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Case Report

Ameloblastic fibrosarcoma – A rarity?

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ABSTRACT

Ameloblastic fibrosarcoma (AFS) is a rare malignant odontogenic tumour classified by the WHO (1992) as an odontogenic sarcoma and defined as "a neoplasm with a similar structure to ameloblastic fibroma (AF) but in which the ectomesenchymal component shows the features of a sarcoma." The first report of AFS was published by Heath in 1887. AFS and related lesions are less frequently diagnosed than odontogenic carcinomas. Approximately two-thirds of AFSs seem to arise de novo, but others have developed in recurrent AF, in which the ectomesenchymal cells retain their embryonic appearance and develop malignant characteristics. We report a rare case of an aggressive odontogenic neoplasm, the incisional biopsy of which showed the features of AF, while the excisional biopsy revealed the features of malignancy, suggestive of AFS.

The purpose of this report is to discuss the diagnostic difficulties, whether AFS is truly an extremely rare tumour as reported earlier and, lastly, should the treatment protocols of AFs be revised, as 44% of AFSs arise from recurrent AFs.

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Introduction

AFS is a very rare odontogenic malignant neoplasm, which is considered as the malignant counterpart of AF, wherein the epithelial component remains benign, while the mesenchymal component shows malignant cytological features.[6] Takeda et al. have suggested that there is a gradual disappearance of the epithelial component as the disease progresses and transforms into a malignancy.[12] Histologically, although there are different sub-types depending on the presence of enamel and dentin, Altini et al. have suggested that they do not differ in their biological behaviour.[1] Molecular analyses of benign tumours with malignant transformation have shown oncogenic aberrations. An alteration in the p53 gene has been reported in the malignant counterpart.[8] AFS shows male predilection, with posterior mandible as the most common site of occurrence.[13]

Case report

A 33-year-old male patient reported to the Department of Oral and Maxillofacial Surgery, with a complaint of pain and swelling on the left side of the face for the past 9 months. History revealed that the patient had noticed a painless swelling 9 months previously. He visited a general dental practitioner locally 6 months earlier where it was clinically diagnosed as a radicular cyst in relation to the decayed permanent mandibular left third molar. The permanent mandibular third molar was extracted, and enucleation of the suspected cyst was attempted under local anaesthesia but could not be completed due to profuse bleeding. Two months later, an incisional biopsy was performed at a local hospital in Hyderabad, and it was sent to the Department of Oral and Maxillofacial Pathology at our institute, where it was diagnosed as AF.

The medical history was insignificant; the patient was a non-smoker and was working in low socioeconomic conditions. Extraoral examination revealed a firm, non-tender and a diffuse swelling measuring 6×7 cm over the left side of the face extending from the angle of the mouth to the preauricular region (Figure 1). On intraoral examination, an exophytic, pedunculated lesion obliterating the buccal vestibule of the upper and lower jaws was noticed (Figure 2). The permanent third molar on the left side was missing, and the mandibular left first and second premolars and molars were mobile.

Investigations

The panoramic radiograph revealed a diffuse multilocular radiolucency extending from the mandibular premolar involving the body of the mandible, entire ramus and coronoid process on the left side. The postero-anterior view revealed the mediolateral expansion of the lesion along with the destruction of the buccal and lingual cortices. Three-dimensional computed tomography (3D CT) of the face and skull revealed an expansile lytic lesion involving the body, ramus and coronoid process of the mandible on the left side with bony septation and large predominantly hypodense soft tissue mass (Figures 3 and 4).

Based on our histopathology report, which was given as AF 4 months ago, it was surgically excised and sent for histopathological examination. The haematoxylin and eosin (H & E)–stained sections of the excised tumour revealed extremely cellular, delicate to dense, predominantly immature connective tissue (CT) stroma encasing few odontogenic epithelial islands, which revealed basal/peripheral cuboidal to columnar cells with hyperchromatic nuclei and reverse polarisation resembling ameloblasts. Polygonal cells in the centre were loosely arranged and resembled stellate reticulum-like tissue. At focal areas, squamous metaplasia of the stellate reticulum-like tissue was also evident. Loosely arranged, highly cellular CT stroma surrounding these odontogenic epithelial islands resembling primitive ectomesenchyme was noted. The cells of the ectomesenchymal component were spindle to ovoid shaped with darkly staining nucleus occupying most of the cells and scanty cytoplasm. It exhibited sarcomatous changes, which included cellular pleomorphism, nuclear hyperchromatism, altered N:C ratio and few mitotic figures (Figure 5). CT stroma in other areas revealed dense bundles of collagen fibres organised in a particular pattern with spindle-shaped fibroblasts and fibrocytes exhibiting focal areas of hyalinisation. Numerous dilated blood vessels engorged with RBCs

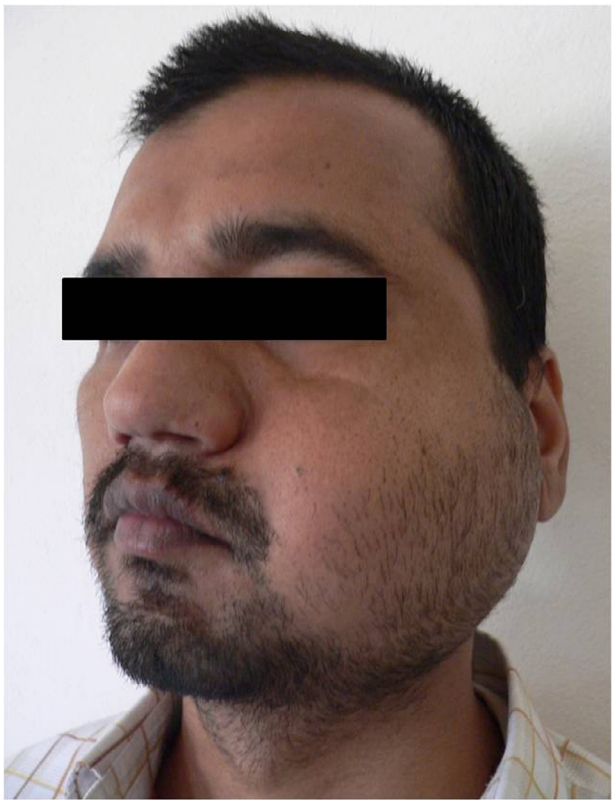

Figure 1. Diffuse swelling over the left side of face extending from the angle of mouth to the preauricular region.

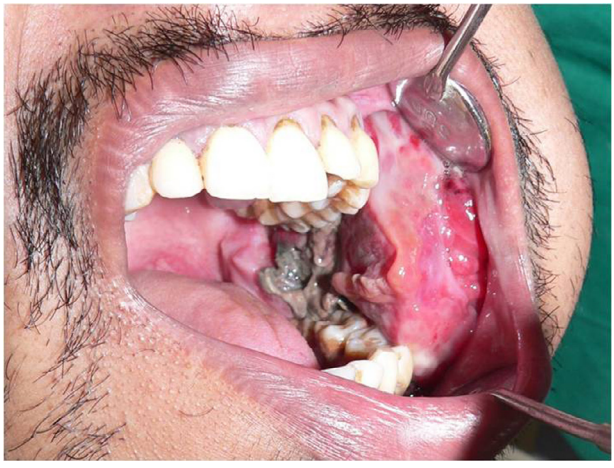

Figure 2. Exophytic, pedunculated lesion obliterating the buccal vestibules of the upper and lower jaws.

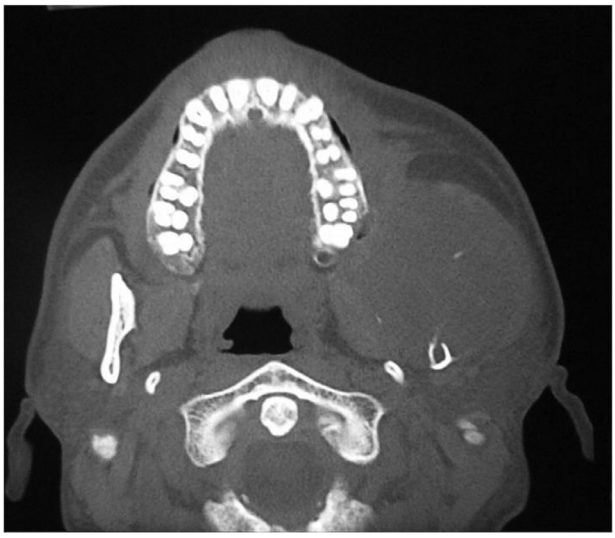

Figure 3. Axial 3D CT section showing expansile lytic lesion involving the left side of the mandible with predominantly hypodense soft tissue mass.

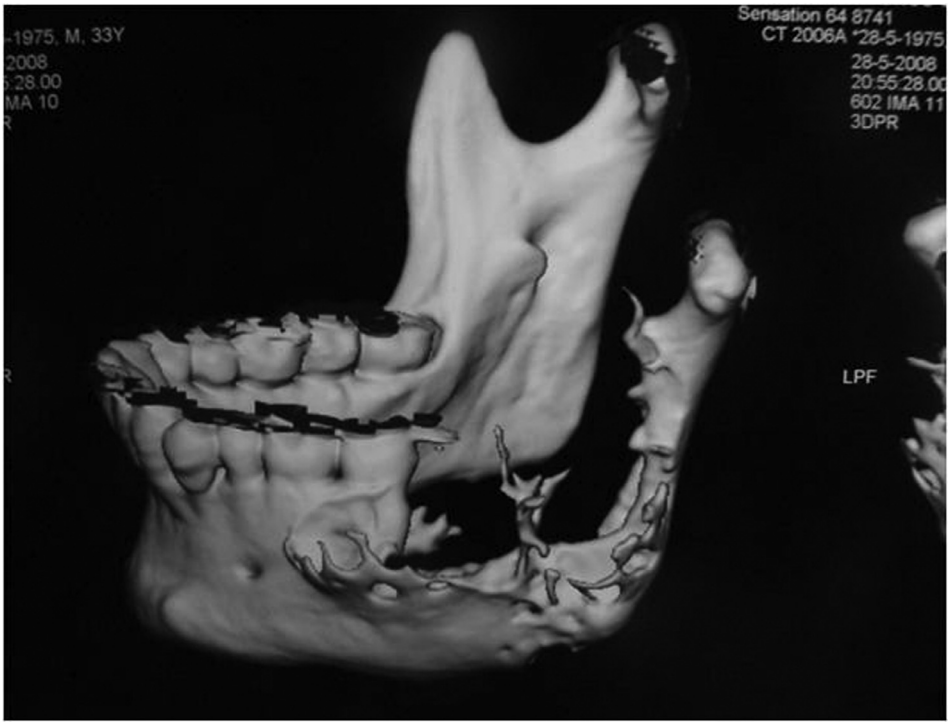

Figure 4. 3D CT image showing lytic lesion involving the body, ramus, and coronoid process of the mandible on the left side with bony septation.

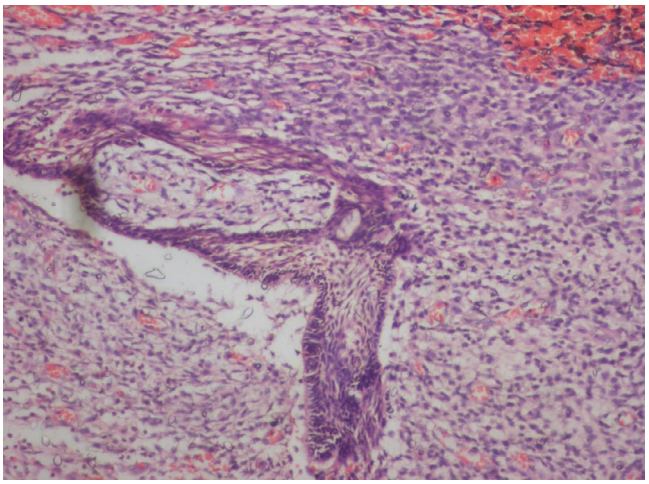

Figure 5. Photomicrograph showing loosely arranged highly cellular CT stroma with sarcomatous changes. An irregular epithelial island is seen exhibiting peripheral tall columnar and centrally polygonal cells with hyperchromatic nuclei and few mitotic figures.

along with areas of haemorrhage were evident. Focal areas also revealed homogeneously eosinophilic, irregularly shaped secretory material of varying sizes. Myxomatous changes were also seen at few places in CT stroma with stellate-shaped cells. The lesion was diagnosed to be AFS.

The case was referred and treated at MNJ Cancer Institute, Hyderabad, with chemotherapy and radiotherapy (Figure 6). There was a recurrence after 2 years, and the patient was admitted in MNJ Cancer Institute, where it was diagnosed as fibrosarcoma, treated and discharged. The patient died after 10 months.

Discussion

To date, less than 100 cases have been documented in the literature.[3] Earlier reported cases have suggested that AFS presents as a painful, intraosseous mass in the posterior region of the jaws.[5] Approximately 80% of tumours were reported in the mandible in contrast to 20% in the maxilla.[6] A male predilection with a male-to-female ratio of 1.6:1 was seen.[7]

Nearly two-thirds of AFSs are malignant de novo, with the average age of patients being 21.9 years, while one-third arises in a pre-existing benign AF, with the average age of patients being 33 years. Most of these patients with AFS have had an enucleation or curettage performed earlier at the same site.[2] The AFSs in general also occur at a later age (mean 27.5 years) as compared to AFs, which occurs at 14.6 years, suggestive of a stepwise malignant transformation.[9] Although it is a malignant tumour, it is relatively benign in that it does not show metastasis and the prognosis is relatively good.[4] However, 20% of patients die, as these lesions are locally aggressive and have a high local recurrence rate (37%).

Histologically, these tumours revealed benign odontogenic epithelial islands in the malignant mesenchymal portion of the lesion. The ectomesenchymal tissue exhibits remarkable increase in cellularity, pleomorphic malignant fibroblasts and numerous atypical mitotic figures.[11]

The present case adds another such rare case of AFS. Our case does not differ from those cases reported earlier in terms of the clinical, radiographic and histological features. As is with most of the cases, our case also appears to have emerged de novo. However, our study differs from other studies in that probably more cases of AFSs have emerged in a pre-existing AF and that these lesions are not as rare as it is described. Although there is no history of the presence of a lump or any evidence of AF, its presence could not be ruled out, as, according to Shafer, most cases of AF clinically are

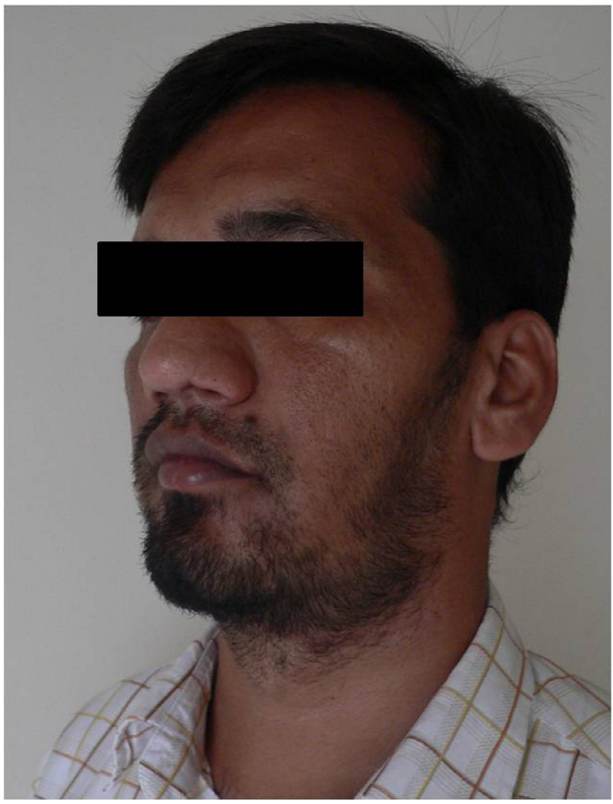

Figure 6. Clinical picture of the patient after chemotherapy and radiation therapy.

asymptomatic and are accidentally discovered during routine radiographic examination,[11] suggesting that, in most cases, the patients are unaware of an existing lesion within the jaws. Therefore, we believe that probably most of the AFSs, which were thought to have emerged de novo, could actually have developed in a pre-existing AF. Hence, the figures suggesting that two-thirds of AFSs emerge de novo and only one-third emerges through malignant transformation of AF may not be true in the strict-sense.

Takeda et al.[12] and Reichart[10] also suggest that there would be a gradual disappearance of the epithelial component as malignant transformation progressed. Hence, it was speculated that some of the previously reported cases of fibrosarcomas of the jaws may be indeed of odontogenic origin.[1] This may be true, and we are in full agreement with them, as in the present case, initially, sections revealed sarcomatous tissue without any evidence of odontogenic epithelial islands giving an impression of a fibrosarcoma. It was only after a thorough search, taking multiple sections from deeper areas of the biopsy specimen, that the tumour revealed few benign odontogenic epithelial islands, suggesting a diagnosis of AFS. The recurrent lesion, after 2 years, was diagnosed as a fibrosarcoma at the cancer institute, which could be due to complete absence of odontogenic epithelial islands, hence strengthening our view that at least some of the cases diagnosed as fibrosarcomas in the jaws in the past could have been AFS.

As we believe that more number of cases of AFSs arise in AF than what is thought, a patient with AF should be treated more aggressively (wide surgical excision) rather than with a conservative approach (curettage or enucleation) as has been described and that the patients should be informed about a possible recurrence and to report any suspicious changes.

Conclusion

AFS has been described as an extremely rare odontogenic tumour and that two-thirds of the cases arise de novo and only one-third of them occurs through malignant transformation of a pre-existing AF. First, for the fact that most of the cases of AFs are asymptomatic and pass unnoticed and are identified on routine radiography, we strongly believe that more number of AFS cases might have occurred in a pre-existing benign AF than what is reported earlier. Second, this tumour may not be as rare as previously reported, as at least some of the cases that have been reported in the past as fibrosarcomas of the jaws or odontogenic fibrosarcomas could be AFS. Hence, we stress the need for a thorough histopathological examination of multiple sections from deeper tissues when a case is suspected to be an odontogenic fibrosarcoma to rule out the possibility of AFS, and we suggest that the AFs should be treated more aggressively than at present.

Conflict of interest

None.

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None.

References

1. Altini M, Smith T. Ameloblastic dentinosarcoma—a case report. *Int J Oral Maxillofac Surg*. 1976;5(3):142–147.
2. Bregni RC, Taylor AM, García AM. Ameloblastic fibrosarcoma of the mandible: report of two cases and review of the literature. *J Oral Pathol Med*. 2001;30(5):316–320.
3. Hu YY, Deng MH, Yuan LL, Niu YM. Ameloblastic fibrosarcoma of the mandible: a case report and mini review. *Exp Ther Med*. 2014;8(5):1463–1466.
4. Huguet P, Castellvi J, Avila M, Alejo M, Autonell F, Basas C, et al. Ameloblastic fibrosarcoma: report of a case. Immunohistochemical study and review of the literature. *Medicina oral: organo oficial de la Sociedad Espanola de Medicina Oral y de la Academia Iberoamericana de Patologia y Medicina Bucal*. 2001;6(3):173–179.
5. Kobayashi K, Murakami R, Fujii T, Hirano A. Malignant transformation of ameloblastic fibroma to ameloblastic fibrosarcoma: case report and review of the literature. *J Cranio Max Surg*. 2005;33(5):352–355.
6. Lai J, Blanas N, Higgins K, Klieb H. Ameloblastic fibrosarcoma: report of a case, study of immunophenotype, and comprehensive review of the literature. *J Oral Maxillofac Surg*. 2012;70(8):2007–2012.
7. Lee OJ, Kim HJ, Lee BK, Cho KJ. CD34 expressing ameloblastic fibrosarcoma arising in the maxilla: a new finding. *J Oral Pathol Med*. 2005;34(5):318–320.
8. Leider AS, Nelson JF, Trodahl JN. Ameloblastic fibrosarcoma of the jaws. *Oral Surg Oral Med Oral Pathol*. 1972;33(4):559–569.
9. Muller S, Parker DC, Kapadia SB, Budnick SD, Barnes EL. Ameloblastic fibrosarcoma of the jaws: a clinicopathologic and DNA analysis of five cases and review of the literature with discussion of its relationship to ameloblastic fibroma.. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod*. 1995;79(4):469–477.
10. Reichart PA, Zobl H. Transformation of ameloblastic fibroma to fibrosarcoma. *Int J Oral Surg*. 1978;7(5):503–507.
11. Shafer WG, Hine MK, Levy BM. Cysts and tumors of odontogenic origin. In: Rajendran R, Sivapathasundharam B, eds. *Shafer's* Textbook of Oral Pathology. 5th ed. New Delhi: Elsevier; 2006:402–403.
12. Takeda Y, Kaneko R, Suzuki A. Ameloblastic fibrosarcoma in the maxilla, malignant transformation of ameloblastic fibroma. *Virch Arch A*. 1984;404(3):253–263.
13. Williams MD, Hanna EY, El-Naggar AK. Anaplastic ameloblastic fibrosarcoma arising from recurrent ameloblastic fibroma: restricted molecular abnormalities of certain genes to the malignant transformation. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod*. 2007;104(1):72–75.