

Unusual Presentations of Adenoid Cystic Carcinoma in Extra-Salivary Gland Subsites in Head and Neck Region: A Case Series

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Abstract Adenoid cystic carcinoma (ACC) is an uncommon malignant epithelial tumor occurring in head neck region, accounting for approximately 10% of all salivary gland neoplasms. Extra-salivary gland involvement of other head neck subsites by ACC may present in unusual modes creating clinical dilemma. Here we present some cases with extra-salivary gland ACC. In this series six cases with extra-salivary gland ACC were included. Three patients presented with external auditory canal and temporo-mastoid region involvement of whom two cases also had intracranial extension. In one case tongue was the site of involvement where it was confused with benign neurogenic tumour. Palatal erosion with formation of oro-nasal fistula was the the manifestation of ACC involving palate. On the other hand sinonasal bleeding mass which came out as ACC on histopathological examination did not cause any palatal erosion and was removed successfully via lateral rhinotomy. In all these cases patients were treated with post-operative radiotherapy and were closely followed up for early detection of any distant metastasis. The experience about the various unusual manifestations of ACC and the therapeutic challenge poised by the cases is discussed here.

Keywords Adenoid cystic carcinoma ·
Extra-salivary gland manifestations · Palatal ·
Intracranial extension

Introduction

Adenoid cystic carcinoma (ACC) is an uncommon malignant epithelial tumor [1], occurring in head neck region, accounting for approximately 5–10% of all salivary gland neoplasms representing 2–4% of malignant occurrences of the head and neck area [2, 3]. Although parotid gland is the single most common site of origin (25%) in head and neck, but most of the ACC arise in minor salivary glands (60%), for which oral cavity (palate) is the most common site of involvement [2]. It has also been reported to occur in breast, lacrimal gland, lung, brain, bartholin gland, trachea and paranasal sinuses in literature. ACC is usually presents insidiously, generally advanced when diagnosed and has a propensity for perineural spread [2]. Surgery and irradiation are the current effective treatment modalities for ACC but locoregional recurrences are frequent [2].

Case Discussion

Patients included in this series were selected from the otolaryngology out-patient department of our institution during the period of 3 years (Sept. 2006–Aug. 2009). During this period six patients were diagnosed as ACC involving extra-salivary subsites in head-neck region. Of the six patients two were male and the rest were female (M:F = 1:2) (Table 1).

In this present series maximum number of extra-salivary gland presentation were involving external auditory canal

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Table 1 Showing distribution of cases according to areas of involvement

Area of involvement by ACC	No. of cases
Temporo-mastoid region	3
Tongue	1
Palate	1
Nose and Para-nasal sinuses	1

**Fig. 1** CT scan of brain showing soft tissue mass in temporal region with underlying bone destruction and intracranial extension

and temporo-mastoid region. Of the three cases two had intracranial extension confirmed on CT scan (Fig. 1). These cases presented with temporo-mastoid mass occluding external auditory canal with multiple discharging sinuses (Fig. 2) accompanied by ipsilateral excruciating pain and facial nerve paralysis. Initially these cases were presumed to be squamous cell carcinoma involving temporal bone. But surprisingly the fine-needle aspiration cytology (FNAC) report came as ACC. These two cases were treated by combined effort of neurosurgeons and ENT surgeons, performing lateral temporal bone resection with removal of intracranial extension by neurosurgeons. The scalp defect was covered by local scalp rotation flap. The other case presented as a solitary mass involving cartilaginous part of right external auditory canal without any facial nerve palsy or intracranial extension. It was removed with adequate margin. The final histopathological report was ACC which was unanticipated from the nature of clinical presentation of the mass. All the six cases included in this study were treated with post-operative radiotherapy in order to improve locoregional control. Of the two cases involving temporo-

**Fig. 2** Temporo-mastoid mass with overlying skin induration and multiple discharging sinuses

mastoid region with intracranial extension developed metastatic ACC of ipsilateral parietal bone, 9 months after primary resection and was operated subsequently. There were no further recurrence or distant metastasis in any of the three cases of ACC of temporo-mastoid region till date.

The mass involving middle and posterior third of tongue was of short duration, about one and a half years and on FNAC was reported as neurogenic tumour but the exact tissue diagnosis was not confirmed. Keeping in mind the location of the mass within the tongue MRI study was undertaken which showed homogenous enhancing mass involving right half of tongue, middle and posterior thirds, encroaching floor of mouth but not extending deeper to mylohyoid muscle and not destroying midline tissue septum of tongue (Fig. 3). Complete resection of the mass was undertaken under general anaesthesia via mandibular split and right hemiglossectomy with supraomohyoid neck dissection was performed. Deep resection plane and proximal resection margin were free from involvement by the tumor.

The case of palatal erosive growth with oro-nasal fistula (Fig. 4) was biopsied from margin of the growth, the histological diagnosis was ACC. The patient was treated with local radical excision and post-operative radiotherapy. Closure of oro-nasal fistula by an obturator was performed after achieving locoregional control of the disease. Tongue and palatal ACC cases were free from any locoregional recurrence or distant metastasis.

The sino-nasal mass presented with recurrent epistaxis in a middle aged female associated with broadening and deformation of the external nasal framework along with development of discharging sinuses over left lateral nasal wall (Figs. 5, 6). The patient suffered from excruciating pain over face on the side of involvement, but surprisingly

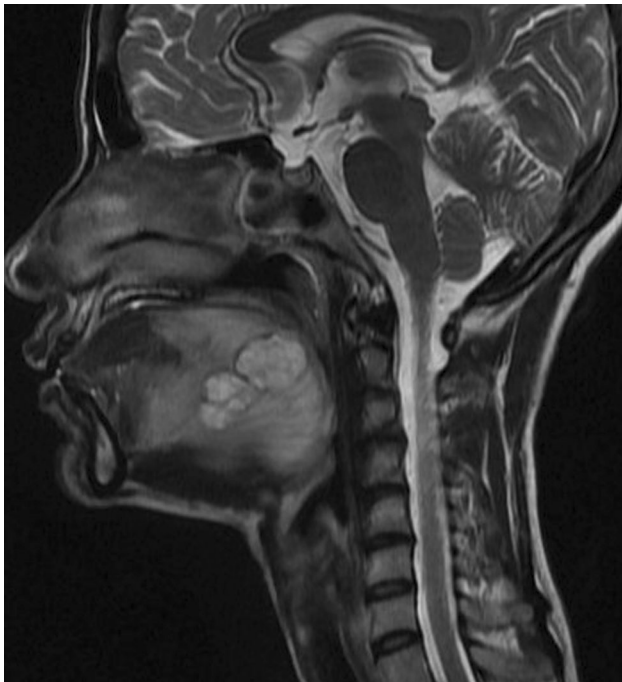


Fig. 3 MRI of tongue showing homogeneously enhancing mass in right half of tongue, middle and posterior third area

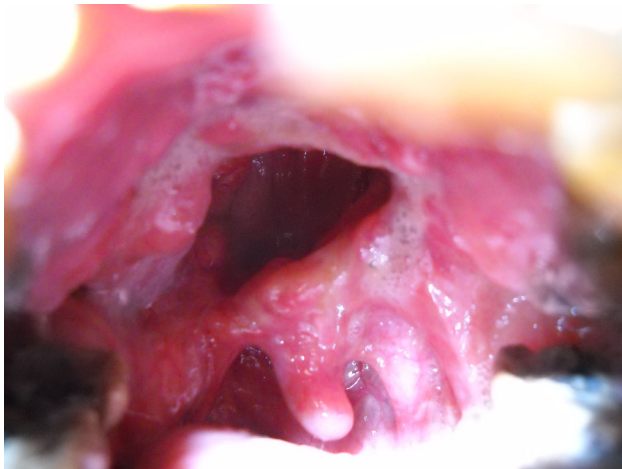


Fig. 4 Palatal erosive growth with oro-nasal fistula

there was no palatal erosion. Although a sino-nasal malignancy was suspected the endoscopic biopsy from nasal mass on histopathology was reported as ACC. We planned for radical surgery to ensure local clearance encompassing complete removal of mass as well as sinus tracts and the surrounding skin along with involved external nasal framework. This was achieved via lateral rhinotomy approach with the wide soft tissue defect covered by forehead rotational flap. The patient developed distant lung metastasis in the form of solitary pulmonary nodule of size 3 × 3 cm in left lung mid zone, located

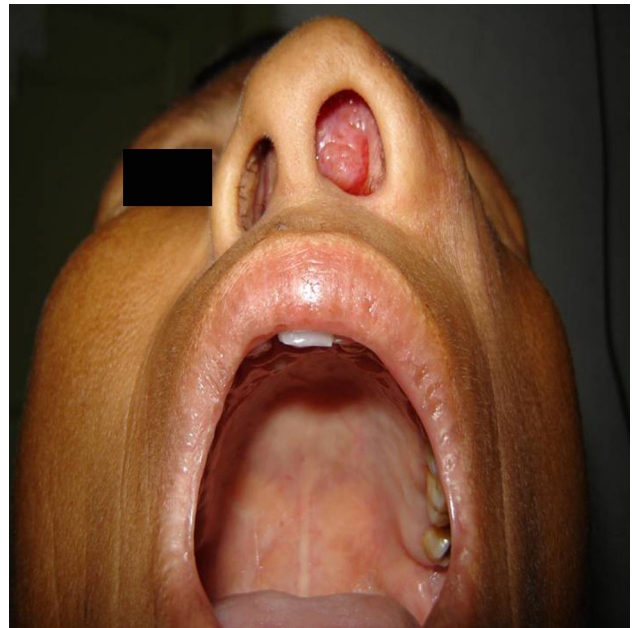


Fig. 5 Left nasal mass with no palatal erosion



Fig. 6 Broadening and deformation of the external nasal framework along with discharging sinuses over left lateral nasal wall

peripherally which was noted in HRCT scan of thorax performed 6 months after primary resection as the patient developed shortness of breath on exertion. The lung metastasis was resected completely by cardiothoracic surgeons, facilitated by its peripheral localization and she is free from any further recurrence till date.

All the six cases were closely followed up for the next 2 years for the development of any local or distant metastasis and is still under follow-up. Pulmonary, hepatic and bone metastasis were searched for in all cases by HRCT scan of thorax, CT scan of abdomen and isotope bone scan respectively.

Discussion

The first case of ACC was described by three Frenchmen (Robin, Lorain and Laboulbene) in two articles published in 1853 and 1854, who also described cylindrical appearance of the tumour [4]. Billroth described that ACC has a great tendency to recur [2]. Spies in 1930 coined the term adenoid cystic carcinoma [5].

Foote and Frazell described ACC located in major and minor salivary glands, tumours were incompletely encapsulated and variations in histology had an effect to indicate the propensity of perineural spread [6]. Conley and Dingman described ACC as “one of the most biologically destructive and unpredictable tumours of head and neck” [7]. The paradoxical clinical behaviours of ACC are, firstly tumour growth is slow, relentless and progressive, secondly surgical resection although feasible multiple local recurrences are the rule and thirdly metastatic spread to regional lymph nodes is uncommon but distant spread to lungs and bones is frequent [2].

Histologically ACC can be categorized into three growth patterns, cribriform, tubular and solid, of which solid variety has worse prognosis due to advanced stage and frequent association with distant metastases [8, 9]. Szanto et al. [10] graded ACC into three grades, cribriform or tubular (grade I), less than 30% solid (grade II) and greater than 30% solid (grade III). Cribriform and tubular variants showed no difference in rate of distant metastases and overall survival when treated with similar modalities. Cribriform variant has more chance of local recurrence [11]. Evesson and Cawson [12] found a discreet predominance of ACC cases in women (F:M = 1.2:1). Our study also corroborated with this finding (F:M = 2:1).

Regarding various extra-salivary subsites of involvement Perzin KH et al. described 16 cases of ACC involving the external auditory canal in a clinicopathological study of which in eight cases it was confined to the external auditory canal probably originating from the ceruminous glands and on incomplete excision nine patients suffered from twenty-six local recurrences. Death was usually caused by intracranial extension of the tumour or by pulmonary metastases [13]. Abe T et al. [14] described a case of ACC with striking intracranial extension where a middle aged woman had external auditory canal ACC excised and developed recurrence of ACC in ipsilateral temporal lobe. In our study there

were three cases involving temporo-mastoid region and external auditory canal with two cases having intracranial extension reflecting the similar scenario.

Tongue as the primary site of involvement by ACC has been described by Kuauhyama Luna-Ortiz et al. [15] in a retrospective analysis of database of sixty-eight ACC cases, of whom eight cases (11.7%) involved tongue—seven cases were female and one case was male with an average age of 51 years. Previous series of ACC of the head and neck region have shown that the frequency in which the tongue has been the site of origin ranged from 3.4 to 17.1% [16–18]. Our study also corroborated with aforementioned data with one case of ACC tongue out of six cases (16.7%) occurring in a middle aged female. Eduardo Costa Studart Soares et al. [19] described ACC involving posterior third of tongue similar to the location of the lesion found in our study. Namazie A et al. [20] performed retrospective review of 14 cases of ACC involving base of tongue. Dolores Carrasco Ortiz et al. [21] reported a case of ACC presenting over dorsum of tongue.

31% of the cases of ACC in head and neck region affect minor salivary glands, particularly the palate [3]. In a report by Spiro et al. [22], of 242 salivary gland ACC cases, 171 patients presented lesions involving accessory glands, while 64 patients (26%) presented the palate as the affected site, being the tongue the second most affected area.

A little less than 5% of sinonasal malignancies are ACC. Maxillary sinus is the most commonly affected site and patients suffer from facial pain that has defied diagnosis for long duration [23]. In our study also the patient of sinonasal ACC suffered from excruciating facial pain.

Possible treatments of ACC include four different modalities: surgical therapy, radiotherapy, chemotherapy and combined therapy (surgery and radiotherapy, radiotherapy and chemotherapy). In most cases combined therapy is the treatment of choice [20, 24–26]. Only surgical removal or radiotherapy in isolation may fail to eliminate the possibility of recidivation in surgical margins, as well as the occurrence of metastasis in cervical lymph nodes, lungs, bones and brain [27]. All the six cases in our study were treated with combined therapy using surgery followed by radiotherapy.

Conclusion

Extra-salivary gland involvement of ACC in head-neck region is not very uncommon and the possibility should be kept in mind in long standing cases, cases with extensive local destruction and in patients suffering from excruciating local pain out of proportion to the extent of disease. Pre-operative diagnosis of ACC either by punch biopsy or

FNAC helps us to plan for radical local excision followed by post-operative radiotherapy which is the treatment of choice for ACC. Moreover we have to maintain prolonged follow-up of the patients in order to be able to find out locoregional recurrences as well as distant metastases at an early stage when it is still possible to salvage the patient.

Conflict of interest None.

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