Case Report

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A RARE, AGGRESSIVE TUMOR OF THE MANDIBLE: CASE REPORT AND REVIEW

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ABSTRACT:

This paper aims to highlight a case of malignant tumor in a 23 year old male patient which is of Odontogenic origin, rare in occurrence and presented with a rapid, aggressive growth phase causing local destruction. Clinically, it is more common in the mandible and histologically, it is similar to ameloblastoma in addition to the presence of prominent cytological atypia and mitotic activity. The main as well as prominent histological features include hypercellularity, high mitotic index, lack of differentiation, vascular and neural invasion. Lymph node involvement with metastasis to various sites (frequently the lung) has also been reported and presented.

Key words: odontogenic, rare, rapid, destruction, ameloblastoma, neural invasion.

INTRODUCTION:

Tumors of the lower oro-facial region may be benign or malignant. Most frequently seen primary malignant lesions include soft tissue and hard connective tissue sarcomas, carcinomas of the salivary glands, especially squamous cell carcinomas (in 90% of the cases) and melanomas.[1-6] Benign lesions may have odontogenic or non-odontogenic origin and if left untreated, some of the lesions will lead to extensive tissue destruction and deformity whereas others will interfere with mastication and will become secondarily infected following masticatory trauma.[7-11]

CASE DETAIL:

A 23 year old male patient reported to the Department of Oral Medicine and Radiology, with a chief complaint of a painless growth over the right lower back tooth region for the past one week. Patient gave history of interference of that growth during chewing food and closing mouth. There was no associated pain, dysphagia, trismus, dysphonia, fever, chills or loss of weight and he didn't give any history of chewing habits. Patient was subjected to biopsy and FNAC of that

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region 1 week back in a private hospital whose report stated that the FNAC was non diagnostic and the histopathology report stated that it was suggestive of *well differentiated squamous cell carcinoma* in relation to right mandibular posterior region.

At present, the intra oral examination revealed that there was presence of an ulcero-proliferative growth in the distal aspect of 47 that extended upto 1cm anterior to the retromolar region. It was covered by pseudomembranous slough in the superior aspect, and the surface of the growth was irregular and had "cauliflower like" appearance with associated erythematous regions and it was tender, rough, and was present with an indurated base. No paresthesia was elicited. There were also no palpable lymph nodes present. Based on the findings obtained, a provisional diagnosis of a *Malignant growth* in relation to the right mandibular posterior aspect was given.

IOPA of 46, 47 region revealed, presence of impacted 48 along with an overlying diffuse radiolucency suggestive of a cystic lesion. OPG revealed presence of a unilocular radiolucency extending from the CEJ of the mesial aspect of 48 upto the CEJ in the distal aspect of 48 that involved the ascending ramus of mandible, with a well defined sclerotic border which was suggestive of cystic lesion (Dentigerous cyst – Lateral Varient).

Later patient was advised CBCT of mandible and CT of Head and neck which revealed similar findings as in OPG. In addition, it revealed that there was no marked buccal or lingual cortical expansion and no bone perforation. MRI of head and neck revealed presence of a hypointense lytic lesion involving the posterior aspect of body and angle of mandible extending to the right retromolar trigone region and the fat plane between the adjacent soft tissue and lesion being maintained suggestive of neoplastic etiology. In addition to this there were presence of tiny lymph nodes involving level I, II, III of both sides of neck and level V of left side.

An exfoliative cytology stated inflammatory cells numerous nucleated squamous epithelial cells amongst which some exhibited nuclear hyperchromatism, anisocytosis and anisonucleosis, suggestive of Class III cytology.

Deeper biopsy section of that region revealed loosely cohesive epithelium resembling stellate reticulum with focal areas of basal cells exhibiting *Ameloblastic differentiation*. The associated fibrous cyst wall exhibited intramural ameloblastomatous islands of varying sizes with few exhibiting cystic degeneration and few with keratin pearl formation. Also, nuclear enlargement, increased nuclear cytoplasmic ratio, increased mitotic activity and focal areas of necrosis were noticed which were suggestive of ***Ameloblastic carcinoma arising from unicystic ameloblastoma.***

The treatment carried out was en bloc resection in the right side of mandible along with supra omohyoid neck dissection involving lymph nodes of level I

and II. Post operative follow revealed absence of recurrence.

DISCUSSION:

Odontogenic malignancies are very uncommon and their diagnosis is based on clinicoradiographic findings and subjective histopathological evaluation. To be regarded as odontogenic, the tumor must arise in the gingival or in the jaws (table: 1).

Malignant ameloblastomas are sub classified into four distinct entities namely

1. Metastasizing ameloblastoma
2. Ameloblastic carcinoma – primary type
3. Ameloblastic carcinoma-secondary type (dedifferentiated), intraosseous
4. Ameloblastic carcinoma-secondary type (dedifferentiated),peripheral

Metastasizing ameloblastoma is differentiated from ameloblastic carcinoma by its benign histological appearance despite its ability to metastasize.

"Primary Ameloblastic carcinoma" has been classified recently by the World Health Organization (WHO) as a tumor that demonstrates the morphological features of ameloblastoma with atypia, regardless of the presence or absence of metastasis. [7] Elzay and Slootweg and Müller use the term Ameloblastic carcinoma to convey the presence of cytologic features of malignancy.

The World Health Organization defines ameloblastic carcinoma as *"a rare primary odontogenic malignancy that combines the histological features of ameloblastoma with cytological atypia*". It is a malignant epithelial proliferation, which is associated with an ameloblastoma (carcinoma ex ameloblastoma) or histologically resembles an ameloblastoma (de novo Ameloblastic carcinoma). They most commonly occur in the mandible (2.14:1), mean age of occurrence is a range from 15 to 84 yrs. It also has a male predilection with male to female ratio to be 1.4:1.

Clinical presentation is variable such as cystic lesion with benign clinical features or large tissue mass with ulceration, bone resorption and tooth mobility. In our case, 47 was not mobile and also there was no appreciable bone resorption.

Radiologically, they appear as either unilocular or multilocular radiolucent pattern with cortical destruction due to their aggressive nature.

Histologically, the microscopic pattern of ameloblastoma along with malignant cytological features is present. These include- increased nuclear to cytoplasmic ratio, nuclear hyperchromatism, and presence of mitosis. Necrosis in tumor islands and dystrophic calcifications may also be present.

In our case, the microscopic features were much relevant and apt for the final diagnosis of ameloblastic carcinoma.

Differential diagnosis might include- *Primary intra alveolar carcinoma, squamous odontogenic tumor, metastatic carcinoma of jaw, central high grade mucoepidermoid carcinoma, acanthomatous ameloblastoma, kerato ameloblastoma*. Hence recognition of typical ameloblastic features is necessary for its diagnosis.

Due to their aggressive clinical course, with perforation of cortical plates of the jaw and extension into the adjacent soft tissues, surgical resection along with radical neck surgery (in case of lymph node involvement) is necessary. Long term meticulous follow up is definitely necessary due to its poor prognosis.

CONCLUSION:

Ameloblastoma is a slowly growing, locally aggressive tumor and any change in its growth pattern should cause suspicion of malignancy. Therefore, complete history and general systemic examination are required to rule out any distant metastasis. Despite the fact of the biological nature of this rare tumor - clinical diagnosis, immediate histological analysis and timely treatment indeed play a major role in enhancing the quality of life of the patient.

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FIGURES:

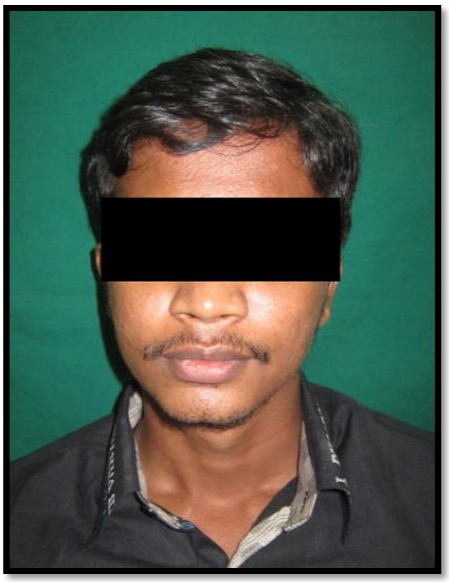

Fig 1: Profile

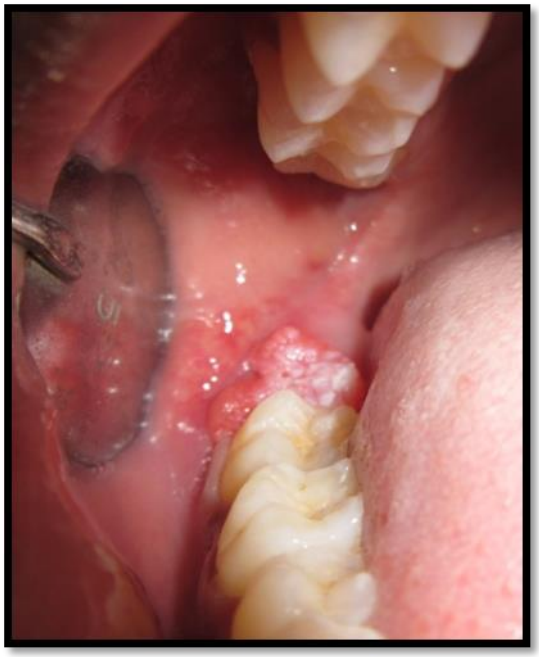

Fig 2: Intra oral picture

Fig 3: Intra oral picture depicting the “Cauliflower“ like growth

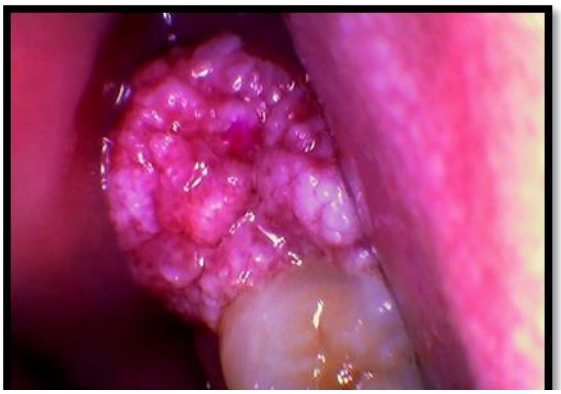

Fig 4: IOPA revealing presence of impacted 48 with radiolucency involving the crown

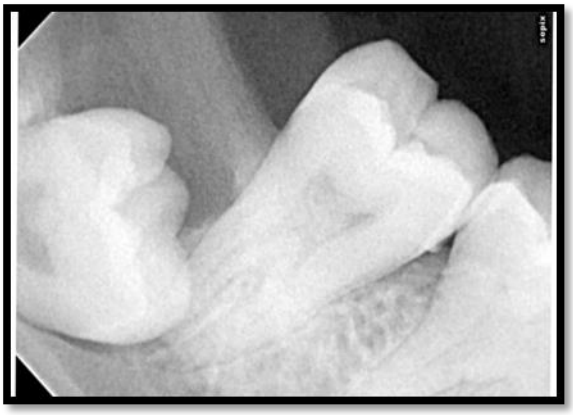

Fig 5: OPG revealing well defined radiolucency in relation to the

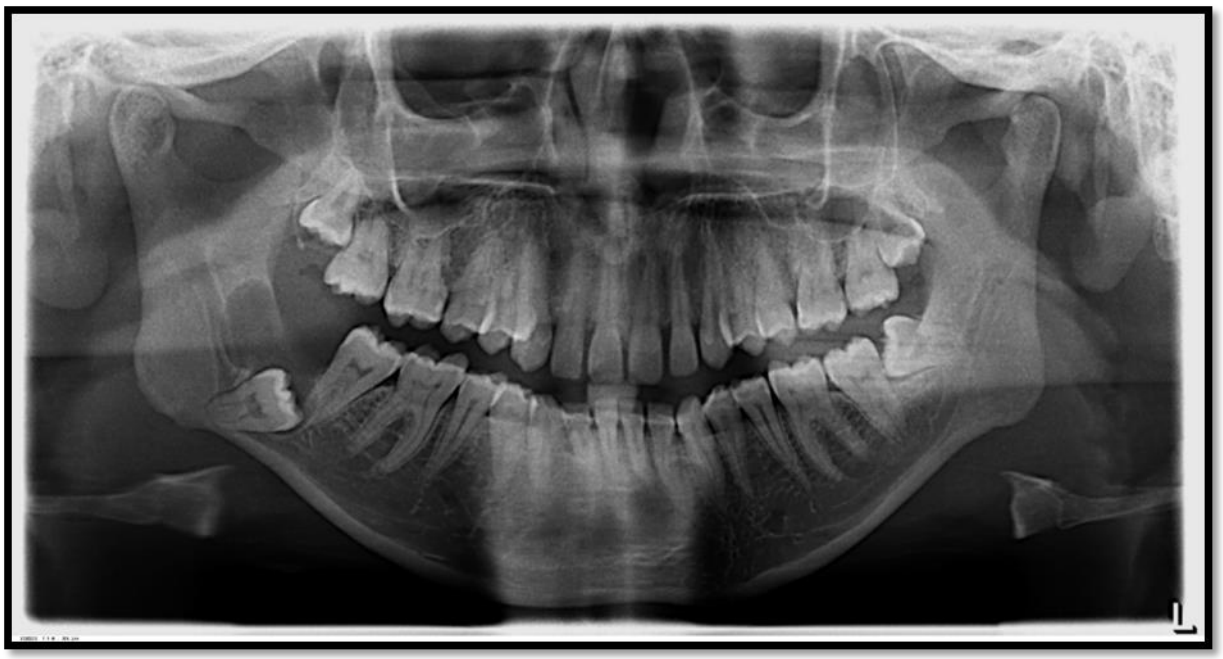

Fig 6: CBCT of mandible with 3D reconstruction

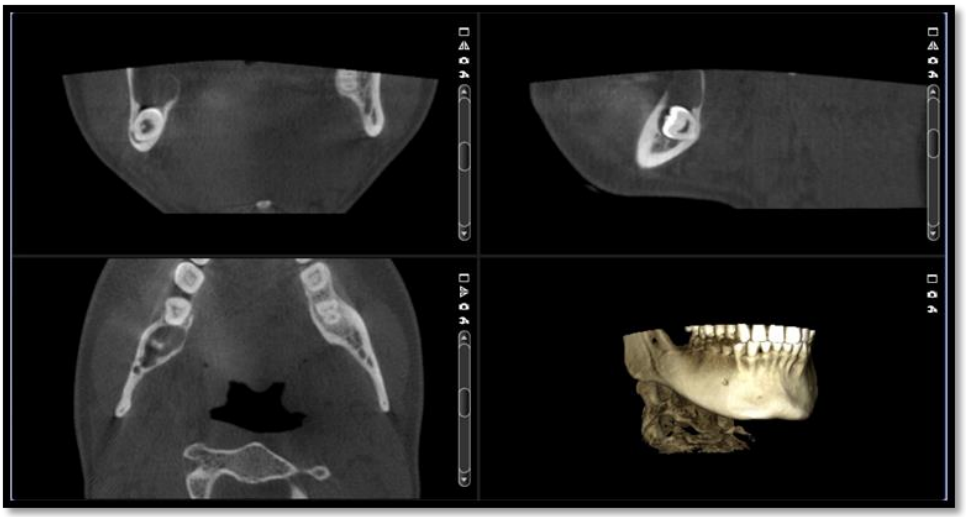

Fig 7: MRI of Head and neck

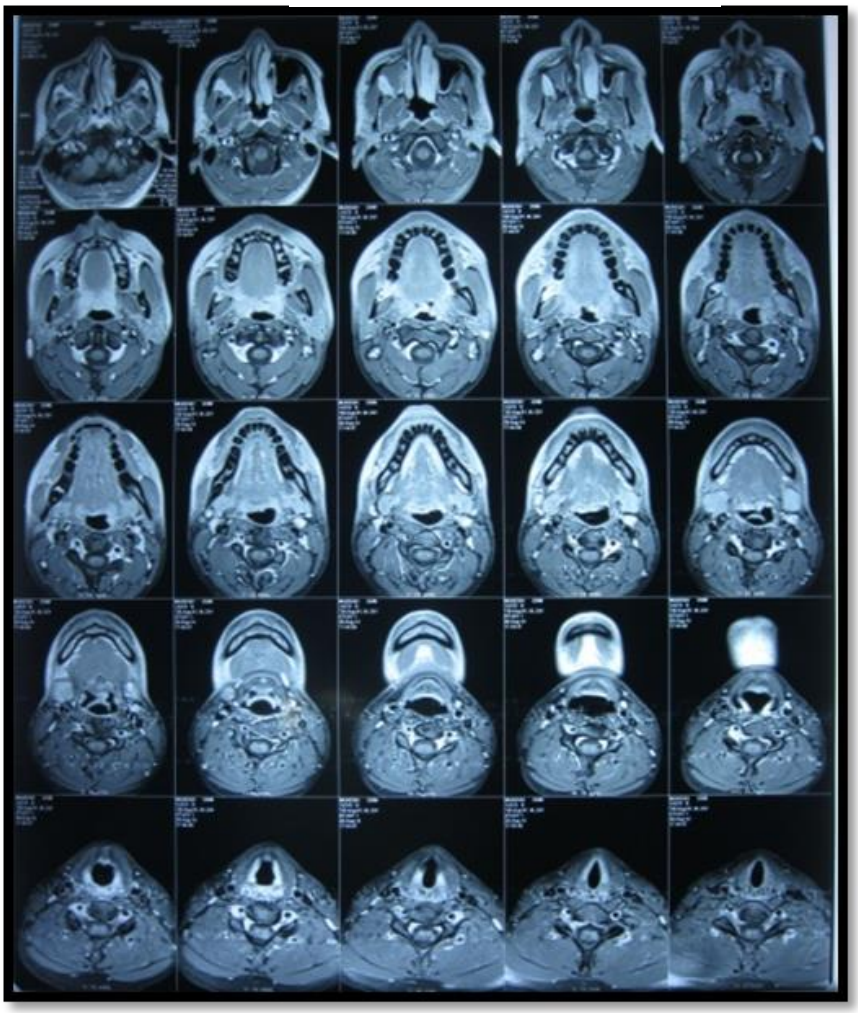

Fig 8: Biopsy specimen

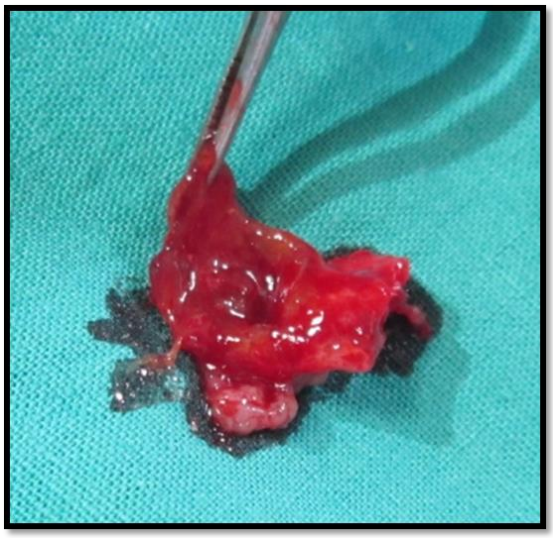

Fig 9: Histopathological

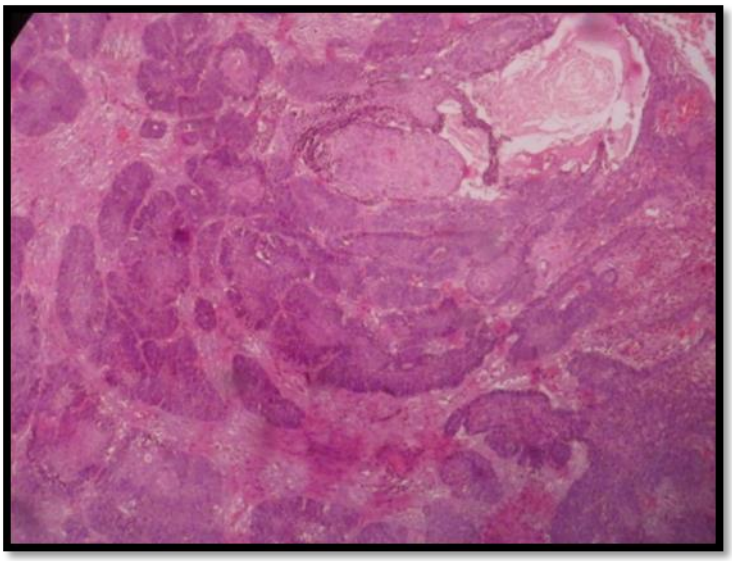

Fig 10: Histopathological

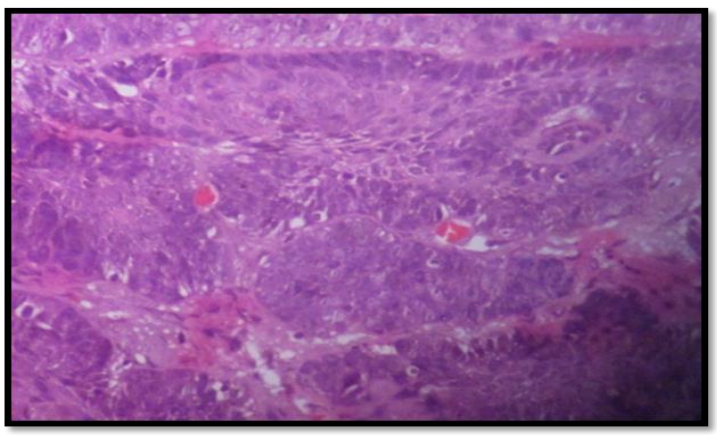

TABLE:

Table 1: Classification by Slootweg and Muller (1984)

Type 1 Primary intraosseous carcinoma ex odontogenic cyst
Type 2
(A) Malignant ameloblastoma
(B) Ameloblastic carcinoma, arising de novo, ex-ameloblastoma or ex-odontogenic cyst
Type 3 Primary intraosseous carcinoma arising de novo
(A) Non keratinizing
(B) Keratinizing