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Ameloblastic carcinoma of the mandible: A rare entity

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Abstract

Ameloblastic carcinoma (AC) is a very rare malignant odontogenic epithelial tumor of the jaws that has histological features of both ameloblastoma and carcinoma. It occurs primarily in the mandible in a wide range of age groups without any gender predilection. The clinical presentation of AC is variable; it may present as a cystic lesion with benign clinical features or as an aggressive tissue mass with extensive local destruction. Surgical En Bloc excision is the treatment of choice. Radiotherapy and chemotherapy seem to be of limited value for the treatment of AC. Long-term follow-up with periodic reassessment is mandatory. We are presenting a case report of AC in a very young patient, with literature review.

Keywords: Ameloblastoma, Malignant ameloblastoma, Ameloblastic carcinoma, Odontogenic tumors.

Introduction

Ameloblastic Carcinoma (AC) is an extremely rare malignant odontogenic epithelial tumor of the maxillofacial skeleton[(1)] reported only 92 cases till 1984 to 2012 in the scientific literature.[(2)] AC is more predilection in males than females with a ratio 2.3:1 without any age predilection ranges from 7 to 91 years.[(2)] It occurs more significantly in mandible than maxilla with a ratio of 5:1, the most common site of occurrence is the mandibular molar region[(3)] and only one case reported in anterior jaw region.[(4)] Malignant variants of the ameloblastoma are extremely rare.

The term ameloblastic carcinoma was introduced by Elzay.[(5)] AC defined as a rare odontogenic malignancy that combines the histological features of ameloblastoma with cytological atypia, even in the absence of metastases.[(6)] According to the WHO criteria may arise as a result of malignant changes in a preexisting benign ameloblastoma (carcinoma ex-ameloblastoma, secondary type) or may develop *de novo* as a primary AC not preceded by a simple ameloblastoma.[(7)] Clinically, AC may arise in the form of a cystic lesion with benign clinical features or as an aggressive in nature with large tissue mass. A painful swelling with rapid growth is the commonest clinical sign, bone destruction and trismus may also be presenting symptoms.[(1,4,6)] In this article we report one such rare case of AC in young male patient affecting left side of the jaw.

Case Report

An 18 years old male patient reported to the department of oral medicine and radiology, with a chief complaint of swelling in the mouth on left side of the lower jaw since 8 months and complains of mild pain during mastication and difficulty in mouth opening, since last 3 months. (Fig. 1A) He was asymptomatic 8 months earlier and noticed a painless swelling on left side of the lower jaw. But, after 3 months noticed rapid growth of the swelling with ulceration over the swelling and also noticed loosening of teeth at the affected region and underwent extraction 1-month back. The patient realized the swelling grew in size even after extraction of the tooth. Patient gives a history of chewing Gutka 2-4 packets daily, since 3 years and usually places the quid against the left cheek.

On extra-oral examination a gross asymmetry of the face with a single, diffused swelling at the left cheek region which was approximately 6 cm x 8 cm was observed. (Fig. 1A) The swelling was extending anterio-posteriorly from the left corner of the mouth to left angle of the mandible and left ear and superio-inferiorly from the ala-tragus line to 1 cm below the left lower border of the mandible. The skin over the swelling was normal in colour and texture. On palpation it was mild tender, firm in consistency and fixed to the underlying structures, with no local rise of temperature. Left submandibular lymphnodes were palpable, measuring approximately 1x1 cm in size, which were non-tender and movable. Intraorally, a single, well-defined, ulcerative lesion which was approximately 4 cm x 6 cm in size was present at the mandibular left gingivo-buccal complex. It was extending anterio-posteriorly from the distal surface of 34 to retromolar area with expansion of buccal and lingual cortical plates, medially 1 cm away from the normal teeth bearing area, on to the floor of the mouth on left side by displacing the teeth (35 and 36) and laterally 2.5 cm away from the normal teeth bearing area and extending deep into the adjacent vestibule and leading to the obliteration of the vestibule and subsequently giving rise the extra oral swelling. The

edges are everted and the surface covered with pseudomembranous slough. Displacement and mobility of 35 and 36 noted. The lesion was mild tender, firm to hard in consistency and fixed to the underlying structures. (Fig. 1B)

Fig. 1: Extra oral swelling with asymmetry on left side of the face (A), Intra oral view shows an ulcerative lesion (B), Post-Surgical Photograph after one year (C, D)

Base line investigations are not contributory. Radiographical investigation such as Mandibular occlusal radiograph (Fig. 2) revealed expansion and thinning of buccal and lingual cortical plates. Slight resorption of the apices of 35 and 36 noted. Orthopantomogram (Fig. 3A) revealed a well-defined, multilocular radiolucency with fine, thin, curved septae was noticed. It was extending from the apical region of 34, posterio-superiorly upto coronoid process and involving the mandibular whole left body, ramus and coronoid process just by sparing the left condyle. Only a thin rim of inferior border of mandible noted with complete destruction of the alveolar bone at the affected area. Displacement of 34, 35 and unerupted 38 which is pushed into the ramus noted. CT scan revealed a lytic lesion involving the entire mandibular left body, ramus and coronoid process with thinning of cortical plates cortex and with cortical breach at multiple sites just by sparing the left condyle.

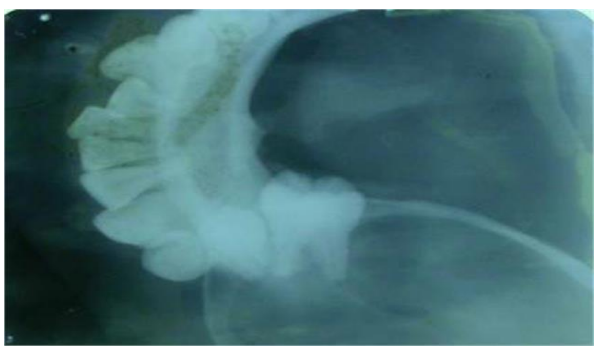

Fig. 2: Mandibular occlusal radiograph showed buccal and lingual cortical plates expansion

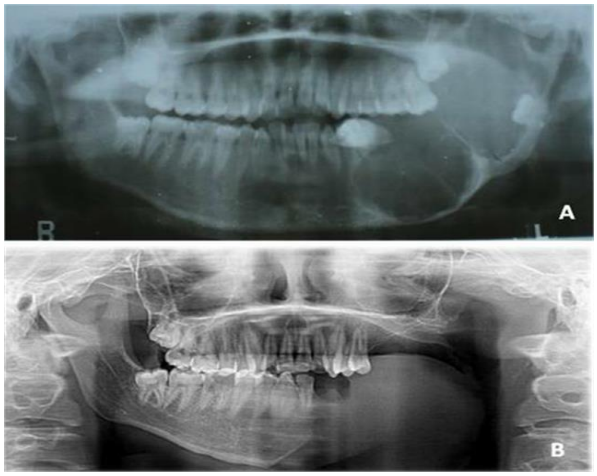

Fig. 3: Orthopantomogram showed a well-defined, multilocular radiolucency with curved septae (A), Post-Surgical pantomograph after one year (B)

Incisional biopsy of the lesion was done (Fig. 4 A & B) and histopathological examination revealed hyperplastic parakeratinised stratified squamous epithelium partially covering the underlying tissue shows large interconnecting strands of epithelial cells like cells which are loosely arranged stellate reticulum like cells, surrounding by columnar ameloblast like cells. Few areas shows premature cystic degeneration and large focal areas of these strands are infiltrated with chronic inflammatory cells. One focal area contains compactly arranged epitheloid cells, which shows hyperchromatism, nucleus and cellular pleomorphism, mitotic figures exhibiting malignant features suggestive of Ameloblastic Carcinoma. (Fig. 5 A & B) Based on the history, clinical, radiographical and histopathology examination final diagnosis of Ameloblastic Carcinoma was made. The patient was surgically treated with hemi mandibulectomy. The patient is under regular follow-up and no metastases reported during the 1-year follow-up period. (Fig. 1C, D & 3B)

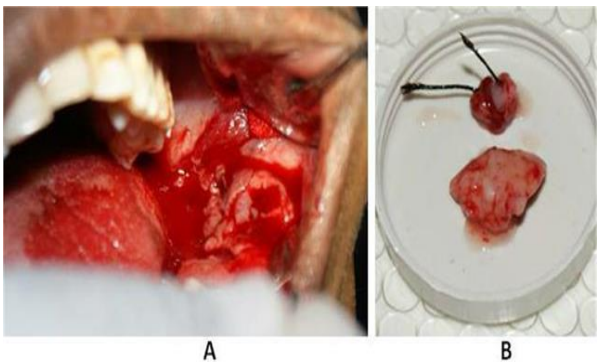

Fig. 4: Incisional biopsy of the lesion was done (A), Specimen (B)

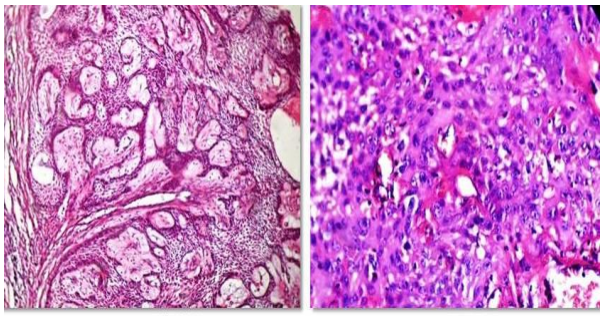

Fig. 5: Histopathological Photographs

Discussion

Ameloblastomas are tumors of odontogenic epithelium, which develops from remnants of dental lamina, enamel organ and from the epithelial lining of an odontogenic cyst or from the basal cells of the oral mucosa. Malignancy in ameloblastomas has been controversial for many years, in part because of its rarity, complicated by the confusion in terminology. 'Malignant Ameloblastoma' is the term referred to those lesions which metastasize inspite of their benign histopathological feature. 'Ameloblastic carcinoma' (AC) is the term applied only for an ameloblastoma with a malignant morphologic features, with or without presence of metastasis.[4] Metastases from ameloblastomas are most often found in the lungs and cervical lymph nodes are the second most common site for metastasis, and rarely spread to vertebrae, other bones and viscera.[8]

The revised histologic classification by WHO in 2005, based on their origin AC is classified as primary type and secondary type. Primary types are those arising *de novo* whereas the secondary type arises from a pre-existing ameloblastoma. The exact mechanism of malignant transformation of secondary type of AC is presently unknown due to its extremely rare occurrence with limited number of cases reported in the literature.[9] Considering the patient's history and absence of metastasis the present case is assumed to be secondary type AC.

AC is more commonly affecting the males with variable age groups ranges from 7 to 91 years.[2] It occurs more significantly in mandible than maxilla with a ratio of 5:1, the most common site of occurrence is the mandibular posterior region[3] the present case was consistent with literature. Clinically, AC's are more aggressive and destructive to adjacent tissues than typical ameloblastomas. The most common clinical sign is swelling associated with pain, rapid growth, trismus and dysphonia[1,6] as reported in the present case.

Radiographically, AC and ameloblastoma has the same features such as a well-defined scalloped border with cortication, the septa are robust and thick and these lesions are unilocular or mostly multilocular giving the appearance of a honeycomb or soap-bubble pattern.[1] The affected teeth show displacement, root resorption, loss of lamina dura and displace normal anatomic boundaries such as the floor of the nose, maxillary sinus and mandibular canal. But the differentiating radiographical feature with ameloblastoma is AC sometimes it represents some focal radiopacities apparently reflecting dystrophic calcifications.[1,10] as in the present case except dystrophic calcification consistent with the literature.

Histologically, AC can be considered a form of ameloblastoma that has lost most of its recognizable microscopic features. Despite areas or foci that resemble ameloblastoma, AC shows changed patterns and cytological features. The presence of sheets, islands, or trabeculae of epithelium and the absence or rare presence of stellate reticulum-like areas should alert the pathologist to the possibility of AC. Round to spindle-shaped epithelial cells with little or no differentiation toward the columnar cells of ameloblastoma further suggest this malignant process. Other features of malignancy include hyperchromatism, large or atypical nuclei, increased mitotic index, necrosis, and calcification, and particularly neural and vascular invasion. Calcifications are rare in ameloblastoma, so the presence of calcifications should be carefully evaluated. The presence of many clear cells (>15% of tumor cells) strongly suggests an AC.[11] In the present case, there was no evidence of regional or distant metastasis but there was histological evidence of typical ameloblastic areas and focal area of compactly arranged epitheloid cells, which shows hyperchromatism, nucleus and cellular pleomorphism, mitotic figures exhibiting malignant features. Although we could not ascertain unequivocally whether AC in our patient developed de novo or from a pre-existing ameloblastoma, we believe the former might be the most likely due to the aggressive behaviour of the lesion within short duration in a very young patient. Although, focal area showing malignant features in a small piece of submitted tissue, tissue from multiple sites or a large piece from representative area might have confirmed the more consistent cytological features and changed patterns of AC.

A wide surgical excision is the treatment of choice. En bloc removal with covering 2-3 cm normal bony margins has been advocated for disease free survival. This method has resulted in local recurrence rates of less than 15%.[12] Cervical lymph node dissection should also be considered when there is obvious lymphadenopathy. Radiotherapy and chemotherapy appear to be of limited value; however, these methods should be considered when there is a locally advanced or metastatic disease that is not amenable to surgical resection.[13] But there is controversy regarding the radiation therapy, as it brings the risk of osseous complications, in the form of osteonecrosis and induced sarcoma.[12]

ACs can recur locally 0.5–11 years after definitive therapy.[14] The prognosis is dominated by the risk of local recurrence, including after a long relapse, and by distant metastases.[3] Distant metastasis is usually fatal and the most common site for a distant metastasis is the lung, followed by bone, liver and brain.[12,14] Long-term follow-up with periodic reassessment is mandatory. Systematic assessment of the chest through periodic imaging is recommended further prevention of recurrent lesion and to improve patient prognosis.[3]

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