

Ameloblastic Carcinom of Maxilla: A Rare Entity

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Abstract : Odontogenic carcinomas of the maxilla are classified as ameloblastic carcinoma, malignant ameloblastoma and primary intraosseous carcinoma. The maxilla is an unusual site for ameloblastic carcinoma. This report is of a patient with a locally advanced ameloblastic carcinoma of the left maxilla. Treatment consisted of left maxillectomy and free-flap reconstruction, followed by postoperative adjuvant external beam radiation. A literature review describing clinical and histological presentation of this rare tumour is presented.

Keywords: Ameloblastoma, ameloblastic carcinoma, maxilla, odontogenic carcinoma, radiotherapy

I. Introduction

Ameloblastoma is a locally aggressive benign odontogenic tumour that originates from the remnants of dental epithelium, 80% of ameloblastomas occur in the mandible and 20% occur in the maxilla. The malignant variant of ameloblastoma 'Primary ameloblastic carcinoma' as defined by the World Health Organization (WHO) is a tumour that demonstrates the morphological features of ameloblastoma with atypia, regardless of the presence or absence of metastasis[1]. Ameloblastic carcinomas may arise denovo, ex- odontogenic cyst, and ex-ameloblastoma. Most of them are known to arise denovo. It may arise in the form of a cystic lesion with benign clinical features, or as a large tissue mass with ulceration and bone resorption.

Ameloblastic carcinomas do not show any age-group predilection. An age range of 15 to 84 years has been reported, mean reported age was age 30 years[2]. There is no sex predilection[3]. Swelling, pain, and trismus are the common presenting symptoms. Radiographic findings include poorly defined radiolucency, with focal radiopacities. These tumors show an aggressive course with extensive local destruction and metastatic spread to cervical lymph nodes and distant metastasis to lung and rarely to brain and bones. Less than 30 cases of maxillary ameloblastic carcinomas are reported in literature[4]. This report is of a patient with a rare ameloblastic carcinoma of the left maxilla.

II. Case History

A 52 year old man presented to our institute with complaints of swelling over left maxillary region and ulceroproliferative growth over left upper alveolar region of 2 months duration[Fig 1]. He is a tobacco chewer and alcoholic. On local examination, 4X5cm ulcero-proliferative swelling in the left upper alveolus was palpable which was bleeding on touch with loss of upper left molars and premolars. Swelling on the left side of maxilla was measuring 6X6 cm. CT scan showed a large hypodense mass on left upper alveolus with erosion of maxilla, pterygoid, and zygomatic bone with extension into infratemporal region[Fig 2]. Initial biopsy taken from the growth was suggestive of squamous cell carcinoma.

The patient underwent maxillectomy with forehead flap reconstruction[Fig 3]. Intra operative findings were 6X5X4cm tumor involving lateral wall of maxilla and extending into oral cavity eroding palate and alveolus from midline to gingivobuccal sulcus in the oral cavity. The tumor was extending to infratemporal fossa and orbital cavity eroding the bones. Orbital soft tissues and maxillary mucosa were free from tumor infiltration. Post operative histopathological examination showed that the lesion proper was composed of cells which were arranged in the form of cords, trabeculae, nesting and pseudo glandular patterns [Fig 3,4]. These cells were polygonal in shape with vesicular nuclei prominent nucleoli and moderate amount of cytoplasm. Some areas within lesion showed nests of cells with peripheral palisading of nuclei and central loosely arranged cells resembling the stellate reticulum. These cells showed moderate degree of nuclear pleomorphism. Zygomatic bone resected margins and posterior mucosal soft tissue margins were involved by malignancy and all other margins were free. Immunohistochemistry (IHC) was done and High molecular weight cytokeratin (HMWK) [Fig 5] and calretinin[Fig 6] were positive and S100 was negative [Fig 7] and hence the case was diagnosed as ameloblastic carcinoma.

Patient was evaluated post operatively with CTscan which showed a 3.8X3.8X3 cm irregular mildly enhancing mass in the region of left maxillary antrum with bilateral enlarged level II cervical lymph nodes. Patient was treated with external beam radiotherapy(RT) by intensity modulated radiotherapy (IMRT) technique

to a total dose of 70.2 Gy in 39 fractions @ 1.8Gy per fraction with concurrent weekly cisplatin chemotherapy. Patient completed treatment and is on regular follow up for the last two years and is disease free.

III. Discussion

The diagnosis, histological features and treatment of ameloblastic carcinoma remains challenging. The term ameloblastic carcinoma was first described by Shafer et al in 1983[5]. There is a lack of pathological features to distinguish ameloblastoma with no malignant potential from malignant histology that may metastasise. In 2005, the WHO revised the classification of odontogenic carcinoma and described primary ameloblastic carcinoma as an odontogenic malignancy that combined the histological features of ameloblastoma with cytological atypia with or without metastasis.

The differential diagnosis for ameloblastic carcinomas at this site includes squamous cell carcinoma, primary intra-alveolar epidermoid carcinoma, and mucoepidermoid carcinoma. Metastases in the jaws from visceral neoplasms and invasion of bone by a tumour from adjacent soft tissue or paranasal sinus are another possibility. Odontogenic tumours are lesions derived from epithelial, ectomesenchymal or mesenchymal elements that are part of the tooth forming apparatus. The etiology of benign and malignant odontogenic tumours is unknown and the majority arise without an apparent causing factor. Malignant odontogenic tumours are classified as odontogenic carcinomas and odontogenic sarcomas.

Ameloblastic carcinoma is characterised by malignant cytological features within histologic pattern of an ameloblastoma. The usual histological features are tall columnar epithelium with pleomorphism, mitotic activity, focal necrosis and perineural invasion and nuclear hyperchromatism. Peripheral palisading with a background of stellate reticulum structure and epithelial lining of cystic spaces can be found. Atypical cells are seen to form nests and broad ribbons which branch and anastomose. They show high proliferative index. Ameloblastomas unlike ameloblastic carcinomas do not have features of necrosis, perineural invasion or high proliferative index. The present case showed polygonal cells arranged in the form of cords, trabeculae, nesting and pseudo glandular pattern. Some areas within lesion showed nests of cells with peripheral palisading of nuclei and central loosely arranged cells resembling the stellate reticulum. IHC showed positivity for HMWK and calretinin in the present case and negative for S100. Secondary ameloblastic carcinomas arise from a pre-existing ameloblastoma and are very rare. They are divided into intraosseous and peripheral types. A pre-existing microscopically proven benign ameloblastoma should be present for a diagnosis of secondary ameloblastic carcinoma. They have nests, strands and follicles of recognisable ameloblastoma type histology within variable degrees of squamous differentiation.

The differential diagnosis also includes primary intraosseous squamous cell carcinoma which is characterised by islands of neoplastic squamous cell carcinoma with moderate differentiation and without prominent keratinisation. Metastatic squamous cell carcinoma should be excluded but there are no histopathological features to distinguish a primary intraosseous squamous cell carcinoma from a metastatic squamous cell carcinoma. However, the epithelium of the squamous odontogenic tumour lacks any cytological evidence of malignancy. Squamous cell carcinoma arising in the lining of an odontogenic cyst closely resembles oral squamous cell carcinoma. The present case was initially diagnosed as squamous cell carcinoma on biopsy.

Kruse et al have reported 26 cases of maxillary ameloblastic carcinomas in their review article[4]. Mean age at diagnosis was 54.4 years with marked prevalence in the 41-80 year age group, 26.9% of patients had lung metastases and local recurrence was seen in 23% patients. 77% of the patients were followed up for a median follow-up period of 54.3 months (range 6- 156 months) and 23% died of disease after a median time of 62.7 months (7 months-10 years).

Wide local excision is the treatment of choice with 2 or 3cm bony margins[6]. Currently most clinicians treat ameloblastic carcinoma with surgery and postoperative radiation[7,8]. Indications for adjuvant radiation include close or positive margins, stage T3 or T4 tumours, nodal involvement, extra capsular extension. Radiotherapy and chemotherapy have a limited value in the management[9,10]. However these methods should be considered when there is a locally advanced or metastatic disease that cannot be resected.

Follow-up is essential because recurrence and metastasis to the lungs and regional lymph nodes have been reported in the literature[11]. Several instances of recurrence have been reported in literature, and thus a long follow-up is necessary for these tumours.

IV. Conclusion

Ameloblastic carcinoma of maxilla is a rare head and neck cancer and can be cured with multimodality treatment and good followup.

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Fig 1: 4X5cm ulcero-proliferative swelling in the left upper alveolus with swelling in the left maxillary region

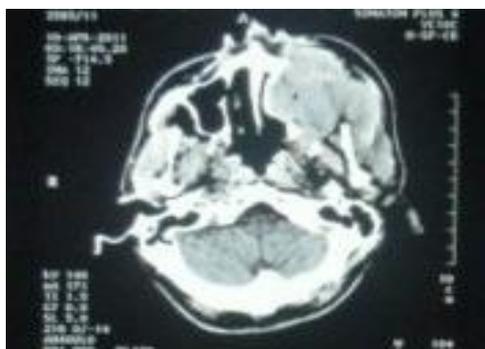


Fig 2 : CT scan showing a large hypodense mass on left upper alveolus with erosion of maxilla, pterygoid, and zygomatic bone with extension into infratemporal region

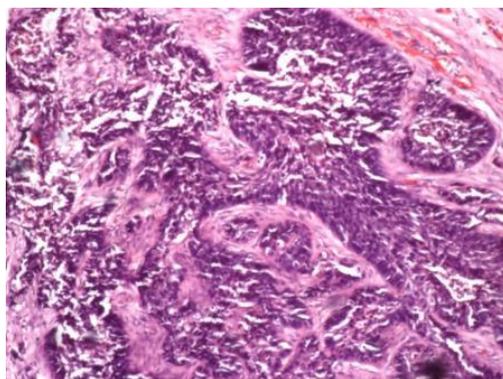


Fig 3:H&E 100X island of ameloblastic cells with peripheral palisading and central stellate reticulum.

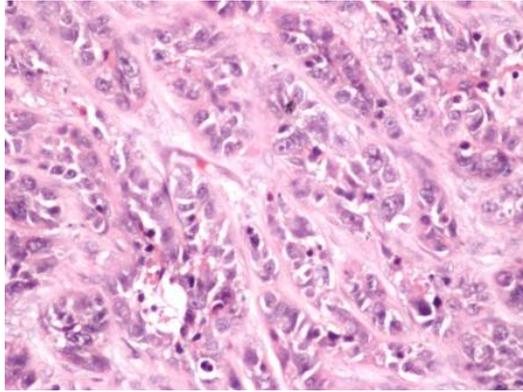


Fig 4 :H&E 400X lesion proper composed of polygonal cells arranged in nests, cords and trabeculae infiltrating surrounding structures)

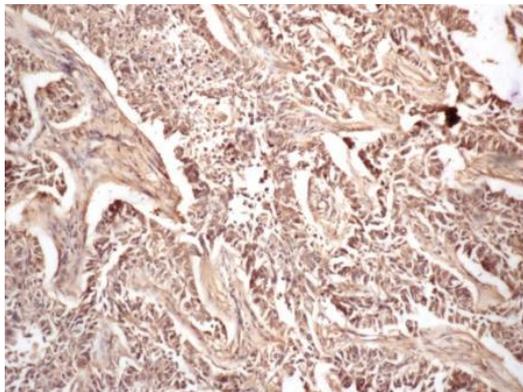


Fig 5: 100X IHC with calretinin, neoplastic cells showing cytoplasmic positivity

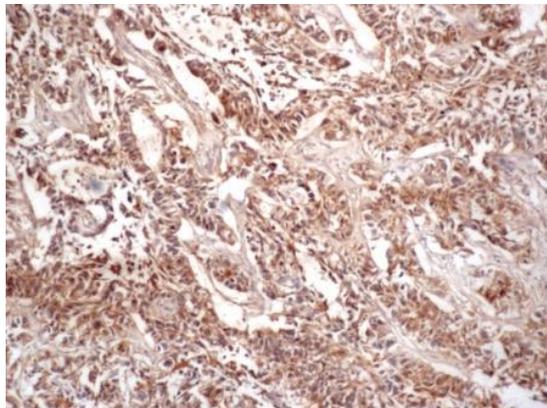


Fig 6:100X IHC with cytokeratin, neoplastic cells showing cytoplasmic positivity

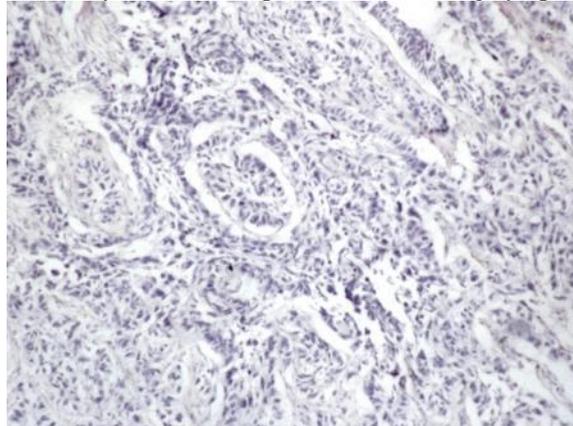


Fig 7:100X IHC with S100, neoplastic cells showing cytoplasmic negativity)