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Ameloblastic fibrosarcoma of mandible: A journey of a benign tumour to an aggressive malignancy

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Abstract
Ameloblastic fibrosarcoma (AFS) is a rare malignant odontogenic neoplasm. It is composed of a benign epithelial component and a malignant ectomesenchymal component and most frequently seen in the third and fourth decades of life. Mandible (posterior part) and maxilla is most commonly involved. We report here a case of AFS originating in the mandible of long term duration of eight years. The presentation suggests malignant transformation of benign ameloblastic fibroma in the present case. The patient was treated by surgical excision followed by radiotherapy.

Keywords: ameloblastic fibrosarcoma, mandible, odontogenic tumour, radiotherapy, surgical resection

Introduction
Odontogenic tumors may arise from epithelial, ectomesenchymal, and mesenchymal elements of the tooth-forming apparatus. Ameloblastic fibrosarcoma (AFS) is a rare malignant odontogenic neoplasm composed of benign epithelium and malignant mesenchymal component [1]. AFS was first described by Heath in 1887. AFS frequently occurs in the third and fourth decades of life. It commonly present with a painful swelling of the jaw bones and posterior part of the mandible is commonly affected. AFS affects both males and females. Although AFS was thought to be the malignant transformation of ameloblastic fibroma (AF) [2], but it may occur de-novo without any pre-existing lesion. Radiographically, the tumor shows an expansile destructive radiolucent lesion with ill-defined margins suggesting malignancy [3]. Prognosis of AFS is usually good with surgical resection by taking adequate margin followed by post-operative radiotherapy. Adjuvant treatment in AFS is necessary to prevent local recurrences. Distant metastasis is very rare but local recurrences common. We report here a case of AFS after a long term benign jaw swelling of eight years originating in the mandible. Very few cases of benign ameloblastic fibroma have been reported in the literature with a history of malignant transformation, including a recent report of malignant transformation of ameloblastic fibroma to an ameloblastic fibrosarcoma in a span of two years [4]. The purpose of this case report is to make oral surgeons aware that a long standing ameloblastic fibroma can turn into malignancy.

Case Report
A 21-year old male presented in the out - patient department of head and neck oncology of our institute with the history of swelling in the left lower alveolus for eight years duration. The swelling had rapidly increased in its size from the last two months and since then the patient was on a liquid diet. The patient was a tobacco and betel-nut chewer. On general examination, the patient was of average built without any co-morbid conditions. Intra - oral cavity examination revealed an ulcero-proliferative growth completely obstructing the opening of the mouth (Fig.1). It was extending from the right canine to left molar teeth of the lower jaw. Skin of the lower jaw was free from tumor infiltration and there were no palpable neck nodes.
Computed tomography (CT scan) of the oral cavity showed a large expansile lytic lesion involving left hemimandible, with extension to midline region. CT also showed that the floor of mouth (FOM) was indented by the mandibular lesion and there was associated thickening of left lower gingivobuccal sulcus (LGBS). Pre-operative punch biopsy was done, which revealed features of ameloblastic fibroma (AF).
The patient underwent complete excision of the mass along with a left extended segmental mandibulectomy and reconstruction was done with titanium plate (Fig. 2). The whole specimen was removed in toto. Post-operative histopathological examination showed features of ameloblastic fibrosarcoma. All the surgical cut margins were free of tumour infiltration. The final diagnosis of AFS was made after post-operative histology report. The patient was further treated by post-operative external beam radiotherapy (EBRT). The patient received 56 Gray/23 fractions and the patient could not tolerate further EBRT due to severe muco-cutaneous reactions. The patient is on follow-up for the last three years without any signs of local recurrence. The patient's mouth opening was adequate and he was taking diet normally.

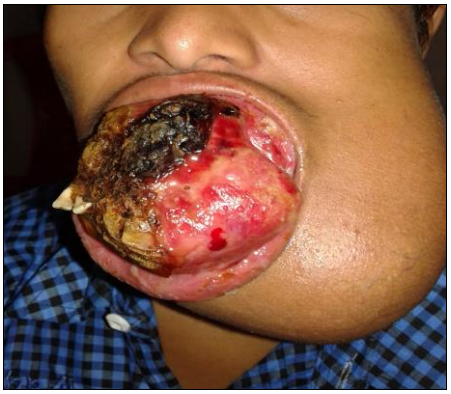

Fig 1

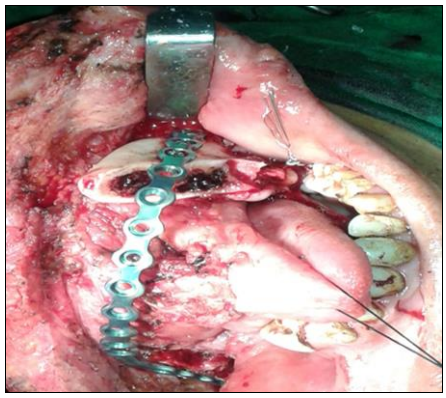

Fig 2

Discussion

Mandible is the most commonly involved bone in AFS and occurs in the third decade of life [5]. In our case also mandible was involved. AFS occurs in a wide range of age group from 3 to 83 years (mean being 27.3 years). Our patient was 21 years at the time of presentation. According to the cases reported so far, there is no predilection for either gender. However, Bregni *et al* reported a male preponderance-to-female in the ratio of 1.6:1 [6]. Our patient was male. Although AFS involves either the mandible or maxilla, but mandible is most commonly involved accounting for 79% and the maxilla in 21% of cases, and posterior part of the mandible is commonly involved [7]. In our present case, the mass was involving the left hemi-mandible just crossing the midline. However, as there was no involvement of the skin or floor of the mouth, local repair with plate could be done without necessitating repair with a pedicled or free flap. Pain and swelling of the effected side of the face are the usual presentation in AFS [7]. But in our case there was no history of associated pain and patient presented with a huge ulcero-proliferative growth occluding the oral aperture. Computerized tomography usually shows a destructive radiolucent pattern with ill defined borders suggestive of malignancy in AFS [8]. In our case, CT scan showed a large expansile lytic lesion in the mandible.

The exact pathogenesis for AFS is not known, half of the known cases states that it may be the transformation of a pre-existing ameloblastic fibroma after surgical resection. Muller *et al* reported 44% of AFS patients had a previous diagnosis of AF [9]. Thus, serial sampling of the surgical specimen of AF is helpful for early diagnosis of AFS. In our case also, pre-operative histopathology revealed AF, and serial sampling of the resected specimen revealed the pathological characteristic of AFS and the final diagnosis could be made. The long presentation history and a pre-operative biopsy of AF suggest that the present case is a malignant transformation from AF. Recently biomarkers associated with cell proliferation (Ki67, PCNA, and c-KIT) and apoptosis (BCL2) are used to overcome difficulties in the diagnosis of the tumor with low malignant potential in the mesenchymal component without considering clinical and radiographic findings [10].

Radical surgical excision is the treatment of choice once the diagnosis is confirmed followed by post operative adjuvant radiotherapy or chemotherapy to prevent recurrences. AFS is a locally aggressive disease and infiltrates the bone and adjacent surrounding soft tissue. The present patient underwent wide surgical resection followed by EBRT. The patient was free from disease at the two years following the completion of treatment without any restriction of mouth opening and hence leading a good quality of life.

Conclusion

We present this rare case of AFS with epithelial dysplasia presenting with rapidly progressing disease. In our limited experience, radical surgery followed by radiotherapy provided a good therapeutic outcome.

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