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RADIOLOGY OF SKULL BASE NEOPLASMS

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CT scanning and MR imaging are used routinely to stage skull base neoplasms preoperatively, define extent of tumor, identify perineural spread, plan surgery and radiotherapy, and evaluate the postoperative patient. Arteriography and interventional radiology also have a role in the identification and treatment of certain skull base neoplasms, particularly juvenile nasopharyngeal angiofibromas and paragangliomas. The skull base can be divided into three components: (1) anterior, (2) central, and (3) posterior. Pathology differs in each region, and CT scanning and MR imaging often demonstrate characteristic imaging features suggestive of the diagnosis. These imaging features are highlighted.

The emergence of skull base surgery in recent history has improved dramatically the prognosis and outcome of patients with skull base pathology. This has been made possible by technologic advances and the adoption of a team approach combining the surgical expertise of otolaryngologists and neurosurgeons with the diagnostic skills of neuroradiologists and pathologists. For many years, the standard treatment of the patient with skull base pathology was surgical exploration to determine the pathology and extent of tumor spread. This approach carried inherent risks of morbidity and mortality, including cranial nerve or vascular injury, cerebrospinal fluid (CSF) fistula, and infection. The nature of the extensive surgical procedures required for staging often prevented these patients from definitive therapy, because the surgical risks outweighed the benefits and were not considered warranted in many cases.[4] Advances in technology combined with the adoption of a team approach between surgical specialists and consultants have allowed a more aggressive approach to lesions that previously were considered unresectable or untreatable, thereby improving patient outcome.

Advances in neuroradiology have contributed to the development of skull base surgery by helping to stage neoplasms preoperatively, define extent of tumor spread, identify imaging features characteristic and suggestive of certain pathologic entities, define a surgical approach, and evaluate for tumor recurrence postoperatively. The development of two modalities, CT scanning and

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MR imaging, has made this possible. These imaging modalities are now widely accessible, even in smaller community hospitals, and are the primary modalities used to evaluate the skull base. Arteriography is a useful adjunct in the workup of vascular lesions, often helpful in diagnosing paragangliomas and angiofibromas. Interventional radiology also has a role in treating vascular lesions. A balloon test occlusion of the internal carotid artery (ICA) may be performed preoperatively if sacrifice of the vessel is anticipated, and intravascular embolization with particles helps to minimize blood loss at the time of surgery.

This article discusses the state-of-the-art imaging techniques and their applications in imaging neoplastic processes of the skull base. A brief anatomic review is followed by a discussion of the characteristic CT scanning and MR imaging features of skull base tumors. The roles of angiography and interventional radiology as they apply to skull base pathology are discussed and illustrated. Radiologic evaluation of the postoperative patient also is discussed, emphasizing the role of positron emission tomography (PET) imaging in differentiating between post-treatment changes and residual or recurrent tumors.

IMAGING TECHNIQUES

The skull base is an undulating surface of limited craniocaudal dimension oriented in the axial plane. For this reason, CT scans of the skull base should include coronal images to evaluate for transgression of the skull base by a lesion. Because of the thinness of the skull base in the craniocaudal direction, slice thickness should be kept to a minimum. Contiguous 3-mm thick slices in the coronal and axial planes are usually sufficient, but thinner collimation (1 mm) is possible if necessary (often used for preoperative planning for endoscopic sinus surgery). Bone and soft tissue algorithms are used with wide window settings (>2000 HU) for bone images and narrow settings (200–400 HU) for soft tissue images. Intravenous contrast is indicated for neoplastic processes in the skull base because it increases the sensitivity of detecting pathology. It is administered by power injection with delayed imaging to maximize intravascular enhancement. Precontrast imaging is usually not necessary, except perhaps in the setting of fungal sinusitis, which is intrinsically dense.

CT scanning is well suited in defining bony anatomy of the skull base because of its high-resolution bone algorithms and is superior to MR imaging in determining bone involvement and destruction. One of the imaging criteria provided by CT scanning that may be suggestive of the type of pathologic condition is the effect of the mass on the adjacent bone.[21] Although there are exceptions to the rule, the reaction of bone to the disease process is often a clue to the pathology. For example, squamous cell carcinoma, the most common of the sinonasal neoplasms, characteristically destroys bone aggressively. This entity and its infections may have features of aggressivity, which include bony destruction, and engulfment of the bone within the mass. Conversely, lesions that cause expansion of the paranasal sinuses and smooth remodeling of adjacent bone are more likely to be benign entities such as mucoceles or sinonasal polyposis. It should be emphasized that there is no absolute correlation between the biologic aggressiveness of the lesion and bone response. For example, sinonasal melanoma tends to have a benign imaging appearance even though its 5-year survival rate is only 5% compared with the 25% survival rate for squamous cell carcinoma, which is associated with bony destruction.[21] A combination of bony destruction and remodeling is associated with chronic, relapsing processes such as aggressive chronic infection or

fungal sinusitis. These lesions usually are associated with reactive sclerosis and bony thickening.

In general, MR imaging is the primary imaging modality for any skull base process that is suspected to involve the brain or leptomeninges or may interface between the brain and extracranial head or neck structures.[16] In comparison with CT scanning, MR imaging easily can image the skull base in any plane and is more sensitive in detecting leptomeningeal, dural, or cranial nerve involvement, marrow space processes, and intracranial spread. It also is better suited to differentiating retained secretions from tumor in processes involving the paranasal sinuses and mastoid air cells. Pulse sequences, such as fat saturation techniques, are useful in suppressing high signal from fat, maximizing tissue–tumor contrast. This is particularly useful in distinguishing bone marrow or orbital fat from tumor. The intravenous administration of gadolinium helps to maximize the contrast between normal tissue and pathology and is used almost universally in skull base studies. Multiplanar, post-gadolinium T1-weighted images with a small field of view and fat saturation are used routinely in skull base imaging.

Virtually all arteriography in ENT-related head and neck disorders is preceded by CT scanning or MR imaging. The main indication for diagnostic arteriography is to characterize and confirm the diagnosis of a mass lesion seen on other imaging studies.[16] Paragangliomas and juvenile nasopharyngeal angiofibromas are hypervascular lesions that have a near pathognomonic appearance on arteriography (Fig. 1). The diagnosis of these lesions usually can be made from the arteriogram, obviating the need for biopsy. The other indication for diagnostic arteriography is evaluation of definitive endovascular therapy or endovascular

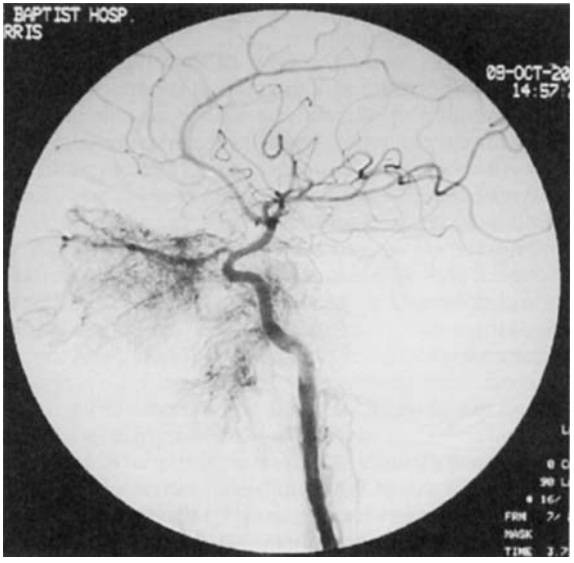

Figure 1. Juvenile nasopharyngeal angiofibroma. A lateral projection from a cerebral arteriogram reveals a large hypervascular mass in the nasal cavity and nasopharynx in this patient with recurrent epistaxis.

therapy as an adjunct to definitive surgical management of a lesion. Preoperative embolization may be invoked to minimize intraoperative bleeding during the resection of such vascular lesions as juvenile nasopharyngeal angiofibromas, paragangliomas, or skull base meningiomas.

PET is another adjunct that has gained popularity, and it is particularly useful in postoperative patients in differentiating between residual or recurrent tumor versus post-treatment changes which may have an identical appearance on CT scanning or MR imaging. Fluorodeoxyglucose (FDG) PET is the most commonly used agent and is taken up by metabolically active tumors that use glucose as a substrate. PET is a functional examination (as opposed to an anatomic examination) that takes advantage of the fact that metabolically active tumor and adenopathy demonstrate FDG uptake, whereas post-treatment scar or granulation tissue does not.[15] Current limitations of PET include limited sensitivity in detecting small lesions, but this issue may be addressed in the near future with the advent of newer imaging agents.

SKULL BASE ANATOMY

The important aspect of the normal anatomy of the skull base is found in the skull base apertures and their contents.[12] The skull base is composed of five bones: (1) the ethmoid, (2) sphenoid, (3) occipital, (4) paired temporal, and (5) paired frontal bones. The skull base can be divided into three compartments: (1) anterior, (2) central, and (3) posterior. This breakdown is convenient because surgical approaches and pathologic entities vary in each compartment. Craniofacial approaches are used in resections of the anterior skull base, and lateral or transpetrous approaches are used for lesions of the central or posterior skull base. Occasionally, a transphenoidal approach may be used for sphenoid or sellar lesions such as pituitary adenomas.

The anterior skull base forms the floor of the anterior cranial fossa and includes contents of the orbits, ethmoid air cells, nasal cavity, and frontal sinuses. It is composed of the orbital plates of the frontal bones, cribriform plate of the ethmoid bone, the planum sphenoidale, and paired lesser wings of the sphenoid bone.[14]

The central skull base is perhaps the most complex and clinically important portion of the skull base because cranial nerves and vessels (the internal carotid artery and its collaterals) traverse the skull base here. The body of the sphenoid bone forms the crux of the central skull base. It is located medially and contains the sella turcica and sphenoid sinus. Its two greater wings spread out laterally forming the floor of the middle cranial fossa. The smaller lesser wings of the sphenoid bone form the anterior clinoid processes. Inferiorly, the central skull base includes the medial and lateral pterygoid processes and the pterygopalatine fossae. Numerous foramina are situated along the plane and intersecting the sphenoid body and greater wings.[3] They include the following paired structures: foramina rotundum, ovale, spinosum, vidian canal, superior orbital fissure, optic canal, and carotid canal. These apertures of the skull base are conduits for cranial nerves and blood vessels. The foramina are also potential pathways for perineural spread of head and neck neoplasms into the cranium.

The posterior skull base forms the floor or "bowl" of the posterior fossa and is composed of the clivus anteriorly, the petrous and mastoid portions of the temporal bone anterolaterally, and the occipital bone laterally and posteriorly. The clivus receives contribution from the sphenoid and occipital bones, separated by the spheno-occipital synchondrosis. The clivus is separated from the

petrous apex of the temporal bone by a cartilaginous synchondrosis, the petro-occipital (petroclival) fissure. Several important foramina of the posterior skull base include the foramen magnum, internal auditory canals, hypoglossal canals, and jugular foramina.[13] The facial (cranial nerve VII) and vestibulocochlear (cranial nerve VIII) nerves exit the cranium through the internal auditory canals, and cranial nerve XII exits the posterior fossa through the anterior and obliquely oriented paired hypoglossal canals. The paired jugular foramina are actually focal widening of the posterior petro-occipital fissure consisting of two parts: (1) the anteriorly located pars nervosa, which transmits the glossopharyngeal nerve (cranial nerve IX), and (2) the pars vascularis, transmitting the jugular vein, and the vagus and spinal accessory nerves (cranial nerves X and XI, respectively).

SKULL BASE PATHOLOGY

Lesions of the anterior skull base can be divided into four categories: (1) aggressive sinonasal diseases that extend intracranially (which are the most common lesions), (2) lesions arising from the bone of the skull base, (3) uncommon intracranial lesions that extend inferiorly into the paranasal sinuses and nasal cavity, and (4) developmental lesions.[21] Lesions arising from the central skull base tend to be from the following categories: sellar and suprasellar lesions extending inferiorly into the skull base, sphenoid sinus lesions, lesions arising from the sphenoid bone, and lesions arising from the dura overlying the planum sphenoidale and clivus. Posterior skull base lesions include lesions that arise from the clivus, petro-occipital fissure, and jugular foramen.

Anterior Skull Base Lesions

Sinonasal lesions comprise the majority of anterior skull base lesions. Neoplastic lesions of sinonasal origin may be divided into tumors of epithelial and nonepithelial origins. The former includes squamous cell carcinoma and the inverted papilloma. Nonepithelial neoplasms include minor salivary gland tumors, melanoma, olfactory neuroblastoma, rhabdomyosarcoma, large cell lymphoma, extramedullary plasmacytoma, and metastases from renal, breast, and lung carcinomas.[21] These lesions can extend intracranially by direct extension through the cribriform plate and floor of the anterior cranial fossa.

Squamous cell carcinomas account for 80% of the malignancies affecting the paranasal sinuses, and most of these tumors occur in the maxillary antrum.[11] It is an aggressive process that often demonstrates bony destruction on CT scan and intense homogeneous enhancement on postcontrast imaging, which helps to distinguish this entity in the majority of cases from benign inflammatory processes, such as sinonasal polyposis, and mucoceles, which do not erode bone and enhance peripherally. MR imaging is useful in differentiating tumor from post-obstructive secretions. Because of its aggressive nature, extension through the cribriform plate into the anterior cranial fossa is possible. MR imaging is better suited in evaluating intracranial extension. It should be noted that meningeal enhancement in itself does not necessarily indicate neoplastic invasion, because reactive dural enhancement may occur as a result of fibrovascular changes. The following criteria are suggestive of dural invasion: (1) nodular, irregular enhancement, (2) greater-than-5-mm thickness of the dura, (3) pial enhancement, and (4) parenchymal enhancement or vasogenic brain edema.[7]

Minor salivary gland tumors and melanoma are the next most common malignancies to involve the sinonasal cavity after squamous cell carcinoma. The minor salivary gland tumors account for approximately 10% of sinonasal neoplasms and show a wide variety of histologic types, the most common of which is adenoid cystic carcinoma. CT scanning may show bony destruction, and postcontrast MR imaging is of particular use in identifying perineural spread of malignancy.

Sinonasal melanomas arise from melanocytes that have migrated from the neural crest to the sinonasal mucosa during embryologic development.[1] They account for less than 2.5% of all melanomas and are more common in the nasal cavity than the paranasal sinuses, often arising from the nasal septum or turbinates. Melanomas tend to remodel bone, although elements of frank bone erosion may be present. Because of their rich vascularity, there is robust enhancement on postcontrast CT scanning and MR imaging. Another characteristic and helpful finding is the T1-weighted high-signal intensity associated with melanoma resulting from one of two factors: (1) the paramagnetic effects of melanin (70% of cases; the other 30% of cases are amelanotic), or (2) intratumoral hemorrhage.[22] The relatively benign appearance suggested by the paucity of bone destruction is deceptive. In contrast, the prognosis is poor with a median survival time of 24 months.

Olfactory neuroblastoma (esthesioneuroblastoma) is a rare tumor of neural crest origin arising from the olfactory mucosa in the superior nasal vault. There is a bimodal incidence with peaks of occurrence between the second and sixth decades of life. Patients present with nasal obstruction, epistaxis, and a decrease in olfactory function. These lesions have a propensity for crossing the cribriform plate where they may invade the brain and seed CSF spaces. Cases that cross the cribriform plate necessitate a craniofacial approach so that the tumor can be resected en bloc.

The role of imaging in these patients is to map the tumor to anticipate surgical boundaries. On CT scans, these lesions appear as solid, homogeneously enhancing superior nasal cavity masses with or without bony erosion or destruction of the cribriform plate. Calcifications may occur in the tumor mass.[19] On MR imaging, these tumors have an intermediate signal intensity, and the T2-weighted signal intensity is higher.[6] In some of the larger tumors in which there is intracranial extension, peripheral tumor cysts can occur at the margins of the intracranial mass and, when present, are suggestive of the diagnosis (Fig. 2).[23]

Rhabdomyosarcoma is the predominant tumor involving the head and neck in the pediatric population, and intracranial extension is a common finding (Fig. 3). MR imaging shows a soft tissue mass in the sinonasal region, infratemporal fossa, or nasopharynx, often with bone destruction. On CT scans, they enhance mildly but demonstrate avid enhancement on MR postcontrast, T1-weighted images.[24] Lymphoma also occurs in the sinonasal region, usually appearing as bulky soft tissue masses that enhance to a moderate degree (Fig. 4).

Juvenile angiofibromas are benign, vascular, nonencapsulated nasopharyngeal tumors arising almost exclusively in adolescent males. They arise from primitive vascular tissue at the sphenopalatine foramen, and despite their benign histology, tend to be very aggressive locally with a propensity to insinuate themselves into and through foramina and fissures. Although they do not frankly invade bone, they do erode and remodel bone.[2] They also have a tendency to recur if not completely resected. Their signs and symptoms are related to extension of the tumor into the nose, paranasal sinuses, orbit, and skull base. These patients generally present with nasal obstruction and epistaxis. Other clinical findings include nasal discharge, facial deformity, cranial nerve palsies, exophthalmos, and anosmia.[5] CT scanning shows an enhanced nasal or nasopharyngeal mass, which may extend into the pterygopalatine fossa (Fig. 5). MR imaging is useful in

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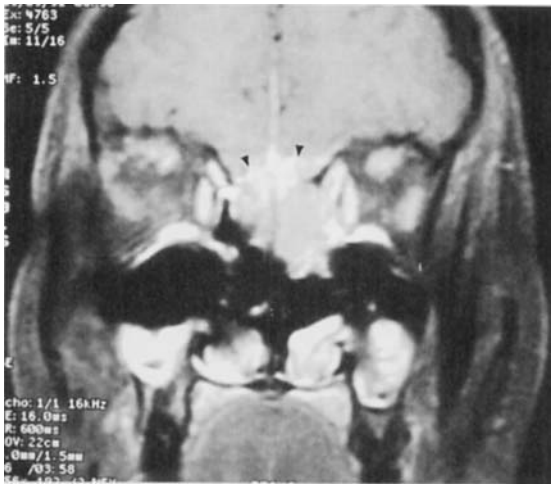

Figure 2. Esthesioneuroblastoma. An intensely enhanced sinonasal mass crosses the cribriform plate with irregular adjacent dural enhancement (*arrowheads*). A noncontrast CT scan shows destruction of the cribriform plate, and MR imaging of the brain and cervical spine reveals intracranial and spinal metastases from CSF seeding.

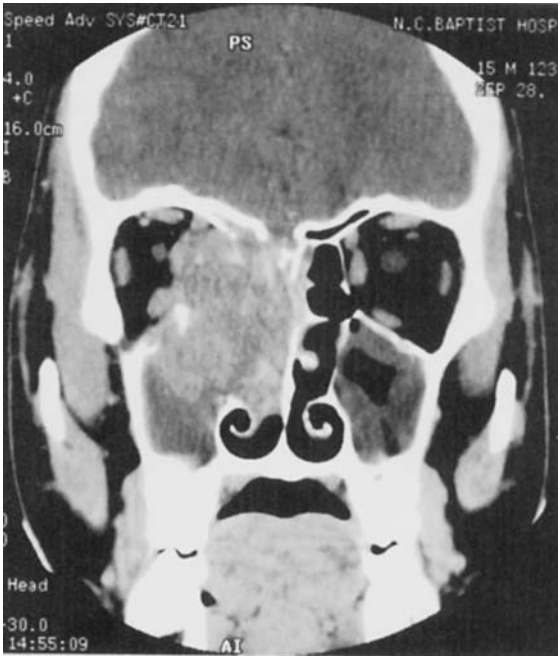

Figure 3. Rhabdomyosarcoma. This heterogeneously enhanced sinonasal mass has aggressive features, including bone destruction of the right lamina papyracae, cribriform plate, and medial wall of the maxillary sinus, and extension into the right orbit. Rhabdomyosarcoma is one of the most common head and neck malignancies in the pediatric population.

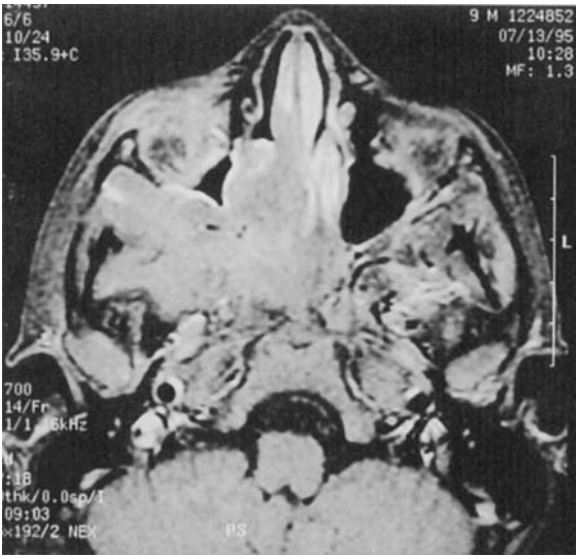

Figure 4. Lymphoma. A postgadolinium T1-weighted MR image with fat suppression reveals a homogeneous, mildly enhanced mass in the nasal cavity with extension into the right masticator space. Angiography shows a hypovascular mass. Biopsy proved Burkitt's lymphoma.

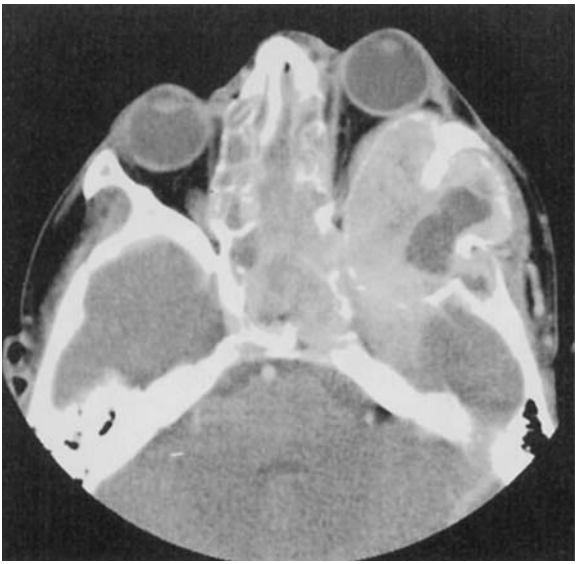

Figure 5. Juvenile nasopharyngeal angiofibroma. Axial postcontrast CT scan reveals a large, heterogeneously enhanced sinonasal mass with extension into the middle cranial fossa and left orbit, causing severe proptosis.

patients in whom intracranial extension is suspected. Features in MR imaging include flow-related voids in the tumor related to high vascular flow. MR imaging also helps differentiate frank sinus invasion from sinonasal obstruction and retention of secretions.

Fibrous dysplasia is one example of a fibro-osseous lesion, which is intrinsic to the skull base. Fibrous dysplasia arises from bone and widens the medullary cavity, leaving the cortex intact (Fig. 6). Chondrosarcomas are rare lesions of the anterior skull base, but when encountered usually arise from the nasal septum or junctional regions of the facial bones. Other examples of fibro-osseous lesions include ossifying fibroma, osteoblastoma, and Paget's disease.

Meningiomas are an example of an intracranial lesion that may extend extracranially. They arise from the intracranial dural lining of the anterior cranial fossa and in rare instances may extend extracranially.

The developmental lesions of the anterior skull base include the cystic nasal glioma and cephalocele. Nasal gliomas are rare, and cephaloceles in this portion of the skull base are frontoethmoidal in nature and are classified in three subsets: (1) nasofrontal, (2) nasoethmoidal, and (3) naso-orbital. MR imaging is the best modality in characterizing cephaloceles.

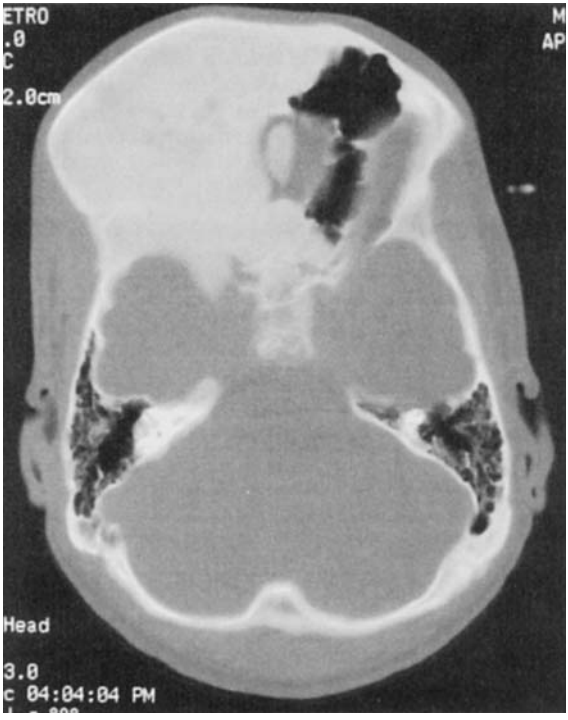

Figure 6. Fibrous dysplasia. The expansive, *ground-glass* appearance of the right anterior skull base, involving the frontal and sphenoid bones, is characteristic of fibrous dysplasia. Note the preservation of the cortical margins, a characteristic feature of fibrous dysplasia.

Lesions Involving the Clivus and Petro-Occipital Fissure

The majority of lesions involving the clivus and petro-occipital fissure arise from the bone marrow space and include chordomas, chondrosarcomas, plasmacytomas, non-Hodgkin's lymphoma, and metastases. Other lesions including pituitary macroadenomas and craniopharyngiomas originate from the sella or suprasellar region and extend inferiorly into the clivus. Meningiomas arise from the dura overlying the posterior clivus and may spread by direct extension into the posterior aspect of the clivus.[13]

Chordomas are malignant neoplasms arising from the remnants of the embryonic notochord. More than one-third of these lesions arise from the skull base, whereas the rest involve the sacrum and spine. Presenting symptoms of skull base chordomas include orbitofrontal headaches, visual disturbances, and ophthalmoplegia caused by the mass effect on the optic and cranial nerves passing through the cavernous sinus. Cranial neuropathies of the trigeminal, facial, and vestibulocochlear nerves also may be present from extension into the cerebellopontine angle and internal auditory canal. These slow-growing lesions are difficult to resect completely because of their proximity to the cranial nerves and ICAs. Unfortunately, they are also resistant to radiotherapy. Histologically, the tumor has chords or rests of vacuolated cells known as physaliphorous cells, which contain mucin and glycerin. On CT scan, the chordoma shows bony destruction of the clivus without sclerosis, separating tumor from adjacent bone. Calcifications frequently are present and may be large. The MR image appearance is somewhat variable, hypointense, or isointense on T1-weighted MR images and sometimes contains cystic areas of hemorrhage or mucoid material (Fig. 7). The lesions appear bright on T2-weighted MR images.[4]

Chondrosarcoma is another lesion with a predilection for the skull base, which is not surprising because the skull base is derived from cartilaginous

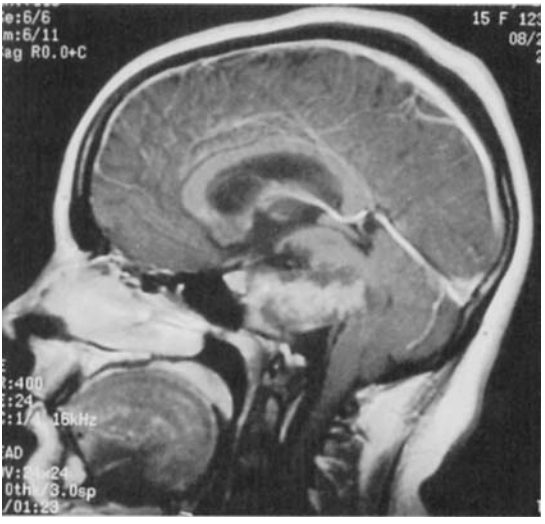

Figure 7. Chordoma. A large, heterogeneously enhanced mass involves the clivus, extending in exophytic fashion into the posterior fossa with significant mass effect on the pons and posterior fossa structures. This 15-year-old patient presented with headaches and cranial neuropathy.

elements or enchondral bone.[13] Most chondrosarcomas originate from cartilaginous elements residing in the petro-occipital fissure or at the junction of the nasal septum vomer with the sphenoid or ethmoid sinuses or along the undersurface of the basisphenoid (Fig. 8). Like chordomas, these lesions rarely metastasize, and the common presenting symptoms include cranial neuropathy, which most commonly involves the sixth cranial nerve. Prognosis depends on histopathologic grade, site, and extent of involvement. Petro-occipital chondrosarcomas are typically low-grade lesions with a much better prognosis than chordomas. Noncontrast CT scanning frequently is helpful in identifying calcified ringlets or arcs that are characteristic of chondroid matrix, which, if present, help to suggest the diagnosis of chondrosarcoma. Unfortunately, the degree of calcification is variable, and some chondrosarcomas are noncalcified soft tissue masses with bony destruction of the clivus. On MR imaging, these tumors have intermediate T1-weighted MR images and high-intensity T2-weighted MR images. Location may help differentiate chondrosarcoma from chordoma, the former originating lateral to the clivus and the latter in the midline. This sign is limited in cases of large lesions.

Plasmacytomas are rare tumors that are usually found in bone marrow resulting from neoplastic proliferation of a single clone of plasma cells with monoclonal immunoglobulin production. At the time of diagnosis, the patient usually already has multiple myeloma or will develop it soon. Neoplastic involvement of the skull base and calvarium in patients with multiple myeloma is common. On CT scan, the plasmacytoma appears as a nonspecific destructive skull base lesion. MR imaging shows intermediate intensity on T1- and T2-weighted MR images and diffuse enhancement on postgadolinium T1-weighted MR images (Fig. 9).

The skull base is frequently a site of bony metastases, most frequently involving renal, breast, and lung carcinomas (Fig. 10). Malignancies involving the clivus

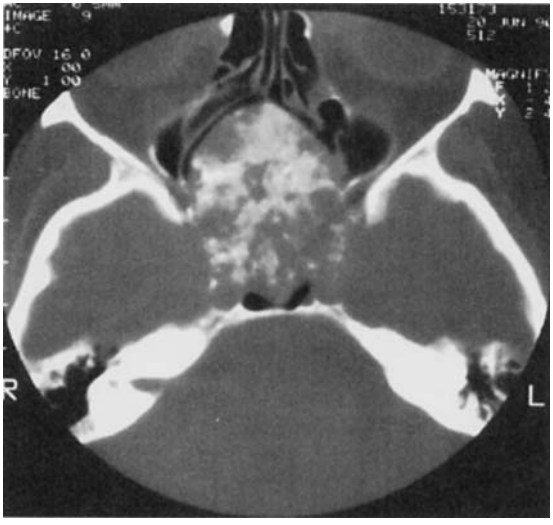

Figure 8. Chondrosarcoma. This patient with a sinonasal mass filling the posterior ethmoid air cells presented with nasal stuffiness. Noncontrast CT scan shows the characteristic *arcs and rings* of chondroid matrix suggesting the diagnosis of chondrosarcoma.

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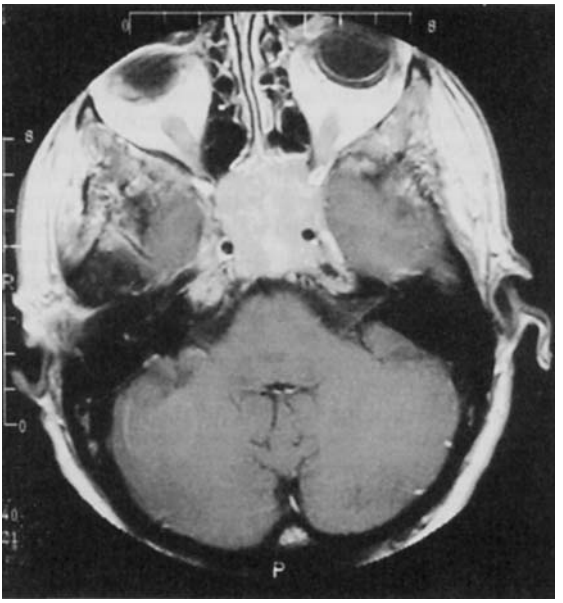

Figure 9. Plasmacytoma. A large enhanced mass on this postgadolinium T1-weighted MR image expands the clivus and represents a plasmacytoma. Many of these patients will eventually develop multiple myeloma.

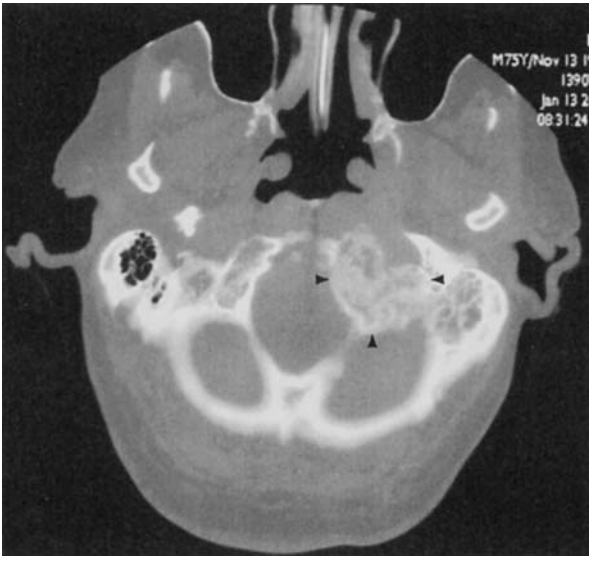

Figure 10. Metastases. A sclerotic, destructive mass (*arrowheads*) involves the left occipital condyle and hypoglossal canal in this patient with metastatic colon carcinoma. The patient had left hypoglossal paresis and denervation atrophy of the left tongue.

in the pediatric population include Ewing's sarcoma, osteosarcoma, lymphoma, and rhabdomyosarcoma.

Intracranial lesions involving the clivus include the pituitary macroadenoma and clival meningioma. Large pituitary macroadenomas may erode the floor of the sella and extend into the clivus. There may be mass effect on the optic chiasm superiorly, causing headache and visual disturbances. Clival meningiomas are uncommon lesions (most commonly parafalcine or overlying the convexities) that usually manifest as part of a larger lesion involving the sphenoid bone.[13] CT scans may show hyperostosis of the adjacent bone (Fig. 11). Meningiomas are isointense on T1-weighted MR images and enhance vividly after gadolinium administration. A "dural tail" enhancement pattern away from the central tumor bulk is frequently present.

Lesions Involving the Jugular Foramen

The jugular foramen is located along the posterolateral aspect of the petro-occipital fissure and is host to a unique differential diagnosis of lesions. The most common lesions include paragangliomas, nerve sheath tumors, and meningiomas.

The paraganglioma, or glomus tumor, is the most common lesion of the jugular foramen. Paragangliomas are vascular neoplasms that arise from the neural crest-derived chemoreceptor cells of the autonomic nervous system in the head and neck. Nomenclature of these lesions reflects the region of origin (e.g., lesions

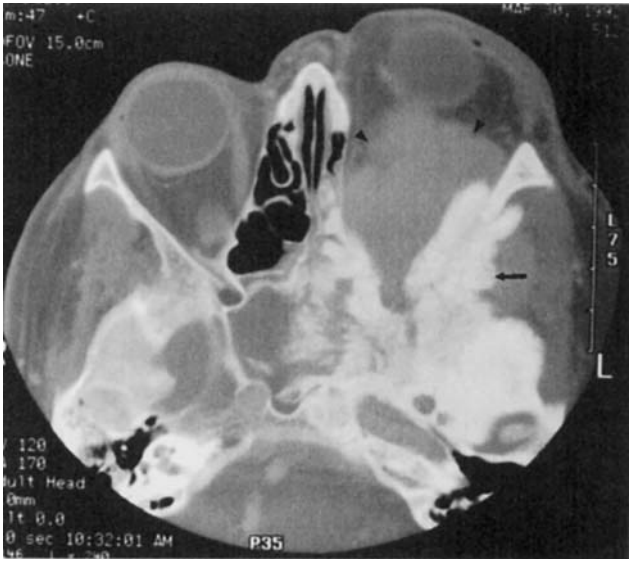

Figure 11. Meningioma. A 59-year-old woman with a large sphenoid meningioma and intraorbital extension presented with blindness in the left eye and severe proptosis. Marked hyperostosis of the left sphenoid wing and lateral orbital wall (*arrow*) is characteristic of a meningioma. Note the large soft-tissue mass representing meningioma extension into the orbit (*arrowheads*).

arising from the jugular foramen are called glomus jugulare tumors, and paragangliomas in the middle ear are referred to as glomus tympanicum lesions) (Figs. 12–14). These tumors are histologically identical to the glomus vagale and carotid body tumors. A hereditary disposition has been identified in 15% to 20% of cases.[8] Synchronous or metachronous paragangliomas are present in 10% of cases, necessitating a careful search for other tumors. Approximately 1% of paragangliomas secrete physiologically activated catecholamines.[25] The most common presenting symptoms include pulsatile tinnitus, conductive hearing loss, and cranial nerve deficits. Glomus jugulare tumors cause irregular lytic bony destruction of the jugular foramen, which is apparent on CT scan. They enhance vividly on postcontrast CT scans and MR images, and prominent flow voids on MR images give the lesion a salt-and-pepper appearance. Frequently, the hypoglossal canal is invaded, resulting in twelfth nerve paresis.

Meningiomas and schwannomas are less common lesions of the jugular foramen. Meningiomas are more common in women in their third or fourth decade of life. These lesions may have a high density on precontrast CT scan and enhance vividly. Adjacent hyperostosis may be present. Schwannomas are rare neurogenic neoplasms arising from the ninth, tenth, or eleventh cranial nerves exiting the jugular foramen. They cause smooth enlargement of the foramen without bony destruction and enhance markedly on gadolinium-enhanced MR images. These lesions, like glomus jugulare tumors, may cause compression and paresis of the ninth, tenth, and eleventh nerves.

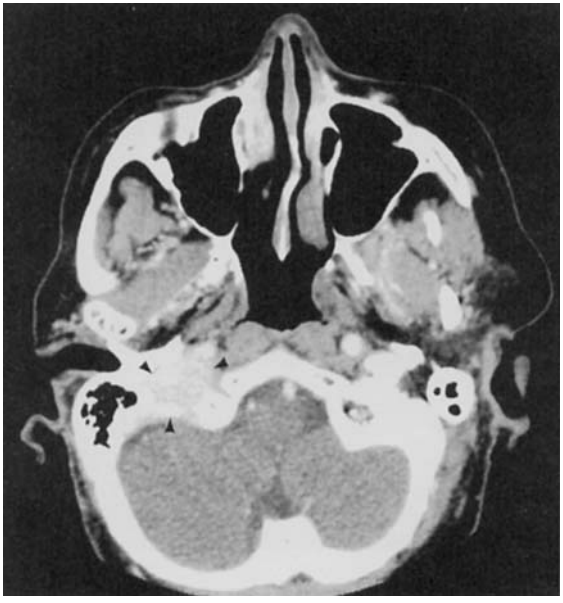

Figure 12. Glomus jugulare. Axial postcontrast CT scan reveals an enhanced mass (*arrowheads*) at the right jugular foramen. Bone window images reveal jugular foramen erosion characteristic of glomus jugulare (versus smooth enlargement of the foramen associated with schwannomas and meningiomas).

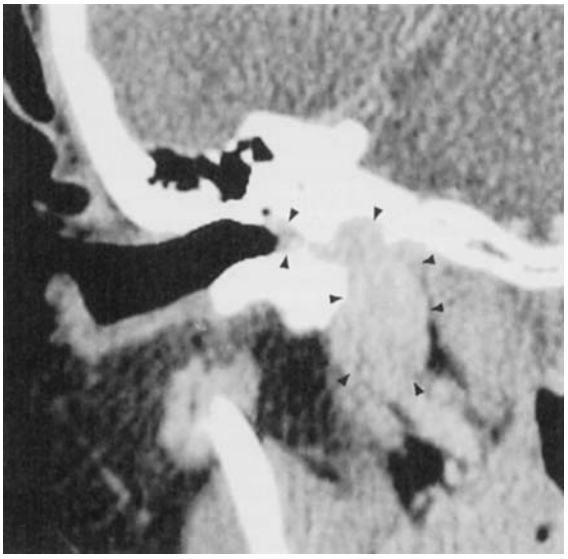

Figure 13. Glomus jugulotympanicum. Coronal postcontrast CT scan reveals an enhanced mass at the jugular foramen with extension into the hypotympanum and middle ear (*arrowheads*).

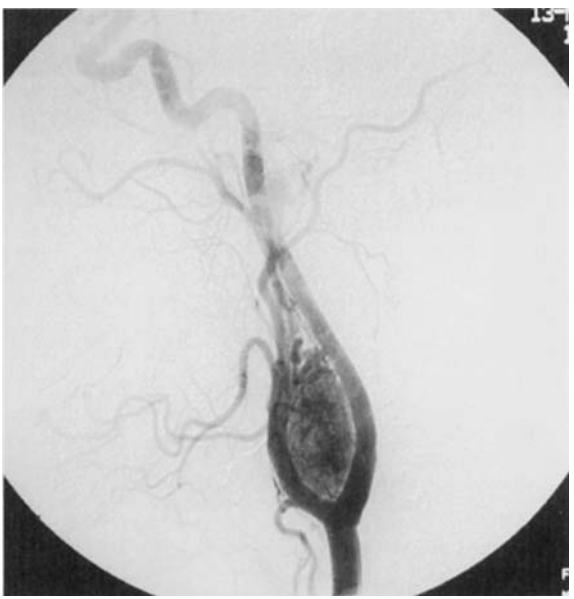

Figure 14. Paraganglioma. This carotid injection from a cerebral arteriogram reveals the characteristic hypervascularity associated with paragangliomas in this carotid body tumor situated at the carotid bifurcation. A small glomus jugulare, not apparent on this image, was present on the postcontrast CT scan. Paragangliomas are multiple in 15% to 20% of patients.

Tumor Spread Involving the Skull Base

Tumor spread to the skull base may occur by three methods: (1) direct extension from the nasopharynx, (2) perineural spread along the nerve sheaths of cranial nerves, and (3) hematogenous metastases. Nasopharyngeal carcinoma may invade the skull base through the sinuses of Morgagni, which are paired natural gaps in the barrier of the pharyngobasilar fascia that permit spread of disease to the skull base and beyond (Fig. 15). Once the neoplasm has invaded the clivus, the tumor may extend into the intracranial vault, invade the dura, and cause cranial nerve palsies. Because of its relative late clinical presentation, patients may present with cranial neuropathy. Sixteen percent of patients present with an abducens (cranial nerve VI) palsy, and 14% of patients present with a trigeminal (cranial nerve V) palsy. Ninety-five percent of all nasopharyngeal tumors are squamous cell carcinomas.[20] The other 5% include lymphoma arising from the lymphoid rich adenoids, minor salivary gland neoplasms, and rhabdomyosarcoma in the pediatric population.

Skull base bone destruction is best visualized with CT scan, and this modality is used at the Wake Forest University School of Medicine for evaluating patients with nasopharyngeal carcinoma, primarily because concomitant imaging of the neck is obtained for staging purposes. Because of MR imaging's increased sensitivity in detecting cranial nerve involvement, dural invasion, bone marrow infiltration, and brain parenchymal invasion, it is the first study of choice for cases in which the patient presents with a cranial neuropathy with an intracranial or perineural spread of disease.

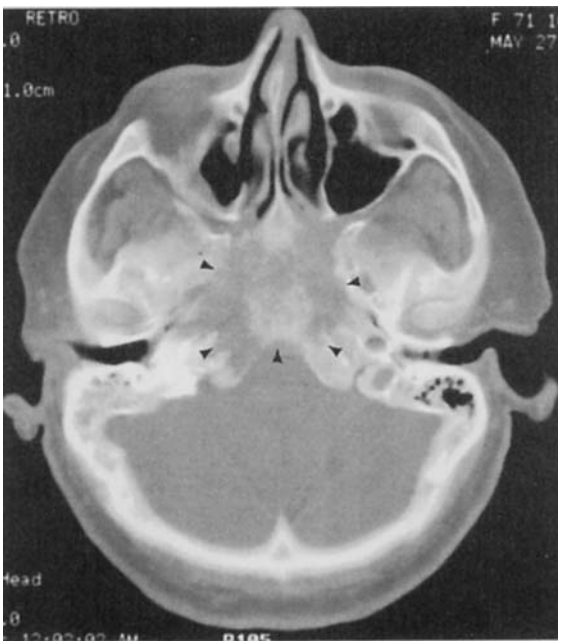

Figure 15. Nasopharyngeal carcinoma. There is permeative, lytic bone destruction of the basisphenoid (*arrowheads*) in this patient with nasopharyngeal carcinoma and skull base invasion. Postcontrast CT scan revealed bilateral cavernous sinus invasion.

Perineural spread is the dissemination of tumor from the primary site along tissue planes of cranial nerves, the neural sheaths of which serve as a conduit through which neoplasms may spread intracranially.[9] Perineural spread may occur in any head and neck malignancy, including salivary gland tumors, mucosal lesions, and skin carcinoma. Infrequently, infections such as rhinocerebral mucormycosis may extend to cranial nerves in perineural fashion.[17] Although adenoid cystic carcinoma is notorious for spreading in this fashion, squamous cell carcinoma is a very common lesion that spreads in this way. The second and third divisions of the trigeminal nerve and the descending facial nerve are most commonly affected by perineural spread. Lesions involving the mandible or infratemporal fossa can spread in retrograde fashion along V3 through the foramen ovale and into Meckel's cavity and the cavernous sinuses (Figs. 16 and 17). Maxillary or hard palate lesions can extend in retrograde fashion along infraorbital or superior alveolar nerves, pass through the pterygopalatine fossa, and extend intracranially through the vidian canal or foramen rotundum. The greater and lesser palatine nerves comprise another potential pathway for perineural spread, especially for soft palate lesions. Aggressive parotid lesions may spread along the descending branch of the facial nerve.

Perineural spread is a serious finding and is associated with decreased survival.[10] Identification of this process may indicate the need for a wider resection or may indicate nonresectability, thereby necessitating radiotherapy. Imaging evidence of perineural spread includes the following: (1) widening or destruction of and excessive enhancement within neural foramina (rotundum, ovale, palatine,

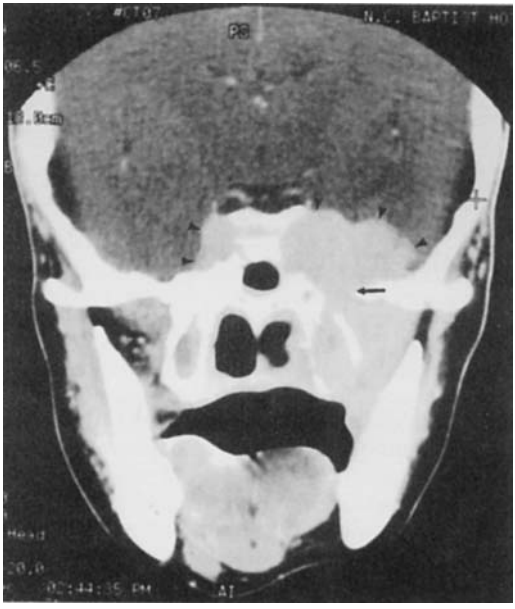

Figure 16. Perineural spread of adenoid cystic carcinoma (*soft palate*). Adenoid cystic carcinoma is notorious for perineural spread of tumor. Coronal postcontrast CT scan reveals tumor infiltration of the left infratemporal fossa (masticator space) and pterygopalatine fossa, with intracranial spread through the foramen ovale (*arrow*) into the left middle cranial fossa and bilateral cavernous sinuses (*arrowheads*).

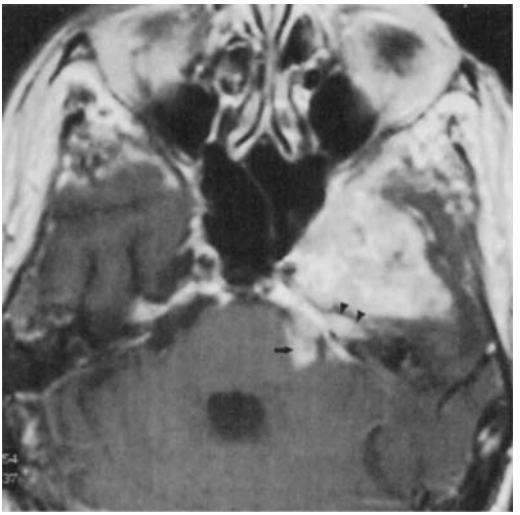

Figure 17. Perineural spread of adenocarcinoma arising from the parotid gland. Intracranial spread of malignancy is apparent on this postgadolinium T1-weighted MR image. There is spread of tumor in the left middle cranial fossa, cavernous sinus, internal auditory canal (*arrowheads*), and cisternal portion of the trigeminal nerve (*arrow*), resulting from perineural spread along the mandibular division of the trigeminal nerve through the foramen ovale.

stylomastoid, or vidian canal), (2) replacement of fat density on CT scan or signal intensity on MR imaging and abnormal enhancement or widening of the pterygopalatine fossa or pterygomaxillary fissure, and (3) expansion or abnormal enhancement of Meckel's cavity or the cavernous sinuses.[17] Postgadolinium axial and coronal T1-weighted MR images with fat suppression best identify perineural spread, although CT scan frequently demonstrates this entity. CT scanning is better at detecting cortical bone destruction associated with perineural spread.

INTERVENTIONAL RADIOLOGY

Interventional radiology offers many procedures that may improve the outcome of skull base surgery. Preoperative embolization may be useful in limiting intraoperative blood loss in the following skull base tumors: intracranial meningiomas, paragangliomas, and juvenile nasopharyngeal angiofibromas. Intravascular embolization entails the use of microcatheters to gain access to distal feeding vessels in which the embolic agent can be deployed to occlude intratumoral vessels. Various embolic agents may be employed, usually some type of microparticulate such as polyvinyl alcohol foam particles (Fig. 18). Risks of intravascular embolization include stroke, blindness, cranial nerve paresis, and skin necrosis, and these procedures require vigilance in identifying dangerous anastomoses and collaterals that may become apparent during the embolization.

Intravascular embolization is a particularly useful procedure for juvenile nasopharyngeal angiofibromas that classically are associated with a large blood loss. Major supplying vessels include the nasopharyngeal and nasal branches of the internal maxillary artery, with contributions from the accessory meningeal, ascending pharyngeal, and ascending palatine arteries. Definitive treatment of angiofibromas is total surgical resection. Because these lesions are highly vascularized,

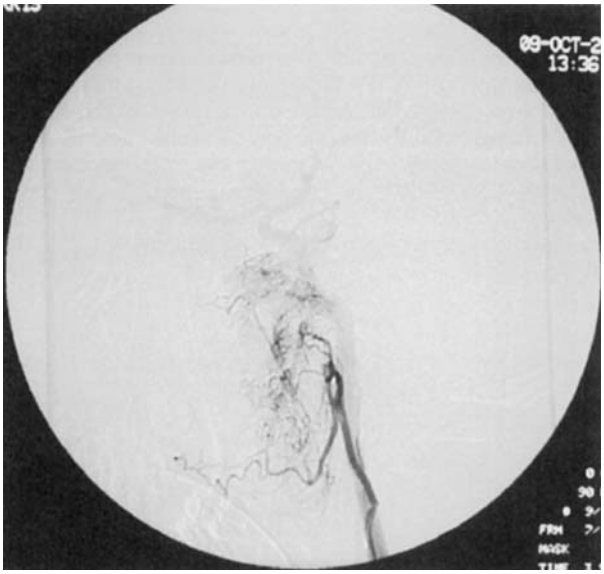

Figure 18. Juvenile nasopharyngeal angiofibroma. Selective arteriogram of the ascending pharyngeal artery reveals tumor hypervascularity. This lesion was embolized preoperatively with polyvinyl alcohol particles through a microcatheter to minimize intraoperative blood loss.

preoperative angiography and transcatheter embolization play critical roles in facilitating aggressive surgery by decreasing intraoperative blood loss and lesion size thereby increasing visibility in the operative field. Similar endovascular procedures may be used for preoperative embolization of meningiomas and glomus tumors.

CT-guided biopsy is a minimally invasive procedure that may be an alternative to surgical or open biopsy. It can be used to acquire tissue from lesions in difficult locations, including the pterygopalatine fossa and parapharyngeal space (Fig. 19).

SUMMARY

Many imaging modalities play a role in evaluating patients with skull base pathologies. CT scanning and MR imaging are the major modalities used to characterize skull base neoplasms. Pathologic entities vary in the different regions of the skull base, and imaging characteristics on CT scans and MR images frequently are suggestive of the diagnosis. Arteriography characterizes paragangliomas and angiofibromas, and occasionally these lesions can be treated preoperatively by endovascular embolization. PET has a role in differentiating recurrent or residual tumors from post-treatment changes (Fig. 20). In many cases, the use of these modalities helps to optimize patient outcome.

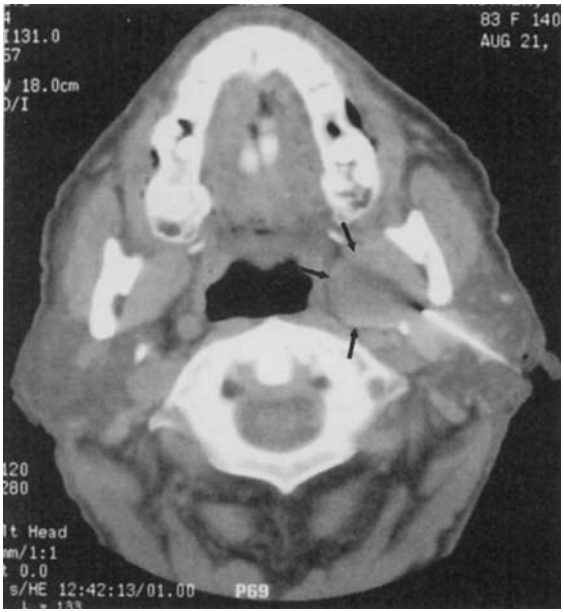

Figure 19. CT-guided biopsy. Percutaneous biopsy of head and neck masses is possible using CT scanning for localization. CT scanning was used to obtain tissue from a bilobed mass in the left parapharyngeal space (*arrows*). Biopsy yielded a milky fluid suggestive of a branchial cleft cyst.

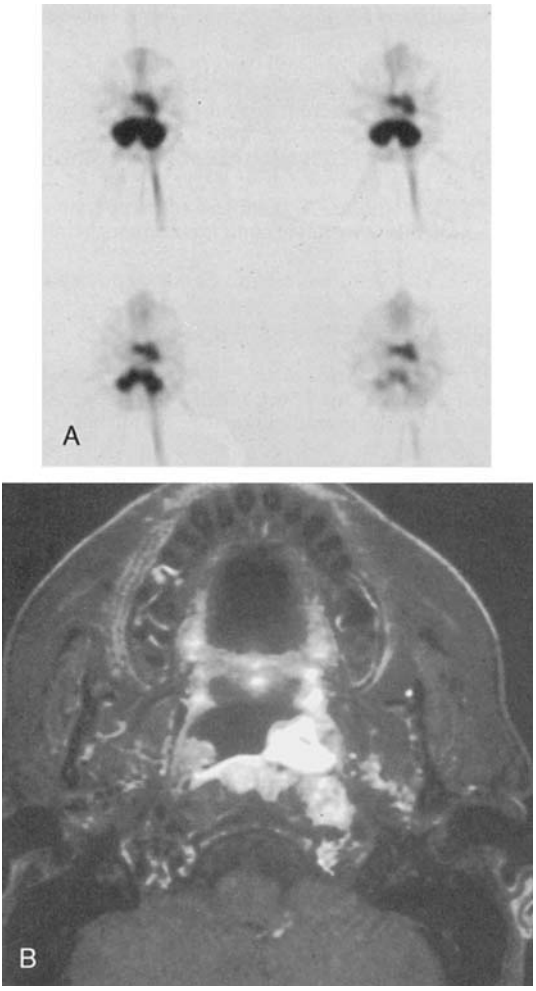

Figure 20. *A*, Axial fluorodeoxyglucose (FDG) PET images of the nasopharynx and posterior fossa reveal abnormal FDG uptake in the left nasopharynx (squamous cell carcinoma). Note normal brain uptake in the cerebellum. *B*, Postgadolinium T1-weighted MR images show corresponding enhanced left nasopharyngeal mass and medial and lateral retropharyngeal nodes.

References

1. Barnes L, Verbin RS, Gnepp DR: Diseases of the nose, paranasal sinuses, and nasopharynx. *In* Barnes L (ed): Surgical Pathology of the Head and Neck, vol 1. New York, Marcel Decker, 1985, pp 403–451
2. Connors JJ, Wojak JC: Juvenile Nasopharyngeal Angiofibromas, Interventional Neuroradiology: Strategies and Practical Techniques. Philadelphia, WB Saunders, 1999, pp 121–130

3. Curtin HD: Central skull base: RSNA Categorical Course in Diagnostic Radiology. Neuroradiology, 2000, pp 143–151
4. Curtin HD, Som PM, Braun IF, et al: Skull base. *In* Som PM, Curtin HD (eds): Head and Neck Imaging, vol 2. St. Louis, Mosby, 1996, pp 1233–1299
5. Davis KR, DeBrun GM: Embolization of juvenile nasopharyngeal angiofibromas. Seminars in Interventional Radiology 4:309–320, 1987
6. Derdeyn CP, Moran CJ, Wippold FJ, et al: MRI of esthesioneuroblastoma. J Comput Assist Tomogr 18:16–21, 1994
7. Eisen MD, Yousem DM, Montone KT, et al: Use of preoperative MR to predict dural, perineural, and venous sinus invasion of skull base tumors. Am J Neuroradiol 17:1937–1945, 1996
8. Fuller AM, Brown HA, Harrison EG Jr, et al: Chemodectomas of the glomus jugulare tumors. Laryngoscope 77:218–238, 1967
9. Ginsberg LE: Imaging of perineural tumor spread in head and neck cancer. Semin Ultrasound CT MR 20:175–186, 1999
10. Goepfert H, Dichtel WJ, Medina JE, et al: Perineural invasion in squamous cell skin carcinoma of the head and neck. Am J Surg 148:542–547, 1984
11. Grossman RI, Yousem DM: Neuroradiology: The Requisites. St. Louis, Mosby, 1994, pp 359–376
12. Harnsberger HR: Handbook of Head and Neck Imaging, ed 2. St. Louis, Mosby, 1995, p 400
13. Harnsberger HR: The posterior skull base, with emphasis on the clivus, petro-occipital fissure, and jugular foramen: RSNA Categorical Course in Diagnostic Radiology. Neuroradiology, 2000, pp 153–166
14. Jinkins JR: Atlas of Neuroradiology Embryology, Anatomy, and Variants. Philadelphia, Lippincott, Williams, & Wilkins, 2000, pp 86–88
15. Keyes JW, Watson NE, Williams DW: FDG PET in head and neck cancer. Am J Radiol 169:1663–1669, 1997
16. Mancuso AA: Diagnostic imaging. *In* Meyerhoff WL, Rice DH (eds): Otolaryngology: Head and Neck Surgery. Philadelphia, WB Saunders, 1992, pp 27–118
17. McLean FM, Ginsberg LE, Stanton CA: Perineural spread of rhinocerebral mucormycosis. Am J Neuroradiol 17:114–116, 1996
18. Parker GD, Harnsberger HR: Clinical-radiologic issues in perineural tumor spread of malignancies of the extracranial head and neck. Radiographics 11:301–308, 1989
19. Regenbogen VS, Zinreich SJ, Kim KS, et al: Hyperostatic esthesioneuroblastoma. J Comput Assist Tomogr 12:52–56, 1988
20. Rice DH: Neoplastic disease. *In* Meyerhoff WL, Rice DH (eds): Otolaryngology: Head and Neck Surgery. Philadelphia, WB Saunders, 1992, pp 531–532
21. Som PM: Anterior skull base: RSNA Categorical Course in Diagnostic Radiology. Neuroradiology, 2000, pp 133–141
22. Som PM, Brandwein M: Sinonasal cavities: Inflammatory diseases, tumors, fractures, and postoperative findings. *In* Som PM (ed): Head and Neck Imaging, vol 1. St. Louis, Mosby, 1996, pp 190–210
23. Som PM, Lidov M, Brandwein M, et al: Sinonasal esthesioneuroblastoma with intracranial extension: Marginal tumor cysts as a diagnostic MR finding. Am J Neuroradiol 15:1259–1262, 1994
24. Yousem DM, Lexa FJ, Bilaniuk LT, et al: Rhabdomyosarcomas in the head and neck: MR imaging evaluation. Radiology 177:683–686, 1990
25. Zak F, Lawson W: The paraganglionic chemoreceptor system. *In* Physiology, Pathology, and Clinical Medicine. New York, Springer-Verlag, 1982, pp 287–411

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