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Case Report

AMELOBLASTIC CARCINOMA PRESENTING CLINICALLY AND HISTOLOGICALLY AS PLEXIFORM AMELOBLASTOMA – A RARE AND INTERESTING CASE REPORT AND REVIEW

***Sasidhar Reddy V., Maheshwar G., Karthik Reddy and Abhishek Reddy**
Department of Oral and Maxillofacial Surgery, Meghna Institute of Dental Sciences, Mallaram, Nizamabad, Andhra Pradesh, INDIA
**Author for Correspondence*

ABSTRACT
Ameloblastic carcinoma is an extremely rare, aggressive malignant epithelial odontogenic tumor with characteristic histologic features and behaviour that dictates a more aggressive surgical approach than that of ameloblastoma. However, reliable evidence of its biologic activity is currently unavailable and our understanding of the histologic features of ameloblastic carcinoma is somewhat vague due to the scarcity of well-documented cases. Because the lesion is usually found unexpectedly after an incisional biopsy or the removal of a cyst, a guide to differential diagnosis is not usually useful. The identifying features of ameloblastic carcinoma must be known and recognized by dental practitioners. Wide local excision is the treatment of choice with selective regional lymph node dissection with close post operative periodic follow up. There are no definitive recommendations regarding radiotherapy and chemotherapy reported in literature. This article discusses a case of ameloblastic carcinoma which presented clinically and histologically as plexiform ameloblastoma.

INTRODUCTION
In 1983, Shafer introduced the term ameloblastic carcinoma to describe ameloblastomas in which there had been histologic malignant transformation. Ameloblastic carcinoma is an extremely rare, aggressive malignant epithelial odontogenic tumor with characteristic histologic features and behaviour that dictates a more aggressive surgical approach than that of ameloblastoma. However, reliable evidence of its biologic activity is currently unavailable and our understanding of the histologic features of ameloblastic carcinoma is somewhat vague due to the scarcity of well-documented cases. It is seen to occur primarily in the posterior mandible in a wide range of age groups with mean age of 30.1 years and with no sex or race predilection reported (Corio *et al.,* 1987). The most common sign described has been swelling, although others include associated pain, rapid growth, trismus and dysphonia. Involvement of the maxilla by ameloblastic carcinoma seems to be less frequent than that of the mandible (Slootweg and Muller, 1984; Lee *et al.,* 1990; McClatchey *et al.,* 1989; Andersen and Bang, 1986; Daramola *et al.,* 1980). It may present as a cystic lesion with benign clinical features or as a large tissue mass with ulceration, significant bone resorption and mobility of teeth. Because the lesion is usually found unexpectedly after an incisional biopsy or the removal of a cyst, a guide to differential diagnosis is not usually useful. The identifying features of ameloblastic carcinoma must be known and recognized by dental practitioners. The tumour cells resemble the cells seen in ameloblastoma, but they show cytologic atypia, lacking the characteristic arrangement seen in ameloblastoma. The clinical course of ameloblastic carcinoma is typically aggressive, with extensive local destruction, lymph node involvement and metastasis to various sites. Wide local excision is the treatment of choice with selective regional lymph node dissection (Dhir *et al.,* 2003) with close post operative periodic follow up. There are no definitive recommendations regarding radiotherapy and chemotherapy reported in literature. This article discusses a case of ameloblastic carcinoma which presented clinically and histologically as plexiform ameloblastoma.

CASES
A 40 year old female patient reported to our centre with a complaint of painless swelling in the lower left part of face causing asymmetry and associated with numbness of lower lip since six months (figure 1).

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Examination revealed a hard, non tender and non fluctuant swelling over lower half of left side of the face approximately 10cm × 5cm in size. Swelling was extending from corner of the mouth to tragus of the ear. Intraorally left lower buccal sulcus was obliterated by a hard swelling. Radiographic examination revealed a large multilocular radiolucency extending from left lateral canine to left third molar region involving an impacted second molar present at the lower border of mandible (figure 2). Computed tomography images show a lesion approximately of size 10 cm antero posteriorly and 6 cm mediolaterally involving both cortical plates of the mandible. Lower border of mandible is also seen to be involved in CT scan and lingual cortical perforation is noticed (figure 3). Aspiration yielded a straw colored fluid typical of a dentigerous cyst or ameloblastoma. A differential diagnosis of ameloblastoma, odontogenic keratocyst, and dentigerous cyst was made and incisional biopsy was performed under local anesthesia. Histopathological examination revealed presence of a stellate, reticulum-type odontogenic epithelium arranged in anastomosing sheets and cords as a tangled network, enclosing cysts of various sizes suggestive of plexiform ameloblastoma. Owing to the protocol for treatment of ameloblastoma enbloc resection of mandible with minimum of 1cm normal bony margins was planned. Segmental resection of the mandible was performed intraorally under general anesthesia and a mandible reconstruction plate was adapted and fixed to the mandible. The resected specimen (figure 4) was sent for histopathology; microscopic examination revealed epithelial islands and strands in connective tissue stoma with areas of peripheral palisading columnar cells with a vacuolated cytoplasm and reverses polarized nuclei. In some areas cytological malignancy, nuclear pleomorphism and a high degree of mitotic rate were observed. Diagnosis of ameloblastic carcinoma was based on presence of ameloblastomatous differentiation of tumor cells. The patient was sent for radiotherapy after removal of the reconstruction plate. One year follow-up showed excellent healing and no signs of recurrence of the lesion.

DISCUSSION

Ameloblastoma is a benign locally aggressive odontogenic tumour, believed to originate from the remnants of dental epithelium. This tumour accounts for 1% of all cysts and tumours of the jaw and is the second most common odontogenic tumour after odontoma. The question of malignancy in ameloblastoma has been the subject of considerable discussion and controversy for many years. There can be little argument that an ameloblastoma that metastasizes is malignant, even if the tumour shows benign histological features. In other instances, ameloblastoma has been considered to be malignant on the basis of an aggressive clinical course in the absence of metastasis. These lesions often show unusual or atypical histological features (Regezi *et al.*, 1978).

Carcinomas derived from ameloblastoma have been designated by a variety of terms, including malignant ameloblastoma, ameloblastic carcinoma, metastatic ameloblastoma and primary intra-alveolar epidermoid carcinoma. Clinically, ameloblastic carcinomas are more aggressive than most typical ameloblastomas. Perforation of the cortical plate, extension into surrounding soft tissue, numerous recurrent lesions and metastasis, usually to cervical lymph nodes, can be associated with ameloblastic carcinomas. This tumour, developing within bone, probably originates from odontogenic epithelial remnants (Corio *et al.*, 1987). Although, the primary intra-alveolar carcinoma and the ameloblastic carcinoma exhibit some clinical differences, their histologic features are similar enough to suggest a histogenetic relation. It is possible, then, that the primary intra-alveolar carcinoma may represent simply a less differentiated, usually nonkeratinizing form of ameloblastic carcinoma, both lesions being derived from odontogenic remnants (Corio *et al.*, 1987).

Two types of typical ameloblastoma must be considered in the differential diagnosis of ameloblastic carcinoma, the acanthomatous ameloblastoma and the so-called kerato-ameloblastoma containing prominent keratinizing cysts that may cause some alarm and distract the pathologist from the otherwise ameloblastomatous feature.

An additional consideration in the differential diagnosis is the squamous cell carcinoma arising in the lining of an odontogenic cyst (Dhir *et al.*, 2003; Regezi *et al.*, 1978).

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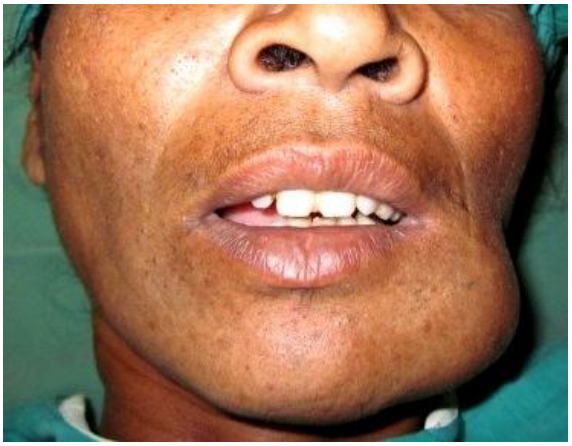

Figure 1: Clinical picture showing facial assymtery

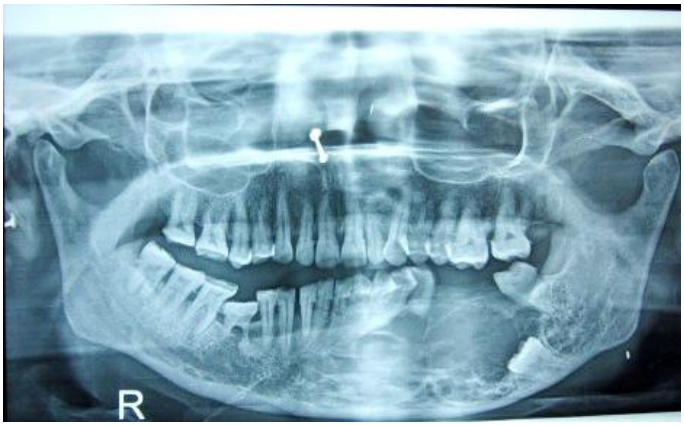

Figure 2: Orthopantomogram showing large multilocular lesion

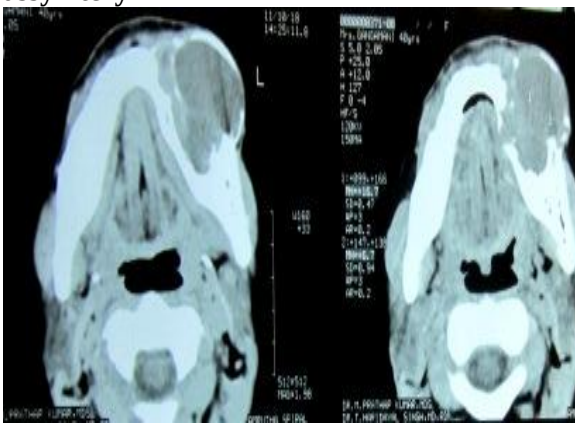

Figure 3: CT scan showing lingual cortical perforation

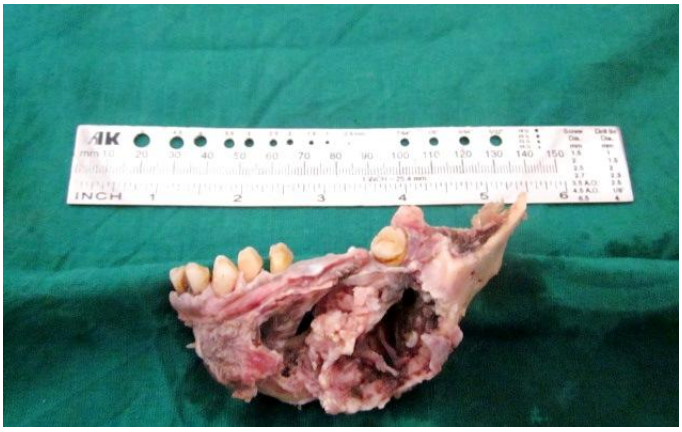

Figure 4: Resected specimen

Histologically, this lesion tends to more closely resemble oral squamous cell carcinoma than what is described for ameloblastic carcinoma. Whether ameloblastoma may transform biologically and histologically from a classic ameloblastoma to a malignant lesion remain controversial. Nevertheless, it is important that, in the future, these lesions be accurately identified, differentiated from malignant ameloblastoma and followed so that their natural history and prognosis can be further defined.

Conclusion

Ameloblastic carcinoma is an aggressive odontogenic tumour that requires aggressive surgical treatment. The clinical and biological differences between conventional ameloblastoma and ameloblastic carcinoma are significant and can be useful to distinguish between the two entities when the pathological diagnosis is not certain. It is reasonable to assume that this case illustrates the malignant portion in the spectrum of ameloblastomas. Cases of ameloblastoma should thus be studied carefully, correlating their histologic pattern with biologic behaviour to detect subtle changes in histology that may predict aggressive behaviour. The treatment of ameloblastic carcinoma is controversial, but the recommended surgical treatment usually requires jaw resection with 2- to 3-cm bony margins and consideration of contiguous neck dissection, both prophylactic and therapeutic. Documented case reports with meaningful follow-up are rare. Meticulous follow-up is essential because recurrence and metastasis in the lung and regional lymph nodes have been reported. Presurgical radiation therapy has been suggested to decrease tumour

size, but chemotherapy is as yet unproven. Reconstruction of the post-resection defect may proceed as one would normally expect following any head or neck carcinoma resection. Sufficient time should be allotted before reconstruction because of potential tumour recurrence.

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