3) Recurrence must be proved by biopsy. (4) There must be direct access to the tumour site for visualization, delivery of the laser and follow up. (5) Follow-up with a microscope is a must as an outpatient.

Patients who had no pre-existing cavity, such as maxillectomy cavity or orbital exnteration, had to have surgical access before laser delivery through a transmaxillary sinus exenteration procedure or a transnasal route. The technique used continuous wave CO_2 laser with suprapulse modes. The laser was used to remove all mucosa in the involved area for a wide field around the recurrence.

Thebone was heated white, in the continuous wave de, to be sure no tumour was missed, or was removed with the supra pulse mode. Where a breach of dura was produced, the defect was identified using the microscope and a plug of fat and muscle, taken from the upper lip, was accurately placed to stop the leak. Postoperatively, the area is packed.

Follow up comprised biopsy and microscopic examination. Any early detected recurrence was treated as an outpatient. The procedure can be repeated more than once for recurrences.

This technique causes minimal destruction to the surrounding tissue and produce little morbidity to the patient in the post-operative period. The laser is administered with or without a general anaesthesia.

The CO_2 laser cannot be used for en block resection of cancer at the base of the skull. Distinct specimen margins cannot be identified after the use of the laser because of the tissue vaporization.

Rontal and Rontal finlly concluded that the use of the surgical laser provided through the operating microscope is an easily controlled means of tumour removal. Good follow-up is possible and is easily accepted by both the physician and the patient. As seen in these cases, properly selected patients followed closely can have surprisingly long-term palliation, living comfortable and productive lives.

Complications of this technique were:

1- CSF rhinorrhoea is the most serious complication and is treated by a muscle fat plug followed by intravenous antibiotics.

2- Introperative bleeding which originates from the sphenopalatine or internal maxillary artery. This can be avoided by early identification of the vessel and clipping it.

3- Indiscriminant fulguration of posterior ethmoid vessels or pressure packing could lead to amaurosis.

4- perichondritis of thyroid cartilage with subsequent web formation.

5- laser burn of teeth enamel or of a lip.

Ossoff et al (1983) conducted a retrospective view of all patients undergoing CO_2 laser surgery by members of the Department of Otolaryngology- Helad and Neck Surgery at Northwestern University Medical School from January 1, 1980 through December 31, 1981. On basis of study of 204 cases as regards technical problems and complications encountered, Ossoff et al developed a list of precautions for laser surgery: 1- Some

exposure to laser education is necessary for all involved staff. 2- Protection of the patient and operating room personnel from corneal injury by protective glasses. 3- All exposed skin and mucous membrane of the patient outside the surgical field has to be protected from any misdirected laser impacts. Protection of the endo-tracheal tube from either direct or reflected laser beam irradiation is of primary importance as it may cause ignition of oxygen tube leading to catastrophic intraluminal air wayfire. 4- Adequate smoke evcuation from the laryngoscope, bronchoscope or oral cvity to facilitate proper visualization. 5-Stabilization of the microscope stand to prevent the microscope with attached laser from falling over.

From the above mentioned precautions Ossoff et al evolved a safety protocol which had to be revised several times to reach the present safety protocol shown in table 14.

Table (14): Present Safety Protocol.

+'	Rusch red rubber endotracheal tube wrapped with reflective metallic tube is used in cases where general anaesthesia is required.
2-	Cuff inflated with methylene blue colored saline.
3-	Cuff protected with saline-saturated cottonoid which have to be counted like surgical sponges at the end of the procedure.
4-	Use of the operating platform to reduce the risk of airway fire.
5-	Use of the lowest concentration of oxygen permitted by anaesthesiologist.

6- Protection of the cornea.

7- Protection of the skin. This can be achieved by a double layer of moistened surgical towels.

8- Adequate smoke evacuation.

Operating under the directives of the safety protocol, Ossoff et al had an overall rate of complications of 6%.

Hyperbaric oxygen:

Already it has been stated that hypoxia causes cells to be protected from irradiation. Treatment in a hyperbaric oxygen chamber with the patient breathing at a pressure of 30 lb/in² (3 atmospheres absolute) has been shown to improve significantly the response of metastatic lymph nodes in the neck and also of extensive tumours of the head and neck that normally do not respond well to conventional radiotherapy. The procedure is time consuming and troublesome for some patients especially those suffering with claustrophobia as patient is placed in a small pressure chamber for at least one hour. Absolute immobility is essential for accuracy of treatment and this, coupled with barometric problems such as difficulty in clearing the eustachian tubes and variations in temperature within the chamber, make the treatment a considerable trial, especially for the more intelligent patient. In view of this, the course of treatment is carried out six sessions given twice a week over a period of three weeks, the patient receiving

a total dose of 3600 rad, which is approximately equivalent to the standard dose of 6500 rad given in daily fractions over six and a half week, (DBL Skeggs 1979).

Hypoxic cell sensitizers have recently been introduced experimentally. These include metronidazole (Flagyl), RO-07-0582 (misonidazole), and others. Metronidazole sensitizer is only moderately efficient and failry high concentrations are required to produce worth while radiosensitization for clinical use. Clinical trial use of RO-07-0582 has been carried out in the United States and other countries. The amount of drug administered is limited by the immediate symptoms such as anorexia, nausea, vomiting and neuropathy. These trials are still under trial and need sometime to be evaluated (Wang 1981).

Hyperthermia:

Experimentally, it has been found that hypoxic cells are more sensitive to elevated temperature than normally oxygenated cells. When hyperthermia is combined with x-rays, the OER is reduced. currently, local heating of the tumour can be given in conjunction with localized radiation therapy.

H.D. Fairman (1982) published the results of his study on 22 patients of head and neck cancer (predominantly squamous), including one case of antral carcinoma, treated by adriamycin and bleomycin with adjuvant hyperthermia to near lethal temperatures (42 - 44°C). Half the patients had previous irradiation while the other half had synchronous irradiation.

Post-irradiation caes had adriamycin i.v. 60 mg/M² and bleomycin 30 mg twice weekly to a total dose of 90-120 mg 50% i.m. and 50% intratumour injection. Synchronized irradiation cases were given almost the same doses. Irradiation was given synchronously on a Cobalt 60 unit. Hyperthermia was effected by a 200 watt microwave Magnetron 200 unit. The author concluded that a high tumour response rate was achieved by this regieme especially when iradiation is synchronized. Further work is needed to get conclusive results.

Radiosensitivity:

The degree of sensitivity to irradation can, to a large extent, be correlated with the histological structure of the tumour. In practice, tumours are grouped as shown in table 15.

Table (15)*

		Dose required
High sensitivity	Tumours of lymphatic origin	2000-4000rad
	Embryonic tumours	
	Highly anaplastic tumours	
Moderate sensitivity	Reticulum cell sarcoma	5000-6500rad
	Squamous cell carcinoma	
	Some adenocarcinomas	
Low sensitivity	Tumours of mesodermal origin, such as fibrosarcoma, osteogenic sarcoma	Minimum of 7000 rad

* Adopted from Scott Brown; diseases of the ear, nose and throat.Basic science.

Radiocurability:

Radiocurability is not a synonym of radiosensitivity. Many of the more radiosensitive tumours, such as anaplastic carcinomas, although responding locllly exceptionally well to the iradiation, may well, by the very nature of their natural history, have dissiminated beyond the bounds of the area being treated. Thus, there may be a successful outcome to the treatment locally but the patient succumbs as a result of wider local extension of the disease or possibly distant metastases.

Complications of radiation therapy:

1- Skin reactions are not severe because with megavoltage irradiation the maximum dosage received occurs at several millimeters below the surface of the skin. These include erythema, desquamation and tanning of the skin. These are exaggerated with the use of actinomycosis D, or any trauma such as washing with soap, exposure to sunlight, or even irritation from coarse clothing.

2- Effects of the irradiation on the normally growing tissues adjacent to the tumour especially the brain, spinal cord and bones. These effects can be early or late and may be potentiated by the use of adjuvant chemotherapy especially antibiotic chemotherapeutics. This effect can even be latent and occur after the completion of radiation therapy (recall phenomenon).

3- Radiation around the eye can cause corneal ulcers, enophthalmos,

keratitis, iritis, conjunctivitis and later cataract.

4- The effects on the upper digestive tract especially when potentiated by actinomycin D, include mucositis, dysphagea, glossitis, pharyngitis, oesophagitis, gingivitis and erythema. Erythema usually gradually progresses to patchy, fibrinous exudation. This is the ideal stage that should be reached by the completion of treatment and shows that the normal tissues are approaching tolerance. If these reactions are intense, it necessitates the use of nasogastric or i.v. feeding (Feldman 1982). These effects can be exaggerated by any form of trauma such as smoking, drinking strong alcohol, infection or the use of irritant mouth washes. Treatment may need be suspended temporarily till the reaction improves.

6- Dental complications include trismus, osteoradionecrosis, xerostomia, teeth decay, atrophy of the zygoma, abnormal occlusion and delay in the development of the maxilla and mandible in irradiated children.

6- The possible development of a second primary cancer in the irradiated field which may develop a number of years after the completion of original treatment.

7- Effects on the ear are either early or late. Early effects include inflammation of the skin of the auricle and external canal and of the mucosa of the middle ear. Late effects include atrophy of the membranous labyrinth especially the annular and spiral ligaments and osteoradionecrosis.

8- Other complications include cellulitis of the check and facial tissues, orbital infections and hypopituitarism.

CHEMOTHERAPEUTIC PROCLEDURES

The term chemotherapy was first used by Paul Erlich in 1907 to describe the use of chemicals against parasitic organisms and he is regarded as the father of chemotherapy. There has been a constant increase in the number of chemical compounds developed and since the beginning of this century, two events are worthy of special note. The first was the development of the radiomimetic alkylating agent, the second was the synthesis of the anti-DNA and anti-RNA drugs referred to as antimetabolites. All of the cancer chemotherapeutic agents available at present are toxic to both tumour and normal body cells. The apparent selective toxicity relates to differences between the body's normal vital stem cells and the tumour cells in respect to the total number and the percentage in a proliferative stage. Following the administration of an effective chemotherapeutic drug, a percentage of the proliferating normal and tumour stem cells will be destroyed. Killing both normal and tumour stem cells recruits non-proliferating and tumour cells into the proliferating pool. If after the elapse of the intervl necessary for the normal stem cell population to return to a non-proliferating state, further chemotherapy is possible, the host may survive repeated course of treatment leading to a progressive destruction of the tumour cell population. Each dose of an effective drug kills a constant percentage of cells, referred to as "first order kinetic kill" and repeated doses of a single agent are not likely to kill the last tumour cell (P. Clifford 1979).

The same drug, which can be curative when only a small number of tumour cells is present, would be completely useless in treating a large advanced tumour. Therefore, chemotherapy to be curative drug treatment should be given immediately after local treatments have removed the bulk of the tumour mass (Price and Hill 1979).

Hill and Price (1975) classified the drugs used in the treatment of head and neck cancer on basis of the effect of 24 hour exposure to various chemotherapeutic agents on the survival of the normal bone marrow stem cells and tumour cells. Drugs could be divided into 2 categories: those which after an initial reduction in survival do not cause increasing damage to normal bone marrow stem cells with increasing dose (class II), and those where the bone marrow stem cell kill does increase with increasing dose (Class III). In both cases, there is maximal selective kill of malignant stem cells.

Anticancer drugs exert their maximal cell killing effects during different phases of cell cycle. This means that drugs can be selected for combination chemotherapy which exert their lethal elffects primarily in different phases of the cycle.

The following are the chemotherapeutics used in head and neck cancer; the first three being the most commonly used.

1- **Methotrexate:**

This is the most extensively used chemotherapeutic in patients with head and neck squamous carcinoma. It acts by tight but reversible binding to dihydrofolate reductase, thereby inhibiting the conversion of folic acid to tetrahydrofolate, a necessary precursor for thymidylic acid biosynthesis. The resulting inhibition of DNA, RNA, and protein synthesis can be avoided by administering citrovorum factor (leucovorin), which is converted directly to tetrahydrofolate, thereby bypassing the methotrexate-induced enzymatic block. Leukopenia and thrombocytopenia are dose limiting toxicities. Overall response rate ranges from 40 to 50 percent in patients with recurrent or metastatic cancer (Wolf and chretien 1981). Intermittent weekly or biweekly schedules of 40 to 60 mg /m^2 achieve better response rates and less toxicity than either daily low dose regimens or intermittent 5-day courses of 5 to 25 mg/day each month, the weekly schedule being superior as demonstrated by comparison. Other recent studies have suggested very high response rates to weekly high dose regimens (1-7.5g/m^2) followed by leucovorin rescue. Methotrexate administration by intraarterial infusion has been extensively studied. The results of single drug and combination chemotherapy will be summarized later with treatment disciplines.

2- **Bleomycin:**

These are a group of antibiotics isolated from streptomyces verticillus and were first investigated for their antineoplastic qualities by Professor Hanna Umezawa (Japan) in 1965. Chemically, bleomycin is a variable mixture of a group of related water soluble glycopeptides.

A unit dosage is used in which 1 mg equals to 1.5-1.75 units (15 units per ampoule). It acts by producing scission and fragmentation of single strand DNA and inhibition of thymidine uptake into DNA. In combination, it renders tumour cells susceptible to attack by other drugs. Recommended dosage is 2 to 3 times weekly administration of 15 units/M^2. A total dosage of 300 units should not be exceeded. It is detoxified and excreted through kidneys even in presence of significant renal impirment (Bone et al 1979).

Bleomycin has a unique specificity against epidermally derived cells. Squamous cell carcinoma cells are approximately 100 times more sensitive to bleomycin than are normal tissue cells or other tumour cells. Other characteristics are a relative lack of hematologic toxicity, a potential cell-cycle synchronizing effect with low dose continuous infusions, and a potential with synergism. Mucositis and pulmonary fibrosis are dose limiting toxicities. Response rate for bleomycin vary from 6 to 45 percent and response durations are generally less than 3 months (Wolf and Chretien 1981).

Best results obtained by bleomycin are in combination with radiation therapy as there is close similarity in effect on DNA. Mucositis resulting of this combination is allowed to heal through rest from treatment which is repeated with both modalities. A much greater reduction in tumour cell mass is obtained with combination than is obtained with radiotherapy alone.

Borre et al reviwed results obtained by some authors who treated patients with maxillary antrum epidermoid cancers with combined bleomycin and radiotherapy compared to other modalities. This is shown in table 16.

Table (16): Comparison of results with combined bleomycin chemotherapy and radiation therapy to other modalities.

Study	No.of cases	Therapy	Results	
Suzuki et al 1976	115	Surgery ± or x-ray	33% 5 year-survival	Non random series
	29	Surgery ± or x-ray + BLM	62% 5 year survival	
Matsumura et al 1973	22	No control group	40% 2-year survival	Low BLM dose various routes of administration

The total dose of bleomycin in the latter series are generally below 150 mg suggesting that treatment with maximal levels (300 units) is important.

3- **Cis - platinum:**

This is the most recently introduced chemotherapeutic drug with significant activity in head and neck squamous carcinoma. The postulated mechanism of action of cis-platinum is related to its alkylating properties, whereby complementary strands of DNA are cross-linked and DNA replication prevented. Dose limiting renal toxicity has been minimized by vigorous hydration and diuresis, Hematologic toxicity is usually moderate and transient (Wolf and Chretien 1981).

Data from trials of single agent cis-platinum indicate a response rate of 25 percent to 30 percent in heavily pretreated subjects. Both high dose (120 mg/m² IV bolus) and low dose (20 mg/m² IV bolus daily x 5 days) regimens appear to have similar overall response rates (Wolf and Chretien 1981). Results achieved with biweekly high dose cis-platinum are comparable to those achieved with weekly high dose methotrexate.

4- **Actinomycin D:**

this is a derivative of streptomyces parvulus. It acts through inhibition of DNA and RNA synthesis.

5- **Vincristine:**

This is an alkaloid extracted from the periwinkle plant. It acts through binding proteins in tumour cells and causes mitotic arrest. It also affects similar proteins making up an important part of nervous tisue.

6- **Cyclophosphamide:**

It is an alkylating agent which acts by substituting alkyl groups for hydrogen atoms especially in DNA, and causing cytotoxicity due to breaks produced int he DNA molecule.

7- **Adriamycin:**

This is an anthracycline antibiotic derived from streptomyces peucetius varcaesius. It exerts its cytotoxic effect by binding to DNA and disrupting the synthesis of RNA (Feldman 1982).

8- **Other single agents:**

These include hydroxy urea, 5-fluorouracil, chlorambucil, methyl CCNU, hexamethylmelamine and 6-macroptopurine. Newer drugs such as mitomycin C, PALA (L.aspartic acid N-(phosphonacetyl) disodium salt), m-AMSA (acridinyl anisidide), vindesine, dibromodulcitol, and gallium nitrate are currently being evaluated in head and neck cancer.

Complications of chemotherapy:

Side effects common to most chemotherapeutic agents are: alopecia, stomatitis, nausea, vomiting and bone marrow depression with subsequent neutropenia and thrombocytopenia.

Individual drug side effects are:

Methotrexate: myelosuppression, septicemia, skin rash and renal toxicity.

Bleomycin: effects on integument and mucus membranes include erythema and rash and less frequently vesiculation, pruritis, hyperpigmentation and hyper-keratosis. These occur in up to 50% of patients often at a total dosage of 150-200 units and at about 2 to 3 weeks. Pulmonary complications range from pneumonitis to pulmonary fibrosis and occur in 10% of cases. They were fatal in 1% of cases. This is more with age over 70 years and a total dose of more than 300 units. Fine basilar râles are the earliest sign. Careful auscultation and weekly chest-x-ray are suggested for early detection. Treatment is by combined antibiotics and corticosteroids (Bone et al 1979).

Actinomycin D: diarrhoea, skin rash and severe inflammation and necrosis when it extravasates outside the vein.

Vincristine: mixed motor-sensory and autonomic neuropathies, depression of deep tendon reflexes, paraesthesias of the fingers and toes, weakness, muscle pain, sensory impairment, constipation, ileus and rarely bowel obstruction.

Cyclophosphamide: haemorrhagic cystitis, urinary bladder fibrosis and even carcinoma of the bladder, water intoxication, immunosuppression and sterility. When used in combination with Actinomycin D, the neutrophil count may fall to zero requiring the use of high dose I.V. antibiotics at the first sign of infection or fever.

Adriamycin: cardiac complications such as arrhythmias, congestive heart failure followed by death. It has a mitogenic, teratogenic and carcinogenic potential. It also potentiates the effects of irradiation and can proudce a "recall phenomenon". Extravasation of the drug at the site of injection can cause a severe chemical burn.

Management of complications of chemotherapy:

1- Blood transfusion with packed red cells are necessary in marrow depression and Hb fall.

2- Platelet transfusion should be given in severe thrombocytopenia (Less than 20,000/mm^3) to prevent bleeding.

3- The early use of high dose I.V. antibiotics.

4- Hyperalimentation to sustain a good nutritional status necessary for wound healing and for facing debilitation resulting from the disease and therapy.

5- Antimetics to treat nausea and vomiting.

6- Local therapy of oral ulcerations and pharyngitis.

7- Good dental hygiene.

8- Temporomandibular joint exercise.

9- Psychosocial support and counselling for both patient and family.

The introduction of the concepts of cell cycle kinetics provided an opportunity for a rationale for combining optimal and safe systemic therapy with the best forms of local treatment.

Price and Hill (1979) have concluded the following principles to be applied in the chemotheapy of head and neck cancer:

(1) Drug treatment should be given when the smallest number of malignant cells i.e. in combination with surgery and/or radiotherapy.

(2) Chemotherapy should be given over periods of 24-36h. in intermittent course (approximately 3-4 week intervals to allow marrow recovery thus reducing toxicity.

(3) A knowledge of the kinetic classification of the antitumor agents is essential for safe chemotherapy. The toxicity of class II agents to bone marrow cells is not dose dependant, therefore they may be added to combinations without reducing their dose. Combinations of class III drugs will be additively toxic to normal bone marrow and so doses should be reduced proportionately.

(4) Giving small daily doses of drugs should be avoided as this leads to drawing bone marrow cells into cycle and killing them.

(5) When selecting drugs for combination therapy a knowledge of their killing effects in different phases of the cell cycle may be helpful.

Multiple drug chemotherapy is now practiced widely, P. Clifford (1979) suggested the following criteria for multiple drug chemotherapy:

1- Each drug in the combination should be active when used alone against the tumour.

2- The drugs should have different mechanisms of action.

3- The toxic effects of the drugs should not overlap. Price and Hill (1979) considered the best response rates in the world are being achieved by using a combination of vincristine, bleomycin, methotrexate, 5-fluorouracil and hydrocortisone given over 24 hours with a subsequent leucovorin rescue. Apart from a response rate of over 70 percent this approach has the advantages that the patient spends only one night out of every 3 to 4 weeks in hospital, absence of severe bone marrow depression, only 6 percent occurrence of alopecia and less nausea and vomiting and the possibility of offering intensive chemotherapy without ruining the quality of the patient's life.

Despite the safety and effectivness achieved by the kinetically based approach, Price and Hill proposed the following precautions which they thought should always be observed in all cases of head and neck cancer receiving chemotherapy:

1- Never give another treatment cycle unless blood count has returned to its original level.

2- Patients with impaired renal function receiving methotrexate must have an extended leucovorin rescue i.e. at least 3 hours longer than normal.

3- Doses of cyclophosphamide, adriamycin and 5-fluorourecil should be halved in patients who have had thoracic, abdominal or pelvic irradiation.

4- Doses of class II agents should be reduced proportionately if more than one of them is included in a combination.

5- Adriamycin should not be given to patients with a history of cardiac failure. The total dose of adriamycin must not exceed 550 mg/m². The dose should be halved in patients who have impaired hepatic function.

6- Patients receiving drugs which are excreted in the urine, e.g. methotrexate and hydroxyurea, must be adequately hydrated and passing urine while having the drug.

7- Bleomycin should not be given to any patients with impaired respiratory function.

IMMUNOTHERAPEUTIC PROCEDURES:

Immunotherapy is the administration of any agent or treatment which stimulates, modifies, or restores in a specific or nonspecific fashion, host immune reactivity and results in the regression or prophylaxis of tumours (Wolf and Chretien 1981).

Immunotherapeutic agents are arbitrarily divided into two categories: those that actively increase host immune reactivity specifically through vaccination with tumour cells or cell extracts or non specifically through administration of agents having this effect, such as bacterial organisms or their products, and those that passively augment immune reactivity.

Passive immunotherapy consists of the administration or removal of serum containing either tumour specific antibodies or antigen - antibody complexes.

Bacille Calmette-Guerin (BCG) and corynebacterium parvum stimulate relatively intact immune mechanisms while levamosile, transfer factor and thymus extracts act to reconstitute impaired or deficient immunologic function.

Morton (1972) believes that adequate and sensitive methods would detect specific antigens in many human cancers.

The host-tumour relationship is complex and attempts to treat patients by immunological manipulations are associated with hazards such as the enhancement of the tumour by antibody, blocking by an excess of soluble antigen and the development of reticuloses (Clifford 1979).

BCG was first tried by Mathe (1969) who reported prolongation of the disease-free interval in children with lymphocytic leukemia. Further studies were made and favourable responses were reported by Donaldson (1973) in a group of patients treated with BCG and Methotrexate of which many patients had recurrent disease following prior surgery or radiotherapy. BCG has been used intradermal, intrapleural, intralesion, intravenous, by scarification, multiple puncture, Heaf gun injection and intracavitary. Uncontrolled manipulation of the immune system in an imperfectly understood situation and death due to BCG hypersensitivity are the dangers encountered (Wolf and Chretion 1981).

Corynebacterium parvum (C. parvum) has been used in the same way as BCG. Fisher et al (1975) have shown that C.parvum given with cyclophosamide produced a more effective tumour inhibition than could be achieved by giving either of these agents separately. Currie and Bagshawe (1970) had previously studied the effects of this combination and had stressed that the maximal antitumour effect could only be achieved by correctly timing the administration of immunotherapy. These studies suggest that cancer chemotherapy as well as its direct effect against the tumour, corrects some tangle in the immune system and as a consequence subsequent immunotherapy is more effective. The overall results of BCG and C.parvum as regards recurrence rate or survival showed no significant improvement.

Levamisole is an antihelminthic imidazole that has immune reconstituting activity believed to be due to biochemical properties similar to the thymic hormone, thymopoietin. It influences cell-mediated immune response (T lymphocytes) but has little effect on the B lymphocyte (humoral) system. It acts by altering lymphocyte cyclic nucleotide levels, altering haptene binding for the induction of delayed hypersensitivity, or influencing the expression of thymocyte antigens on precursor cells (Wolf and Chretien 1981). The effects of this drug are most evident in immuno-suppressed patients, and trials of its effects on patients who have undergone surgery and/or radiation are in progress and resulted in significant increase in disease-free interval especilly for patients with stage IV tumours.

The thymus controls the development and maturation of the T-cell system by secreting a family of polypeptide hormones, thymosin. Thymosin fraction V is the most extensively studied. Thymosin promotes lymphocyte maturation and the induction and functional expression of specific types of lymphocytes such as killer; helper and suppressor cells. The preliminary trials do not indicate a potential benefit of thymosin immunotherapy during or after radiation but these trial are still preliminary and further studies need to be made.

A number of other immunotherapeutic agents are being studied in both animals and cancer patients. These include the interferons, interferon inducers such as poly I:C and pyran copolymer, the lymphokines, tumour antigens, antitumour antibodies and various chemical agents such as glucan, muramyl dipeptide, and isopurinosine.

More precise knowledge and understanding is required before immunotherapy is brought into general use.

TREATMENT DISCIPLINES AND COMBINATION TRIALS:

Since Schuknect (1951) performed total maxillectomy, en bloc resection of maxillary cancer for the first time, a number of reports have been published concerning modified procedures and their long term results. With surgical therapy alone, the 5-year survival never exceeded 50% (Konno et al 1980).

Recently, intra-arterial infusion chemo-therapy via the superficial temporal artery has come into use. When surgery was combined with regional chemotherapy and radiotherapy, long-term results of the treatment of maxillary cancer improved markedly.

As yet, no universally standardized method exists for the treatment of cancer of the maxilla. Many problems are yet to be solved, such as the kinds and dosages of cancer chemotherapeutics, routes of their administration, irradiation dose and the extent of extirpative surgery. Since the majority of the patients conditions are usually diagnosed at an advanced stage, conservative treatment with any single modality in the past has led to many failures. Attempts, therefore have been made to treat this area aggressively, combining irradiation with surgery, chemotherapy and hyperbaric oxygenation. It is also debatable whether radiation should be used postoperatively or preoperatively.

Batsakis (1981) reviewed the results of treatment and 5-year survival of several series treated by several authors with various modalities between 1950 and 1971.

The neoplasms represented were all carcinomas and the series contained selected as well as unsected cases. The near 25% survival average indicates that failure to establish early diagnosis thwarts an effective management.

These trials were summarized with their results of treatment in table 17.

Sato et al (1970) devised a unique combined therapy conssiting of irradiation, chemotherapy and reduction surgery in order to reduce the volume of the tumour, and succeeded in reducing the irradiation dose as well as doses of chemotherapeutic agents without diminishing the effects of radiotherapy and chemotherapy.

Mihashi et al (1972) reported that the 5-year actual survival rate was 62% in patients treated by 6000 rads of ^{60}Co, intra-arterial infusion of 5-FU and maxillectomy.

P. Clifford (1979) reviewed the results of treatment of 102 patients with cancer of the head and neck, including the maxilla, treated at King's College and the Royal Marsden Hospital from lst March 1974 with synchronous VBM and chemotherapy as follows:

Table (17): Carcinoma of the paranasal sinuses: Results of treatment, 5-year survivals.

Author	Form of treatment	Location of tumour	No.of cases	5 - year survival %
Capps (1950)	Irradiation before surgery	Nose and sinusels	52	27.0
Schall (1951)	predominantly surgery	Nose and sinuses	202	26.5
Mattick and Streuter(1954)	Irradiation before surgery	Antrum	68	10.0
Larsson and Martenson(1954)	Irradiation before surgery	Nose and sinuses	294	23.0
Struben(1957)	Surgery before irradiation	Sinuses	76	13.0
Snelling (1957)	Irradiation before surgery	Antrum and ethmoids	115	28.0
Dalley (1957)	Various of combinations of irradiation and surgery.	Antrum and ethmoids	215	28.5
Tabb (1959)	Radical surgery following irradiation	Antrum	60	10.0
Baclesse et al (1960)	Irradiation	antrum	94	20.0
Frazel and Lewis	Radical surgery	Antrum	163	29.0
Zange and Sholtz (1963)	All methods	Antrum	116	25.0
Spratt and Mercado (1965)	Primary irradiation	Antrum	69	15.0
Pointon (1969)	Megavoltage only	Antrum and ethmoids	105	25.0
Pointon(1969)	Intracavitary radium	Antrum and ethmoids	146	28.0
Tabb and Barranes (1971)	Various combinations including chemotherapy.	Antrum	102	27.0

VBM

Vincristine 2 mg I.V.

6 hour later

Bleomycin 30 mg I.M.

18 hours later

Methotrexate 200 mg I.V. infusion/24 hours

folinic acid 50 mg I.V. start ----- 9 mg I.V. 6 hourly X 6

VBM was given in pulses with intervening conventional external supervoltage radiotherapy usually given as 180-200 rads daily for 5 days per week.

All patients had squamous cell carcinoma with T_1 or T_2 lesions + nodes or T_3 or T_4 lesions $\pm$ nodes.

Results of treatment with synchronous VBM and radiotherapy are shown in table 18 in comparison with results of conventional radiotherapy alone in patients treated in the 5-year period immediately prior to 1974.

Table (18): Results of primary treatment with synchronous VBM and DXT.

Patients entered into programme	102	(92)
No local resolution	10	(38)
Local recurrence	18	(28)
Primary treatment failure	28	(66)
Local control plus +ve distant metastasis	3	(4)
Complications of treatment causing death	5	--
Overall distant metastases	8	(17)
Overall local disease control	74	26

* The figures in brackets refer to the comparative group of patients who were treated before 1974 with conventional radiohterapy alone.

After 50 months the disease free survival and crude survival rate were 56.5% and 56% respectively compared to 21.9% and 24.5% respectively for the control group of patients.

P. Clifford (1979) could conclude that there is no need to proceed to surgery after this treatment except in presence of definitive histological proof of residual disease. Also, he concluded that synchronous chemotherapy did not add to the difficulty of subsequent surgery or lead to more complications than is usual following conventional radiotherapy although other authors reported augmentation of complications of radiotherapy when combined with chemotherapy.

Konno et al (1980) analysed the results of treatment of 70 patints with maxillary cancer, they treated by maximum combination of radiation, intra-arterial chemotherapy, subsequent sufficiently extensive maxillectomy and primary reconstruction during the period between December 1971 and May 1979.

1- Radiation therapy was performed with Linac x-ray, 6mV, by cross two field irradiation with the use of a 45° wedge filter. The angle of wedge filter and the cross angle of the directions of the irradiation were modified depending upon the direction in which the tumour spreads. Radiation dosage was 200 rads/day 5 times a week, with 6000 rads/30 day as the goal. In some cases antrostomy is performed after delivery of 1000 rads or before surgery to confirm the diagnosis.

2- For chemotherapy, 250 mg 5-FU dissolved in 20 ml solvent were injected through acatheter inserted retrograde in the superficial temporal artery as one shot 30 minutes before irradiation, 3 times a week for 15 times in average to a total dose of 3750 mg.

3- Maxillectomy was performed 3 weeks after termination of radiation and chemotherapy and was extended as necessary. In some cases where residual or dispersed tumour cells were suspected, 2000 rads of radiation were delivered postoperatively.

4- Reconstruction was performed as follows:

a- In cases where tendons of the pterygoid and temporalis muscles were exposed around the mandible, or the subcutaneous tissue of the face was extensively resected together with the maxilla, or the cranial base was widely exposed, the whole wound was covered primarily usually with a deltopectoral flap.

b- In cases in which the periorbital bones and the orbital periosteum were extensively resected, or the orbital fat was also removed with the resultant exposure of the inferior and medial rectus muscles, the orbital contents were covered with a graft of fascia lata for suspension and then primarily covered usually with a forehead flap.

c- Reconstruction of the palate utilizing the pedicles of the flap or a secondary bone grafting under the flap was performed as necessary, to minimize the postoperative facial disfigurement and functional disturbance.

d- When the tissue defect was not large, the wound surface was covered with a free skin graft.

5- Radical neck dissection was performed only if cervical lymph node metastasis was present and not done prophylactically.

6- In cases of undifferentiated carcinoma, chiefly carcinoma simplex, antrostomy was not performed except for histopathological examination. Instead, a treatment with bleomycin and mitomycin C was used in 3-5 courses when the general condition was stabilized after maxillectomy.

Cervical lymph node metastasis was noted in 7 patients (10%), while orbital exenteration was performed in 23 patients (32.9%).

The actual and relative survival rates are shown in table 18.

Table (18): Long-term survival rate of maxillary cancer after combined therapy.

Survival	All 70 cases		52 cases of squamous cell carcinoma	
	Actual survival rate(%)	Relative survival rate(%)	Actual survival rate(%)	Relative survival rate(%)
3 years	75.2	80.2	85.2	91.1
5 years	64.1	71.9	71.1	80.2

Causes of death after this combined therapy are shown in Table 19.

Table (19): Causes of death after combined therapy.

Cause of death	Squamous cell carcinoma (N=52)	Adeno-carcinoma (N=6)	Undiff-carcinoma	Total no. of cases
Local recurrence	2	1	0	3
Distant metastasis	3	1	5	9
Senility and other causes	3	1	0	4
Total No.of cases	8	3	5	16

Konno et al came to the following conclusions:

1- Although primary reconstruction is opposed by many authors as it may mask early local recurrence, it can be rationalized by the fact that early recurrence, evenwhen detected early, has a poor prognosis. Moreover, primary reconstruction is far more beneficial to patients, as regards to the elffect of reconstruction, financial and physical burden, and time loss, than is secondary reconstruction in patients having postoperative complications such as cicatricial contracture of the face, ocular disfigurement or partial defect not readily closed by prosthesis.

Advantages of primary reconstruction can be summarized as follows:

a- Repair of as much of the facial contour and function as possible.

b- It enables the performance of as much resection of tissues as necessary.

c- It freuently eliminates the need for orbital exenteration even in presence of destruction of the orbital bony wall, since it enables the resection of the orbital bony wall as well as periosteum and a portion of the orbital fat tissue, then cover and suspend the exposed orbital contents with the fascia lata and the flap.

d- It makes the postoperative treatment easy when sectioning the pedicle of the flap after 3 weeks where the wound is covered with a thick flap.

e- Even if the cranial base is widely exposed, that portion can be covered with a flap and supplementary irradiation can be performed as necessary.

2- The high 3-year and 5-year relative and actual survival rates were attributed by Konno et al to be due to low local recurrence rate, which is in turn may be the cause of infrequency of secondary cervical lymph node metastasis.

3- Konno et al believe that it is necessary to intensify the systemic care after surgery in elderly patients so as to prevent death due to senility. Even in T4 cases, they thought that complete cure is possible through extended total maxillectomy providing it is followed by supplemental irradiation.

4- Undifferentiated carcinoma of the maxillary sinus has frequent occurrence of lymph node and distant metastasis. This seems to be accelerated by preoperative procedures such as aspiration and currettage at the primary lesion.

Konno et al, therefore, suggested some other principles for treatment of undifferentiated carcinoma of the maxilla as follows:

a. Antrostomy and handling of the tumour preoperatively should be minimized.

b- Extneded maxillectomy should be performed after irradiation and chemotherapy, especially in cases of T3 up, and should be followed by chemotherapy and immunotherapy.

5- Maxillary cancer in the aged is frequently complicated by systemic disease and is apt to be advanced.

Combined radiotherapy, chemotherapy and total maxillectomy in the aged gave more satisfactory results than combined therapy without surgery especially when old patients find difficulty in attending remote outpatient clinic and are sensitive to pain caused by postoperative local treatment and complications. In contrast to the conclusions of Konno et al (1980), other authors e.g. Sisson and Becker (1981) do not believe that combining preoperative chemotherapy with irradiation adds anything to patient.

The present policy of management of carcinoma of the antrum of the Head and Neck Service at North western University as performed by Sissona nd Becker (1981) includes the following:

1- Exploratory antrostomy is performed on all cases.

2- T_1 and T_2 lesions are treated by either subtotal or radical maxillectomy at the time of exploration if the diagnosis can be firmly established, or within several days of the exploration after permenant sections are reported. Adjuvant high dose methotrexate chemotherapy or postoperative iradiation are added in selected cases.

3- T_1 and T_2 lesions also receive postoperative irradiation when margins of the tumour aresuspected or when recurrent disease is evident.

4- T_3 and T_4 lesions are debulked at the time of an exploratory antrostomy. The medial antral wall is removed to facilitate evacuation and drainage of necrotic tumour during radiotherapy.

5- T_3 and T_4 tumours receive 6000 rads of cobalt 60 irradiation over a period of 4-6 weeks. Four to 5 weks later, an exploratory antrostomy is again performed to ascertain the amount of residual, tumour. While ablative surgery, particularly in T_4 cases, usually includes radical maxillectomy with orbital exenteration, T_3 cases, after irradiation will often resolve completely obviating the need for orbital exenteration. It is mandatory with T_3 and T_4 tumours that the bony borders of the maxillary and ethmoid sinuses are removed completely with the entire floor of the orbit. The orbitl contents are suspended by using a strip of temporalis fascia as a sling or by using a folded dermal graft. Adjuvant chemotherapy is given in selected cases.

6- For massive T_3 or T_4 lesions demonstrating invasion of the anterior cranial fossa, the sphenoethmoid recess or the sphenoid sinus, radical surgery is performed after reviewing the results of preoperative radiation. Craniofacial resection is often perfomed in these cases after anterior craniotomy to determine the resectability of the tumour. If no dural invasion is detected, a partial or complete maxillectomy with or without an orbitl exenteration is performed. This resulted in long-term palliation and, in some cases, cure in 25 patients with stage IV sinus cancer.

7- For all N_1 and N_2 lesions the neck is included in the preoperative irradiation and later is treated surgically by unilateral or bilateral neck dissection.

Patients who refuse treatment, patients with inoperable tumours due to naso-pharyngeal or skull invasion, and patients with distant metastases or unresectable regional metastases can benefit from such procedures as palliative decompression, debulking and drainage through a large antrostomy. Palliative doses of radiation therapy and chemotherapy used in combination with cryosurgery are all helpful in reducing discomfort.

Ervin et al (1981) reviewed most of recent studies on chemotherapy for head and neck, aiming at the comparison of results obtained by single and combined chemotherapeutic agents. The trials reviewed included previously treated and untreated patients. The results of these results are shown in tables 20, 21, 22, 23.

Table(20): Activity of single-agent chemotherapy in untreated head and neck cancer.

Drug Regimen	No.of trials	No.of patients evaluated	>50% response%	Complete response%
Methotrexate leucovorin calcium	3	38	52	3
Cisplatin	2	16	69	0

Table (21): Combination chemotherapy for untreated head and neck cancer.

Drug Regimen	No.of patients	No.of evaluated	>50% response%	Complete response%
Cisplatin methotrexate leucovorin calciurn,bleomycin sulfate.	4	75	77	21
Cisplatin, bleomycin	3	71	76	23
Cisplatin, methotrexate-leucovorin	1	20	60	0
Cisplatin,vincristine sulfate, bleomycin	1	35	80	23
Cyclophosphamide, vincristine, methotrexate, fluorouracile, bleomycin	1	16	58	not reported
Continued Vincristine methotrexate, flurouracil,bleomycin.	1	30	87	not reported

Table (22): Activity of single agents in previously treated head and neck cancer.

Drug Regimen	No.of trials	No.of patients	>50% response%	Complete response%
Cisplatin	3	68	34	9
Methotrexate leucorin calcium	3	73	52	3

Table (23): Combination chemotherapy for previously treated head and neck cancer.

Drug Regimen	No.of trials	No.of patients evaluated	>50% response%	Complete response%
Methotrexate leucovorin calcium,cisplatin, Bleomycin sulfate	7	169	45	12
Cyclophosphamide, methotrexate, fluorouracil,vincristine.	3	77	35	17
Cisplatin, bleomycin	3	22	32	5
Hydrocortisone,vincristine, methotrexate, fluorouracil,bleomycin	1	20	55	not reported

Cisplatin and methotrexate had similar response rates with low complete response rates in both single-agent and combination chemotherapy in previously treated patients. Response rates are slightly superior in untreated patients. The duration of response in all series is 2-4 months. High-dose methotrexate has no benefit more than low-dose methotrate.

There is no notable difference in the partial or complete response rates between single agents and combination chemotherapy. Toxicity being more with combination chemotherapy.

Ahmad et al (1981) analysed retrospectively 59 cases of squamous cell crcinoma of the maxillary sinus during the period 1955 to 1979. Ninety five per cent of patients had T_3 and T_4 lesions. Forty seven patients were treated with irradiation using a cobalt 60 teletherapy in a dose beween6.500 to 7500 rads given during a period of 6-8 weeks given daily, five days a week.

Nine patients had major surgery before irradiation, two had intra-arterial perfusion of hydrogen peroxide in conjunction with irradiation, and one patient had chemotherapy.

Results of treatment are shown in table 24.

Table (24): Initial treatment modality and control of primary tumour.

Treatment	T	No.of cases	Controlled No.(%)	Uncontrolled No.(%)
Radiation	2	1	...	1/1(100)
	3	16	6/16(37.5)	10/16(62.5)
	4	28	9/28(32.1)	19/28(67.9)
	Indefinite	2	...	2/2(100)
Surgery and Radiation	3	4	4/4(100)	....
	4	5	2/5(40)	3/5(60)
Other and Radiation	3	1	...	1/1(100)
	4	2	1/2 (50)	1/2 (50)

The results are somewhat poorer when radiation was used as the only modality of treatment. Actuarial 5-year survival in T_3 cases initially treated with radiation and including cases of recurrence treated by surgery was 38.4%. When surgery was used in T_3 lesions before irradiation, the 5-year survival was 75%.

Survival in T_4 lesions was 33.2% with initial irradiation while combined surgery and radiation yielded 33.3% survival. Overall acturial 5-year survival was 39.3%.

Ahmad et al finally concluded that, in selected cases, surgery followed by irradiation will be of greater benefit than either modality alone. Radiation therapy could cure locally about 1/3 of cases with less favourable condition. For recurrent cases after radiation, surgery yielded increased cures.

Lee and Ogura (1981) reviewed 96 cases of previously untreated primary carcinoma involving the maxillary sinus without metastasis, at the time of diagnosis, who received complete treatment at the Division of Radiation Oncology and the Department of Otolaryngology, Washington University School of Meldicine during the period 1960-1976.

Sixty-one patients were treated by radiation followed by surgery and 35 patients by radiation alone.

Table 25 shows the numbers of patients, stages and primary site control in each modality.

Table (25): Staging, treatment and primary site control.

	T_1	T_2	T_3	T_4	All stages
Radiation then surgery	5/7-71%	12/14-86%	13/21-62%	12/19-63%	42/51-69%
Radiation alone	-	2/3	0/6	3/26	5/35-14%
All patients	5/7-71%	14/17-82%	13/27-48%	15/45-33%	47/96-40%

Radiation alone consisted of 4000-5600 rads tumour close for 9 patients 6000-7000 rads for 25 patients and 8400 rads for one patient. All but 2 patients treated by surgery received their radiation preoperatively and these 2 received radiation after grossly or microscopically incomplete resection. Tumour dose ranged between 4800 and 7100 rads.

In 40% of patients, radiation was delivered through a single anterior port usually combining cobalt and 24 Mv x-rays. A right angled wedged pair of anterior and laterl fields were used in 34%. The remaining patients received right angled pairs without wedges, acutely angled wedges, or lateral ports. Patients presenting with neck nodes received 2400 to 6000 rads to the neck.

Radical maxillectomy with orbital exenteration was carried out in 27 patients. Twenty-one patients had radical maxillectomy with sparing the eye and 13 patients had partial maxillectomy. Radical neck dissections were performed in 3 patients.

the overall 2-year and 5-year NED survivals were 30% and 26% respectively. There were no 5-year survivors in the radiation alone group and only three 2-year survivors.

Table 26 shows the 2-year and 5-year NED survival for both modes of treatment.

Table(26): 2 and 5-year NED survival

Treatment	Overall %
Combined treatment	
2-year NED survival	26/61-43%
5-year NED survival	18/47-38%
Radiation alone	
2-year NED survival	3/35-8.33%
5-year NED survival	0/23-0%

NED survival = No.of patients surviving without any reappearance of cancer.

Robin and Powell (1981) revised 528 cases of carcinoma of the nasal cavity and paranasal sinuses registered at the Birmingham and West Midlands Regional Cancer Registry, England from 1957 to 1972. They compared the results of different modalities of treatment and came to the results shown in table 27, in which the 5-year survival percentage of patients treated by the different methods are recorded. Surgical treatment alone in their study gave the best results, 46.5% in male patients and 50% in female patients and a total of 47.7%, while treatment by radiotherapy alone gave the worst prognosis.

Table (27): Prognosis by treatment:

Treatment	Male Subjects			Female Subjects			Total%
	Total No.	After 5years	%	Total No.	After 5years	%	
Radiotherapy alone	90	15	16.7	68	19	27.9	21.5
Surgery alone	43	20	46.5	24	12	50.0	47.7
Radical surgery and radiotherapy	50	19	38.0	27	9	33.3	36.2
Non-radical surgery and radiotherapy	121	37	30.5	105	42	40.0	35.0
Total	304	91	29.9	224	82	36.6	32.76

Oster (1981) surveyed 15 trials of anterior chemotherapy for locally advanced squamous cell carcinoma of the head and neck including the maxilla in which chemotherpy was followed by radiotherapy and/or surgery. These are shown in table 28.

Table (28): Anterior Chemotherapy Trials.

Source	Patient No.	Chemotherapy	Respone rate(CR+ PR) %	CR %
Schaefer et al 1980	20	Cisplatin	--	--
Spaulding et al 1980	29	Cisplatin & Vincristine sulfate+bleomycin sulfate	100	38
Price and Hill 1980	76	Vincristine&methotrexate+ bleomycin+fluorouracil	75	-
Glick et al 1980	29	Cisplatin + bleomycin	48	0
Tejada et al 1980	66	Methotrexate + Cisplatin	63	5
Kaplan et al 1980	9	methotrexate + Cisplatin +bleomycin+mitomycin	89	-
Ervin and Miller 1980	21	High-dose methotrexate	52	0
Wolf and Makuch 1980	68	Cisplatin + bleomycin	53	7
Hong et al 1979	39	Cisplatin & bleomycin	76	21
Wittes et al 1979	73	Cisplatin ±bleomycin± high dose methotrexate± vinblastine sulfate	52	7
Elias et al 1979	22	Cisplatin & bleomycin, high dose methotrexate	73	18
Tayl[illegible] et al 1979	10	High-dose methotrexate	80	0
Kirkwood et al 1979	24	High-dose methotrexate	54	0

Leone and Ohruma 1979	8	Cisplatin + bleomycin +high-dose methotrexate	85	38
Petrovich et al 1979	12	Vincristine & high-dose methotrexate	-	0
Total %	-	---	65(309/477)	10(39/401)

CR = Complete clinical remission.

PR = Partial clinical remission(> 50%).

Results of most trials were preliminary and it is unclear whether these high response rates could be translated into improved survival and increased frequency of cures.

Oster concluded that anterior chemotherapy is preferred to posterior chemotherapy because it is given at a time when the tumour has a better vascularity and better cell kinetics, and when the patient is in better general, nutritional, performance and immunologic status. He also concluded that chemotherapy conducted with radiotherapy has disclosed unacceptable toxic effects without producing better response rate than those achieved by anterior chemotherapy. Anterior chemotherapy, moreover, may make inoperable tumours operable, may reduce tumour size allowing less extensive surgery and/or radiotherapy, and may permit easy interpretation of the responsiveness of the malignant tumour to chemotherapy. The unsettling aspects of anterior chemotherapy were the residual cancer cells and being a short-term treatment.

Sakai et al (1983) reviewed the history of treatment of maxillary sinus carcinoma in 773 patients treated in the Departments of Otolaryngology, Osaka-Kaisei Hospital, Okobe University medical School and Department of Radiology, osaka University, medical School between 1957 and 1979.

The history of their treatment of maxillary sinus carcinoma is divided into 4 periods each characterized by a major procedure that followed irradiation:

1- From 1957 to 1966: 282 cases were treated by irradiation (7000 rad/35 fractions/7 weeks) followed by maxillectomy performed on 40% of cases. There were hardships imposed upon patients by the radical approach.

2- From 1967 to 1971: 191 patients were treated by radiotherpy (7000 rad/35 fractions/7 weeks) followed 5-FU intraarterial infusion in a total dose of 2000 mg. Maxillectomy was restricted to cases where local recurrence was histologically confirmed but remained as frequent as 40.3%.

3- From 1972 to 1975: 166 patients were treated by irradiation (5000 rad/25 fractions/5 weeks) followed by 5-FU infusion (in 68% of patients) and intrasinal currettage. Maxillectomy to conquer local recurrence was conducted in 30.1%. Despite satisfactory suppression of local recurrence, curretage led to dissimination of tumour cells.

4- From 1976 to 1979: 134 patients were treated by radiotherapy (5000 rad/25 fractions/5 weeks) followed by 5-FU infusion and cryosurgery for reduction of tumour mass. Another regiment used in this period was immunotherapy with the cell wall skeleton of Nocardia Rubra, which appreciably reduced recurrence rates. 21.6% of patients underwent maxillectomy.

Table (29): Statistical Analysis of maxillary sinus carcinoma over four periods.

Period, Radiation dose,therapeutic land mark	No.of cases	Cumulative survival Rate *	Cumulative local non recurrence rate	Maxillectomy	Satisfaction of maxi-llectomy	Neck melastasis	Distant metastasis
1957-1966	282	35.2±2.9%	not	114	40	4	7
7000 rad, maxillectomy		19.8±2.4%	calculated	(40.4%)	(14.2%)	(1.4%)	(2.5%)
1967-1971	191	36.6±3.5%	21.5±3.3%	77	46	5	10
7000 rad, 5-FU infusion		25.1±3.1%	15.8±2.9%	(40.3%)	(24.1%)	(2.6%)	(5.2%)
1972-1975	166	56.5±3.9%	38.9±3.9%	50	22	6	16
5000 rad, 5-FU infusion currettage		39.4±3.8%	34.5±3.9%	(30.1%)	(13.3%)	(3.6%)	(9.6%)
1976-1979	134	64.3±4.3%	48.3±4.4%	29	15	7	4
5000 rad, 5-FU infusion, Cryosurgery		54.4±4.8%	45.2±4.5%	(21.6%)	(11.2%)	(5.2%)	(3.0%)

* Rate ± standard error.

The chronological periods, treatment and results are shown in table 29.

Steady improvement in therapeutic performance is evident in the cumulative survival rates which have been 19.8% for the first period, 25.1% for the second, 39.4% for the third and 54.4% for the fourth period.

Sakai et al (1983), after reviewing the results, believe that the best possible treatment for maxillary sinus carcinoma available at present consists of combination of (1) Co 60 gamma-ray irradiation (5000 rad/25 fractions 15 weeks, 2 portals with Wedge pair filter), (2) continuous intra-arterial infusion of 5-FU in a total dose of 2000 mg during the radiotherapy (practically for 4 weeks), (3) and the reduction of tumour mass mainly by cyrosurgery. Supplementary therapy entails early antrostomy for inspection and daily vacuum aspiration to remove necrotized tissue.

Recurrences are treated by maxillectomy except small lesions that can be controlled by intracavitary irradiation and/or cryosurgery. Cervical metastases, if movables are treated by radical neck dissection.

Sakai et al considered the 45% decline in the need for maxillectomy to be a good achievement towards the general tendency of retention of eating and communicating function as well as appearance Sakia et al reported the following advantages of cryosurgery:

1- The absence of bleeding and severe pain.

2- Retention of room for the surgeon to determine the extension of sites to be frozen, enabled by the repeatd procedures.

3- Less involvement of surrounding normal tissue.

4- Little mechanical irritation which minimizes distant metastases.

5- Suggested suppression of recurrence by the so-called cryo-immunity.

They believe that cryosurgery has significantly contributed to the welfare of maxillary sinus carcinoma patients who now enjoy higher 5 year survival rates than before without facial disfigurement.

Difficulties encountered in crysorugery included judgement of extension of tumour infiltration, protection of neighbouring important organs such as the brain and eyeball from overfreezing, accurate replacement of the probes through a narrow opening, the relief of pain, and the appropriate disposition of sequestrum which remains long after operation.

These problems have necessitated several attentions: (1) The antrostomy should be as wide as possible. (2) Cryosurgery should be divided into several weekly milder procedures to avoid the possibility of invasion of the surrounding organs. (3) Currettage should be done at the end of initial treatment to remove the necrotized or surviving mass.

Introduction of currettage into primary care of maxillary sinus carcinoma has greatly reduced local recurrence rates. Nevertheless, this progress has not necessarily been associated with increased survival rates before the advent of cryosurgery. This is due to reduced distant metastases and reduced mortalities from illnesses other than carcinoma.

Pointon et al (1983) treated 50 patients of proven squamous cell carcinoma of head and neck including mxilla in stages III and IV. Patients were treated with megavoltage (4 mV) irradiation to the tolerance dose given in five daily fractions per week to a total of 15-16 fractions in 3 weeks. Methotrexate 100 mg/m² was administered as a single intravenous injection 24 hours prior to commencing radiotherapy (MTX1) and agin 14 days later (MTX2). Twenty-four hours after each dose, drug concentrations were determined by radioimmunoassay (RIA), Folinic acid was administered to those patients in whom the 24-hour concentration was $> 0.5\ \mu m$.

Complete resolution of disease was achieved in 50% of patients, the median duration of response being 24 months and 45% for 3 years.

Pointon et al believe that this combination has proved to be practical and produced regression of tumour beyond that produced by irradiation alone.

Shidnia et al (1984) made a retrospective study on 121 patients with malignant tumours of paransal sinuses, 98 of which involved the maxillary sinus, treated at Indiana University Medical Center between 1960 and 1980. The center has practiced a combined modality of radiation therapy at a total dose of 60 to 66 Gy in 6-61/2 weeks followed by surgery after 4 weks, which included maxillectomy and neck dissection in cases with lymph node metastasis at the time of presentation. Some patients prior to 1970 had surgery followed by a course of radiation therapy and the patient with overt distant metastasis was treated with palliative radiation therapy alone.

32 patients had a course of radiation therapy followed by surgery. Most of these were treated between 1970-1980 with a pair of anteroposterior and latral fields angled 5-10° with the use of 45° wedge filter to a total of 66Gy in 61/2 weeks. Those who had lymph node metastses at the time of presentation recived a homolateral field of irradiation including the entire neck to a total given dose of 50 Gy, A boost of 10-15 Gy to the lymph nodeswas then given with the apparatus ranging from Cobalt 60 to an electron beam energy of 10 to 13 MeV in 61/2 weeks followed by surgery in 4 week which included a total maxillectomy and neck dissection in the case of neck involvement.

22 patients had surgery followed by a course of a radiation therapy similar to the previous group with the difference that the area of the

surgical defect was filled with bolus material varying from water bags to an acrylic prosthesis.

55 patients received only radiation therapy to a total dose of 30-60 Gy due to the extent of disease and presence of distant metastases.

Results of treatment are shown in table 30 in terms of 30 months survival for different modalities of treatment.

Table (30): Survival of patients according to treatment primaries.

Method of treatment	Maxillary	Ethmoid	Sphneoid	Frontal	Total
Radiation followed by surgery	(9)/30	-	-	-	(9)/30
Surgery followed by radiation	(7)/18	(3)/5	(1)/1	0	(11)/24
Radiation alone	(3)/40	(2)/12	(1)/2	(0)/1	(6)/55
Total	(19)/80	(5)/17	(2)/3	(0)/1	(26)/109

The number in parenthesis represents absolute survival for 30 months. A total of 13/54 patients (24%) in the first 2 groups failed in the primary site and 4/54 (7%) failed in the neck. Of the whole 109 patints analysed, 16 died of distant metastasis. As a whole about 50% of patients died of metastasis and intercurrent disease.

Shidnia et al (1984) concluded that combination of radiation therapy and surgery is the treatment of choice for this primary. With well planned therapy, it probably would not make any difference wether irradiationis before or after surgery. They believe that elective neck radiation should be given after surgery for large tumours (T_3 or T_4) as well as for patients with lymph nodes. There is no evidence that the opposite neck should be treated electively, unless there is a large tumour with bilateral neck nodes or massive unilateral involvement, where postoperative elective radiation therapy can be helpful. They found no correlation between histology and the presence of lymph node metastasis or survival and thought that recurrence in the primary site could be rescued by additional radiation therapy or surgery.

Treatment of certain histological types:

Geopfert et al (1983) on base of their retrospective study of 66 cases of minor salivary gland tumours of the nose and paranasal sinuses, who were treated in the University of Texas, M.D. Anderson Hospital Tumour Institute, have suggested that the treatment of these tumours is best perfomed with ultra-radical surgical resection with adjuvant preoperative and postoperative radiation therapy. Surgical resection includes combined carniofacial exenteration of the anterior cranial fossa and ethmoids and exploration of the middle cranial fossa for removal of possible tumour extension along the maxillary nerve as these tumours characteristically extend perineurally and intraneurally. Adjuvant preoperative and postoperative

radiation is mandatory for local control. Fast neutron irradiation has been used with apparently notable improvement in local control.

The results of treatment with minimum of 2 years follow up are shown in table 31.

Table (31):

Results of treatment (2 years or more)

	No.	Local control No(%)	Survival No(%)
Surgery only	20	9/19(47%)	8/20 (40%)
Planned surgery and irradiation	34	26/34(76%)	16/34(47%)
Irradiation only	2	-	-
Irradiation plus chemotherapy	3	-	-

Lehrer and Roswit (1978) believe that radiation therapy is clearly the treatment of choice in sinus lymphoma with the role of surgery limited in most cases to obtaining biopsy specimens and performing drainage operations. The optimal radiation dosage suggested is 5000-6000 rads in 5-6 weeks. While Fierstein and Thawley (1978) treated a series of 88 patients with head and neck lymphomas, including 15 cases of nose and sinuses, by radiation therapy in doses ranging from 4000-5000 rads. In stage I lymphoma,

adjuvant chemotherapy with a COPP regimen including cyclophosphamide, vincristine, procarbazine and prednisone, was given. All patients with stage IV were treated with chemotherapy. Most patients with localized disease were trated with radiation and in selected cases this was followed by chemotherapy. The majority of sltage II patients were treated with combined therapy. Of the 15 patients with lymphoma of the nose and sinuses, 80% survived and those with localized disease had 100% survival. Fierstein and Thawley suggest treatment with combined chemotherapy and radiation in all stages more advanced than Stage I.

Dramatic remissions occur in over 90% of patients with Burkitt: lymphoma following high dose alkylating agent therapy. Relapse occurs in approximately 2/3 of cases mostly in advanced stages (Batsakis 1981).

Treatment of rhabdomyosarcoma has evoluted from mere radical surgery with wide excision into a multimodal technique consisting of surgery, combined chemotherapy and radiotherapy. However a definitive management of adult cases has not yet been established (Suzuki et al 1984).

The Intergroup Rhabodomyosarcoma Study (IRS) has recommended 6 reigmens (A through E) for the treatment of the tumour based on its clinical group (Feldman 1982).

Regimen A: Following complete resection:

1- Vincristine (2 mg/m², intravenously weekly for 12 doses).

2- Actinomycin D (0.015 mg/kg/day, given I.V. for 5 days, with the course repeated at 12, 24, 36 and 48 weeks).

3- Cyclophosphamide (2.5 mg/Kg/day, given orally on day 42 and continuing up through 24 months.

Regimen B: As in Regimens A plus:

4- Postoperative radiation to the primary site.

Regimen C: Following surgery and postoperative radiation.

1- Vincristine (2 mg/m² I.V weekly for 6 weeks, 6 courses given over 48 weks).

2- Actinomycin D (0.015 mg/kg/day I.V. for 5 days, course repeated 5 times in 45 weeks).

Regimen D: as C plus:

3- Cyclophosphamide : as in Regimen A.

Regimen E ("Pulse VAC"):

1- Vincristine: as in Regimen A.

2-Actinomycin D: (course as in Regimen A and repeated 4 times in 54 weeks).

3- Cyclophosphamide: 10 mg/kg/day, I.v. for days 1 to 5, with the course repeated orally on days 84-90 then 2.5 mg/kg/day orally from day 140 to 24 months).

Regimen F: as in Regimen E plus:

4- Adriamycin (60 mg/m², given I.V. 5 times in 51 weeks).

These regimens are utilized as follows:

1- Patients in group I are treated according to regimen A or B.

2- Patients in group II are treated according to regimen C or D.

3- Patients in group III and IV are treated according to regimen E or F. In addition, all group III patients receive radiation (5000 - 6000 rads in 5-6 weks) to the primary site after 6 weeks of chemotherapy to avoid acute morbidity that can develop from the concomitant use of radiation and chemotherapy as regimens E and F are the most toxic. In group IV patients, radiation is also given to metstsis.

In the IRS, only 3% developed cervical metastsis. This suggests that radicl neck dissection is not indicated. The same was concluded by Schuller et al (1979). The recommended treatment by IRS of cervical nodes is therapeutic surgery and/or radiation of clinically positive nodes. Total excision of lesions in areas as the maxillary sinus where patients have a high risk of developing extensive local disease may be worthwhile. Intrathecal chemotherapy should be given for all lesions with known, suspected or likely CNS invasion (Suzuki et al 1984).

Results of treatment inIRS study:

1- Group I patients: treatment with Regimen A and B gave almost same resutls with only 8% developing recurrences.

2- Group II patients: treatment with Regimen C and D gave almost same results with 19-23% relapses.

3- Group III patients: treatment with Regimen E and F gave almost same results with 47-52% complete remission and 29-34% partial remissions. 36-44% of patients with complete remission, however, developed a relapse later.

4- Group IV patients: 43-50% of patients developed complete remissions while 31-40% developed partial response.

The IRS findings also tend to show the following:

1- Radiation therapy after local resection is not necessary.

2- The use of 3 drugs after 2 years is not more effective than the use of 2 drugs for 18 months.

3- Adriamycin does not seem to improve response or survival in "pulse VAC" therapy (Feldman 1982).

Malignant vascular tumours are treated as a rule by wide surgical excision. Radiotherapy is used as an adjuvant especially after partial excision; for inoperable cases and for distant metastasis and should not be less than 5000 rads in 5 weeks (De Campora et al 1983).

Metastatic tumours of the paranasal sinuses are treated by removal of metstasis together with the primary with good results. When metastases to the sinuses represent one of multiple areas of involvement, palliative treatment, in the form of irradiation, local arterial perfusion or chemo-therapy, may be used. The metstatic tumours of jaws need chiefly palliative

treatment, mainly by radiation therapy, with the intention of providing relief from pain and prolongation of life (batsakis 1981).

PALLIATION:

The final aim of treatment is the relief of human pain on suffering. This premise should remain a primary concern in the treatment of the dying patient.

General measures of palliation include the control of infections by drainage and antibiotics, the use of topical anaesthesia as viscous xylocaine as a gargle or swished about the mouth for reducing discomfort of mucositis, proper oral hygiene and adequate nutrition and hydration. Shoulder pain experienced by most patients following radical neck dissection with resection of the spinal accessory (11th) nerve, can be relieved by shoulder sling, infrared heat and muscular exercise.

Specific measures for pain relief include radiation, chemotherapy, analgesic drug therapy, surgical therapy and stimulation techniques.

Radiation therapy is useful for bony metastasis, especially to a vertebra, and ulcerative malignancies.

Chemotherapy may offer excellent pain relief and is frequently well tolerated. methotrexate, 25 to 50 mg/m², administered intravenously once weekly is usually employed with marked pain relief within 1 or 2 weeks.

Analgesic drug therapy should aim at the supply of ample strength and quantity of drugs to the patient with uncontrolled cancer, while, on the other hand substituting various non-addicting medications to the patient who is free of disease. Because of the problem of developing drug tolerance and the need for further stronger, more effective pain relief as the cancer progresses, the milder analgesics are used first. These include aspirin, acetaminophen, propoxyphene and ethoheptazaine. Tolerance may lead to the shift form one drug to the other. When mild drugs become ineffective, combinations of drugs are used such as acetaminophen and propoxyphene, propoxyphene and phenacetin; aspirin and cafeine. When these we should proceed to the intermediate strength analgesics which include codeine, usually in combination with acetaminophen or APC compound, and pentazocine. Combinations are more effective.

When these drugs fail, more complex treatment forms such as psychotropic drugs, behavioral modification, biofeedback, and/or autogenic relaxation techniques may be needed for use, combinations of anitriptyline, fluphenazine or chlorpromozine and one of the mild or moderate class of analgesics.

The class of strong analgesics include methadone, meperidine, and morphine and produce apathy, somnolence, decreased apetite and altered behaviour. The parenteral use of strong narcotics is the last stage of progression of drug use for pain control.

Surgical palliation is directed at the control of pain and bleeding, reduction of tumour bulk and odour as well as the maintenance of nutritional stability. Reduction of the tumour bulk can be carried out esily with repeated and freuent currettage or electro-coagulation and suction. Cryosurgery, as well, is uselful.

Often simple surgical procedures is tried first as they may give excellent relief of pain. Local blocks in the trigeminal nerve and its branches using alcohol injection can give pain relief for 6 months to one year. Neurectomies also may be useful when the tumour lies within the distribution of the avulsed nerve. Percutaneous radiofrequency rhizotomy of the trigeminal ganglion gives good results in tumours within the distribution of the nerve (Sweat and Wepsic 1974). Major surgical neuroabaltive procedures are considered when the patient has a life expectancy of 3 months or more when other procedures fail. Medullary tractotomy in which the descending root of the trigeminal nerve in the medulla is sectioned has given good results by some authors. Frontal leucotomy, cingulumotomy and thalamotomy are available only in few institutes. They may lead to major personality changes and apathy and have high morbidity and mortality rates.

Patient teaching about pain cause and duration may be helpful to alleviate anxiety. Many adjunctive techniques of pain control are employed with some success. These include behavioral and environmental modifications,

relaxation, biofeedback, hypnosis and acupuncture using needles at certain acupuncture points on the body to achieve anaesthesia.

Nursing nd nutrition:

Nursing including preoperative care, postoperative care, wound care, mouth care, communication, emotional consideration and care of irradiated ptients and patients treated with chemotherapeutics is of utmost importance as an adjuvant to treatment.

Liquid nutritional supplements, vitamins and iron should be given as necessary. The use of gastrostomy must be remembered for palliation although feeding problems are usually short lived occurring after major surgery and as a terminal event.

Nutrition may consist of parenteral therpy (intravenous hyperalimentation), enteral therapy or both. Parenteral nutrition aims at supplying sufficient calories and proteins by infusion through a catheter inserted into the subclavian veins.

Enteral therapy involves feeding through a nasogastric tube, feeding oesophagostomy and gastrostomy. Various types of diets and commercial products are available for tube-fed patients.

From the above discussion of treatment modalities and trials, one can conclude that all patients should not receive the same treatment. Treatment should be individualized to every case based on tumour extension, staging and condition of the patient.

The best results are obtained with preoperative iradiation, anterior chemotherapy and radical surgery.

Preoperative irradiation usually in the form of cobalt 60 iradiation, 5000 rads/25 fractions/5 week helps debulking of the tumour and protects against dissimination during surgical maneuvres. It is best applied with hyperbaric oxygen.

Anterior chemotherapy gives the best results with intra-arterial infusion of 5-FU to a total dose of 2000 mg during radiotherapy given through the superifical temporal artery in shots. Combination of vincristine, bleomycin and methotrexate gave good results but tend to increase the incidence of toxic effects.

Surgery is best carried out 4-6 weeks after the end of preoperative radiotherapy and anterior chemotherapy. Surgery should be radical. Extended radical surgery may be needed for extensive disease while craniofacial resection is indicated when the disease remains on the cribriform plate without intracranial extension. Preservation of the orbital contents with

removal of the orbital floor could be practiced in selected cases with involvement of periorbita, posterior ethmoid cells or orbital apex. Primary reconstruction is now becoming preferrable as it minimizes the postoperative disfigurement and functional disability without affecting the survival rate. Cryosurgery as a means of debulkation of the tumour mass is now becoming popular with the aim of minimizing the need for radical maxillectomy to give patients the chance to live with less cosmetic and functional disability.

Cervical node metastasis is treated by radical neck dissection, there is no reason for elective neck dissection. Supplementary treatment consists of early antrostomy for inspection and aspiration. Recurrences are treated with radical surgery and irradiation, the use of radio-active sutures and CO_2 laser is now performed by some authors with promising results.

Postoperative irradiation and chemotherapy may be given suspected or proven microscopic residuals.

It should be noticed that certain histological types respond better to specific modalities of treatment, e.g. lymphomas respond best to radiotherapy, Burkitt's lymphoma responds best to high dose alkylating agent therapy, rhabdomyosarcoma is best controlled with combinations of resection, posterior chemotherapy and postoperative radiation, while malignant vascular tumours are treated as a rule by wide surgical excision with adjuvant radiation.

SUMMARY

Malignant tumours of the maxilla are hidden within secluded bony cavities and tortuous recesses and may involve and invade many vital structures. These tumours are fortunately uncommon forming 80% of the paranasal sinuses cancer which themselves constitute 0.2-0.8% of all body malignancies and 3% of cancers involving the upper aerodigestive tract. A wide variety of factors have been implicated in the aetiology of these malignancies. Occupational agents including mesothorium, radium agents mustard gas, polycycliaromatic hydrocarbons, nickel, chromates, wood and leather dust are the commonest aetiological factors. Nutritional agents, such as nitrosamines and some food additives, radio-isotopes and smoking are also blamed for these malignancies. Recently immuno-suppression and oncogenic viruses including those responsible for AIDS are included in these factors. Chronic sinusitis and oroantral fistula are rare predisposing factors.

Maxillary carcinoma has an incidence of less than one case/200.000 people per annum with age incidence over 40 years in 95% of patients and sex ratio of 2:1 in favour of males. Squamous cell carcinoma comprises 80-90% of the pathological types. The next common histopathological types are undifferentiated carcinoma, adenoid cystic carcinoma, adenocarcinomas, lymphomas, plasmacytoma, osteosarcoma, malignant mixed

tumour, and rhabdomyosarcoma respectivley. Speed is mainly by direct infiltration of the surroundings aided by the intimacy and unity of the nasal passages and paranasal sinuses. Bone destruction occurs in 70-80% of cases while invasion of the orbit occurs in about 45% of cases. Cervical lymph node metastases and distant metastases are rare.

Metastatic tumours of the maxilla are rare constituting about 1% of malignancies of this area. These are most often arising from primaries in the kidneys, lungs and breast. Direct invasion of the maxilla occurs by direct spread from the surroundings in 7% of cases of cancer of the head and neck.

As yet, there is no generlly accepted classification for tumours of the nose and paranasal sinuses. The different proposed classification are not suitable for a universal adoption without modification.

The most acceptable classification is that proposed by the American Joint Committee (AJC) (1978) and by the Japanese Joint Committee (JJC) 1977.

Malignancy of the nasal chambers and paranasal sinuses often masquerades as a chronic inflammatory condition. Presentation is therefore, variable and depends upon the direction of spread.

Pain, nasal obstruction, nasal discharge, oral symptoms, dental symptoms, cheek swelling and ocular symptoms are the commonest presentations.

The advanced stage at diagnosis is responsible for the poor prognosis. Diagnosis is a difficult thing and is only obtained by maintaining a high degree of suspicion, which should proceed to a full examination and investigation of the patient. Investigations include radiographic imaging particularly computerized tomography which has improved very much the chances of early detection of soft tissue tumours and intrcranial extensions, antral lavage, exploratory antrostomy, endoscopy and biopsy. Exploratory surgery seems to be the only way of early detection of the tumours.

The approach to treatment of the maxillary cancer depends upon the certainity of clinical diagnosis. At the present time there is no standard treatment endorsed by all surgeons. In general, treatment plans should be individualized according to the histology, location, extent of the tumour and various patient factors. A variety of therrapeutic measures are available for the management of malignancies in the maxilla. These include surgical excision, radiation therapy, cryotherapy, laser excision, chemotherapy, immunotherapy and others.

Several trials utilizing different modalities and combinations have been reported. The present most acceptable treatment is preoperative

irradiation with cobalt 60 gamma rays to a total dose of 5000 rads/25 fractions/5 weeks, anterior chemotherapy with intra-arterial infusion of 5-FU for a total dose of 2000 mg during irradiation or a combination of vincristine, bleomycin and methotrexate, and radical surgery extended as necessary including orbital exenteration and craniofacil resection for tumours inserving the ethmoids. Without intracranial extension. Supplementary therapy consists of antrostomy for follow up and suction of necrotic tumour tissue.

There is a recent trend for preservation of orbital contents as this does not worsen prognosis but gives a better chance for a better patients cosmetic and functional ability.

Cryosurgery is recently suggested to be a substitute for radical maxillectomy as debulkation of the tumour as it has less incidence of disfigurement, maxillectomy being carried out for local recurrences and extensive tumours.

Postoperative irradiation and chemotherapy are suggested for local suspiclous and confirmed residuals.

CO_2 laser and radioactive sutures are under trials but seem to be promising.

Good nursing and nutritional care of patients preoperatively and postoperatively can participate a better prognosis.

In fact, further studies are needed for the development of more practical classifications, diagnostic procedures and standard modalities of treatment as those available one are far from satisfactory.

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الملخص العربى

تستتر الاورام الخبيثة لعظمة الفك العلوى داخل تجويفات عظمية مقفله ومتعرجة ويمكنها ان تشمل او تغزو تركيبات حيوية عديدة . ومن حسن الحظ ان هذه الاورام غير شائعة وهى تكون حوالى ٨٠٪ من جميع اورام الجيوب الانفية التى لا تتعدى نسبتها ٢ر - ٨ر٪ من الاورام الخبيثة التى تصيب كافة اجزاء الجسم البشرى وحوالى ٣٪ من امراض الجهاز الهضمى التنفسى وهناك عوامل عديدة تلعب دورا فى احداث هذه الاورام منها عوامل مهنية مثل الميزوثوريوم والراديوم وغاز المسطرد والهيدروكربونات العطرية متعددة الحلقات والنيكل والكرومات والغبار الناتج من الخشب والجلود وهذه العوامل من أهم مسببات هذا المرض . ومن ضمن مسببات هذه الاورام ايضا بعض العوامل المتصلة بالتغذية مثل امينات النتروز وبعض الاضافات التى تضاف الى الاطعمة المحفوظة وكذلك النظائر المشعة والتدخين .

وحديثا اكتشف دور مثبطات الجهاز المناعى وكذلك بعض الفيروسات فى احداث هذه الاورام الخبيثة ومن ضمن هذه الفيروسات تلك التى تسبب مرض (الايدز) وهو مرض ضعف الجهاز المناعى المكتسب وكذلك يعتبر الالتهاب المزمن بالجيوب الانفية من العوامل المؤديه لهذا المرض .

ولا تتعدى نسبة حدوث اورام الفك العلوى حالة واحدة من بين كل مائتى الف نسمة فى العام الواحد وفى ٩٥٪ من الحالات يكون عمر المريض اكثر من ٤٥ عاما ونسبة الاصابة بين الذكور ضعف نسبتها بين الاناث .

وقد وجد ان الاورام الناتجة عن الخلايا البشرية تكون بين ٨٠و٩٠فى المائة من النوعيات الباثولوجية ويلى ذلك الاورام غير مميزة الاصل

والأورام الغدية والأورام الليمفاوية والبلازماسيتوما وأورام العظام الخبيثة والورم الخبيث المختلط واورام العضلات الخبيثة بالترتيب المذكور . وتنتشر هذه الأورام فى معظم الحالات عن طريق الامتداد المباشر الى الانسجة المحيطة ويساعدها فى ذلك التقارب والتوحد بين الجيوب الانفية وممرات الانف المختلفة ، ويؤدى هذا الانتشار الى تدمير العظام فى ٧٠ - ٨٠٪ من الحالات بينما يمتد الورم الى العين فى٤٥٪ فقط من الحالات اما الامتداد الى العقد الليمفاوية بالرقبة أو الى أماكن بعيدة فهو شئ نادر الحدوث .

أما الاورام الخبيثة التى تصيب عظمة الفك العلوى ثانويا نتيجة للامتداد من اماكن أخرى بالجسم فهى نادرة جدا ولا تتعدى نسبتها ١٪ من مجموع الاورام الخبيثة بهذا الموضع وتأتى هذه الاورام الثانوية فى الغالب من الكلى والرئتين والثدى بينما الاورام التىتصيب هذه العظمة كنتيجةللامتداد المباشر اليها من الانسجة المحيطة بها فتكون حوالى ٧٪ من جميع حالات سرطان الرأس والرقبة .

وحتى الان لا يوجد تقسيم عام مقبول للاورام التى تصيب الانف والجيوب الانفية اذ أن كل التقسيمات المقترحة من بعض الاطباء لا يصلح أى منها لاعتماده كتقسيم شامل بدون الحاجة الى ادخال تعديلات عليه الا أن اكثر هذه التقسيمات قبولا هو ذلك الذى اقترحته اللجنة الامريكية المشتركة فى عام ١٩٧٨ وكذلك ذلك الذى اقترحته اللجنة اليابانية المشتركة فى عام ١٩٧٧ .

ومن الشائع ان تختفى الاورام الخبيثة التى تصيب الانف والجيوب الانفية وراء صورة التهاب مزمن ولذلك فان الشكوى التى يأتى بها المريض تتعدى صورها كثيراوتعتمد على الاتجاه الذى يتمدد اليه الورم ، ومن

اكثر الاعراض شيوعا الألم وانسداد الأنف والافرازات الأنفية والأعراض التى تنشأ من الامتداد للفم والاسنان وكذلك تورم الخد والاعراض الناشئة عن امتداد الورم للعين . هذا وتعتبر الحالة المتأخرة التى يكون عليها الورم وقت التشخيص من أهم أسباب تردى نسبة الشفاء منه اذ ان تشخيص الورم فى هذا الموضع يعتبر مهمة صعبه ويمكن التوصل اليه فقط بالاعتماد على درجة عالية من التشكك فى الاعراض ومن ثم اجراء كافة الفحوص اللازمة بما فيها التصوير بالاشعة وخصوصا الاشعة المقطعية التى أدت الى تطور هائل فى امكانيات التشخيص المبكر للاورام التى تصيب الانسجة الرخوة والامتداد الى داخل الجمجمة ، كذلك من هذه الفحوص غسل الجيوب الانفية والعمليات الاستكشافية والمناظير وأخذ العينات من انسجة الورم وتحليلها ، ولكن فى الواقع فان الجراحة الاستكشافية تعتبر الوسيلة الوحيدة للتشخيص المبكر لهذه الاورام .

ويعتمد اسلوب علاج سرطان عظمة الفك العلوى على درجة من التأكد من التشخيص وفى الوقت الحالى لا يوجد أسلوب علاجى موحد متفق عليه بين كافة المعالجين الا أنه على وجه العموم فأن خطة العلاج يجب ان توضع حسب معطيات كل حالة على حده سواء من ناحية نوعية التشريح النسيجى للورم أو موضعه بالتحديد او امتداداته وكذلك بعض العوامل الشخصية الخاصة بالمريض وفى الواقع فهناك عدة وسائل متاحة لعلاج الاورام الخبيثة لعظمة الفك العلوى وتتضمن العلاج الجراحى والعلاج الاشعاعى والعلاج بالتبريد وبأشعة الليزر والعلاج الكيماوى والعلاج المناعى وطرق اخرى .

وقد جرت عدة محاولات علاجية أستخدمت فيها وسائل مختلفة للعلاج وكذلك استخدمت فيها وسائل مجتمعة اختلفت من محاولة لأخرى ولكن الأسلوب الأقرب الى القبول فى الوقت الراهن يتكون من علاج اشعاعى

بواسطة اشعاعات جاما الصادرة من كوبالت ٦٠ المشع وبجرعة تتراوح بين ٥٠٠٠ – ٦٠٠٠ راد مقسمة على خمس وعشرين جلسة تتم فى خلال خمسة أسابيع بمعدل خمسة ايام كل اسبوع وفى نفس الوقت يعطى علاج كيماوى بالعقاقير عن طريق اعطاء محلول وريدى يحتوى على ٥ – فلوروبوراسيل بجرعة كلية ٢٠٠٠ مجم تتخلل العلاج الاشعاعى أو عن طريق اعطاء مجموعة عقاقير تتكون من فينكريستين وبليومايسين وميثوتركسات ثم يلى ذلك كله استئصال جراحى شامل يتحدد مداه حسب الحاجة ويمكن أن يتضمن استئصال العين وتجويفها واستئصال جزء من عظمة الجبهة والرأس فى حالة امتداد الورم ليشمل هذه الاماكن . ويشتمل العلاج التكميلى على عملية استكشاف للجيوب الانفية لمتابعة الحالة ولازالة الأنسجة الميتة المتخلفة عن الجراحة .

ويوجد فى الوقت الحالى اتجاه يميل الى الابقاء على العين ومكوناتها حيث انه قد وجد ان عدم استئصالها لا يؤدى الى تدهور نسبة الشفاء أو تقصير فترة الحياة المتوقعة بالاضافة الى ان الابقاء على العين يعطى فرصة افضل للمريض للابقاء على الناحية الوظيفية والشكلية للعين .

وقد أدخلت حديثا الجراحة بواسطة التبريد حتى التجمد كبديل للاستئصال الجراحى الموسع كوسيلة لتصغير حجم الورم حيث ان ذلك يفيد فى تقليل التشوه الناتج عن الجراحة ومع قصر الجراحة على الحالات التى ينتشر فيها الورم بدرجة كبيرة أو الحالات التى ينمو فيها الورم مرة اخرى بعد الاستئصال . وقد اقترح البعض اعطاء علاج اشعاعى وكيماوى بعد الاستئصال الجراحى للحالات التى يشك فى وجود بعض البقايا منها .

وقد جرى حديثا أيضا ادخال العلاج بواسطة أشعة الليزر والخيوط

الجراحية المشعة وقد أعطت حتى الان نتائج مشجعة ٠

ومن الأهمية بمكان ذكر أن التمريض والتغذية السليمة للمريض قبل وبعد الجراحة يمكن ان يسهما الى حد كبير فى تحسين نتائج العلاج ٠

وفى الحقيقة فان الحاجة مازالت ملحة للمزيد من الدراسات لاستنباط تقسيمات جديدة ووسائل تشخيصية جديدة ووسائل علاجية موحدة ومتفق عليها حيث أن المتاح حاليا مازال غير مرضى٠

الاورام الخبيثة بعظمة الفك العلـــــوى

رسالة مقدمة مـــن

الطبيب / عادل محمد ابو النجا سرحـــان

توطئة للحصـــول علـــى

درجة الماجستير فى جراحة الانف والاذن والحنجرة

تحت اشراف

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كلية الطـب ـ جامعة الازهر

١٩٨٦