Aggressive atypical ameloblastic fibrodentinoma: Report of a case

Girish B. Giraddi, Vipul Garg

Abstract

Ameloblastic fibroma and related lesions constitute a group of lesions, which range in biologic behaviour from true neoplasms to hamartomas. This group of lesions is also sometimes referred to as mixed odontogenic tumors and usually includes ameloblastic fibroma, ameloblastic fibrodentinoma and ameloblastic fibro-odontoma. Despite numerous efforts however, there is still considerable confusion concerning the nature and interrelationship of these mixed odontogenic tumors and related lesions. The malignant counterpart of these lesions namely aameloblastic fibrosarcoma, ameloblastic dentinosarcoma and ameloblastic odontosarcoma respectively are said to arise secondarily in their benign counterpart or de novo. Recurrence of the benign lesion raises the risk towards malignant transformation therefore a radical surgery should be planned inspite of enucleation or curettage. Here we present a case of an aggressive ameloblastic fibrodentinoma which was radically excised in the light of clinical and histological presentation followed by reconstruction of mandible.

Keywords: Ameloblastic fibroma, ameloblastic fibrodentinoma, ameloblastic fibrosarcoma, ameloblastic dentinosarcoma, ameloblastic odontosarcoma

Introduction

Ameloblastic fibroma (AF) and related lesions are defined by the WHO as neoplasms composed of proliferating odontogenic epithelium embedded in a cellular ectomesenchymal tissue that resembles dental papilla and has varying degrees of inductive change and dental hard tissue formation. This group of lesions is also sometimes referred to as mixed odontogenic tumors, and usually includes AF, ameloblastic fibrodentinoma (AFD) and ameloblastic fibroodontoma (AFO). Despite numerous efforts, however, there is still considerable confusion concerning the nature and interrelationship of these mixed odontogenic tumors and the related lesions.[1]

AFD, one of the lesions of the above-described group, is a rare entity and its very existence is not completely accepted. Indeed, AFD is considered by some authors to occupy a stage between the AF and AFO based on the extent of histodifferentiation.[2,3]

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Access this article online	
Quick Response Code:	Website: www.contempclindent.org
	DOI: 10.4103/0976-237X.94557

It is a controversial neoplasm with respect to its biological nature and histological diagnosis. In this 1992-WHO classification of odontogenic tumors, AFD is defined as a neoplasm similar to AF that also shows inductive changes leading to the formation of dentin. AFD has predominantly occurred in the posterior region of the jaw and, especially, the mandibular posterior region. This is usually in association with unerupted molar teeth in childhood.[4]

AFD usually occurs as a slow-growing, asymptomatic swelling in the mandibular posterior region. Occasionally, it may be associated with unerupted tooth. The age at the time of diagnosis falls in the first two decades of life. There is a male preponderance, with a male to female ratio of 2.4-3:1.[5]

Here, we report a case of aggressive AFD that presented as a rapidly growing mass associated with pain and presenting with resorption and perforation of the lingual cortical plate.

Case Report

A 17-year-old female reported to the Department of Oral and Maxillofacial surgery with a rapidly growing painful expansile swelling of the right body of the mandible measuring about 6 cm x 4 cm x 3 cm.

The swelling extended from the corner of the mouth to the angle of the mandible [Figure 1].

Examination of the oral cavity revealed a bony hard swelling extending from the right first premolar to the angle area. The buccal and lingual vestibule was obliterated and the expansion of lingual cortical plate was relatively higher. There was grade II mobility with respect to 44 and 45, while the 46, 47 and 48 were absent. The patient gave an unclear history of exfoliation of the posterior tooth. There was no evidence

of mucosal ulceration. Cervical lymph nodes of the same side were enlarged, firm, tender and mobile.

The orthopantomogram revealed a well-defined radiolucency measuring about 4.5cm x 2.5cm, associated with the impacted molar [Figure 2]. The borders were not sclerotic. The substance of the radiolucent area showed few flecks of radioopacity. The radiograph showed expansion and thinning of the cortex.

Computed tomography revealed a voluminous, space-occupying, expansile lesion at the posterior part of the right mandible. The tumor had caused expansion of the buccal and lingual cortical plate with perforation at some places. Minute calcifications were evident within the lesion [Figure 3].

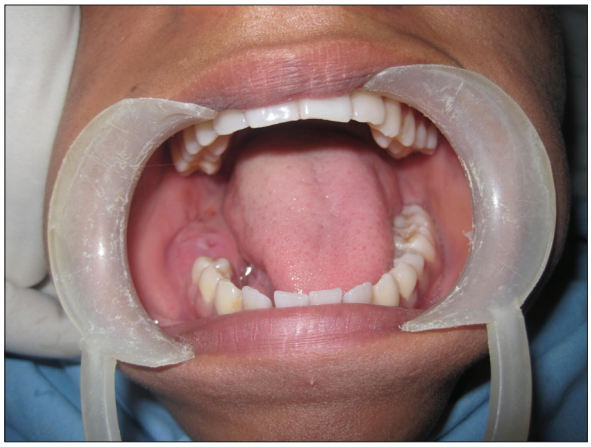

Figure 1: Pre-operative intraoral view shows buccal and lingual expansion in the posterior mandible

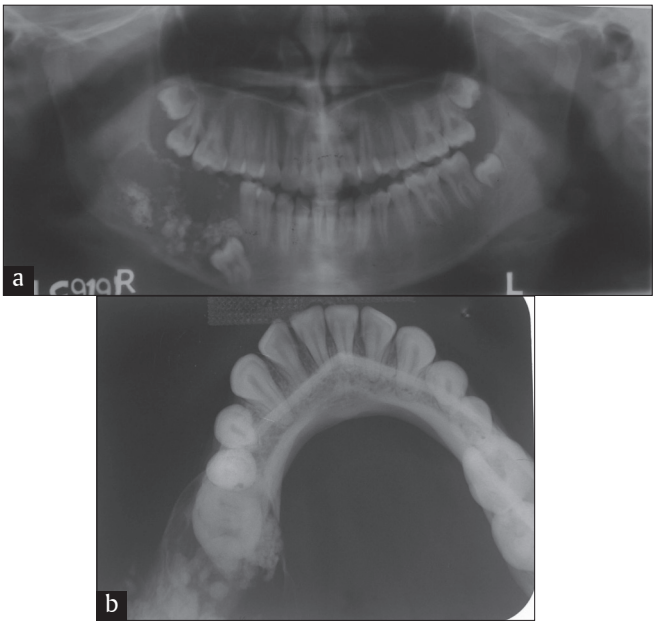

Figure 2: (a) Panoramic radiography shows an ill-defined radiolucent lesion with scattered foci of calcified material that contained several radioopaque bodies of varying sizes and shapes and an impacted first molar (right side) (b) Occlusal view shows expansion and thinning of the buccal and lingual cortical plate

A pre-operative differential diagnosis of calcifying epithelial odontogenic tumor, mixed odontogenic tumor, calcifying odontogenic cyst and fibrosseous lesion was given.

Incisional biopsy was performed and submitted for histopathological examination.

The section on low-power examination revealed neoplastic proliferation of odontogenic epithelial and mesenchymal tissue. On higher-power magnification, the odontogenic epithelial component consisted of multiple follicles and islands of tall columnar to cuboidal ameloblast-like cells with reversed nuclear polarity. The mesenchymal component was primitive connective tissue resembling the dental papilla of the tooth germ. Juxtaepithelial hyalinization was evident in few areas. A fibrillar pattern reminiscent of dentinal tubules was noted in the focal area as marked [Figure 4]. Enamel formation could not be identified even on multiple sections.

The most striking finding was the presence of increased cellularity with mild pleomorphism of the dental papilla-like cells in few areas [Figure 5]. However, in view of the absence of mitotic figures and, secondly, in correlation to the clinical

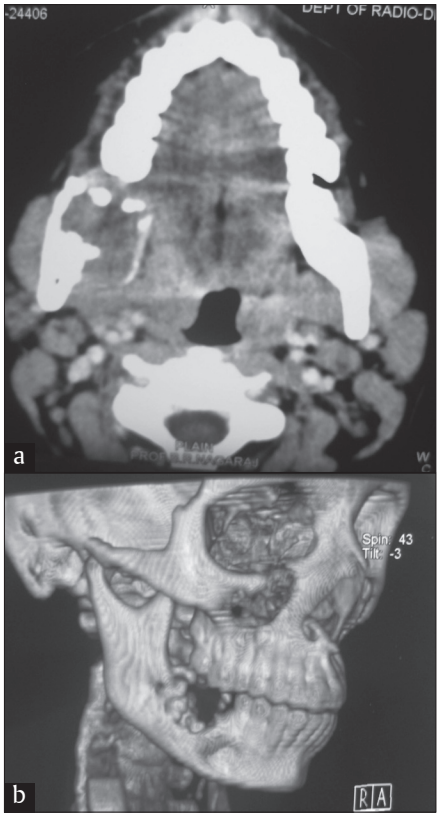

Figure 3: (a) Computed tomography shows an expansile lesion of the right mandible (b) Tomographic reconstruction image shows the extent of the lesion

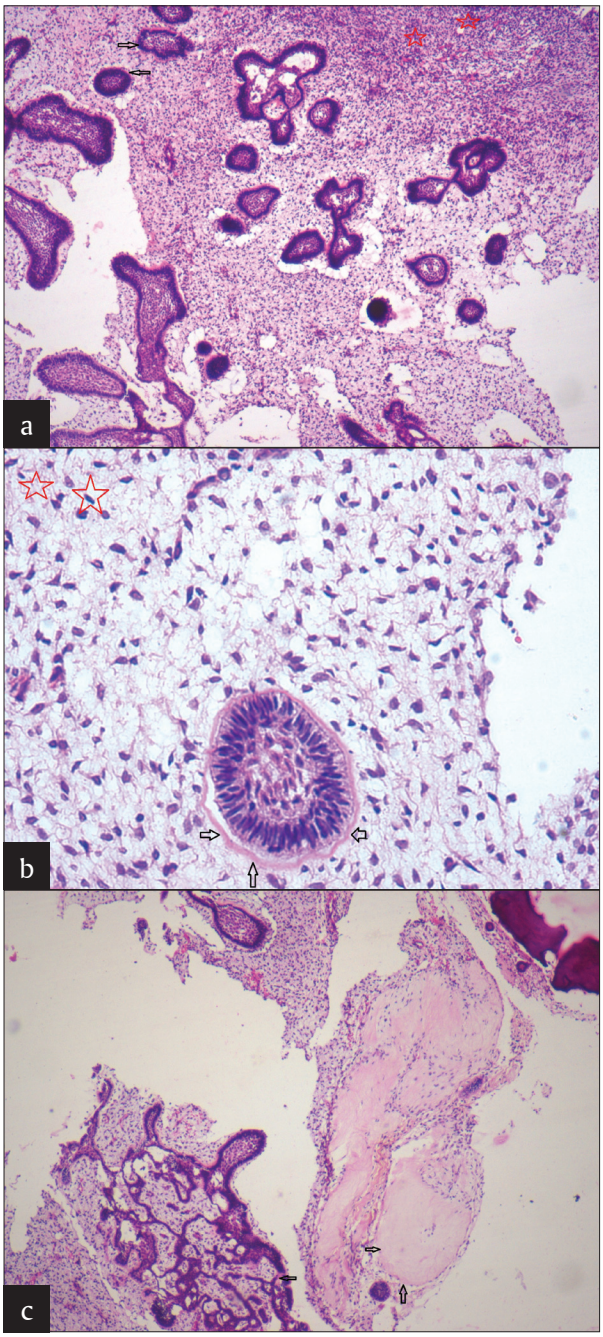

Figure 4: (a) Microscopic appearance of the lesion (hematoxylin and eosin). Islands of odontogenic epithelium with peripheral cells resembling ameloblasts and central cells resembling the stellate reticulum (arrows) of the enamel organ amid highly cellular loose connective tissue, similar to the dental papilla (stars) – ×40 original magnification (b) Dental hard tissue formation adjacent to the epithelial island, ×200 (c) Abundant dentinoid matrix – ×40 original magnification

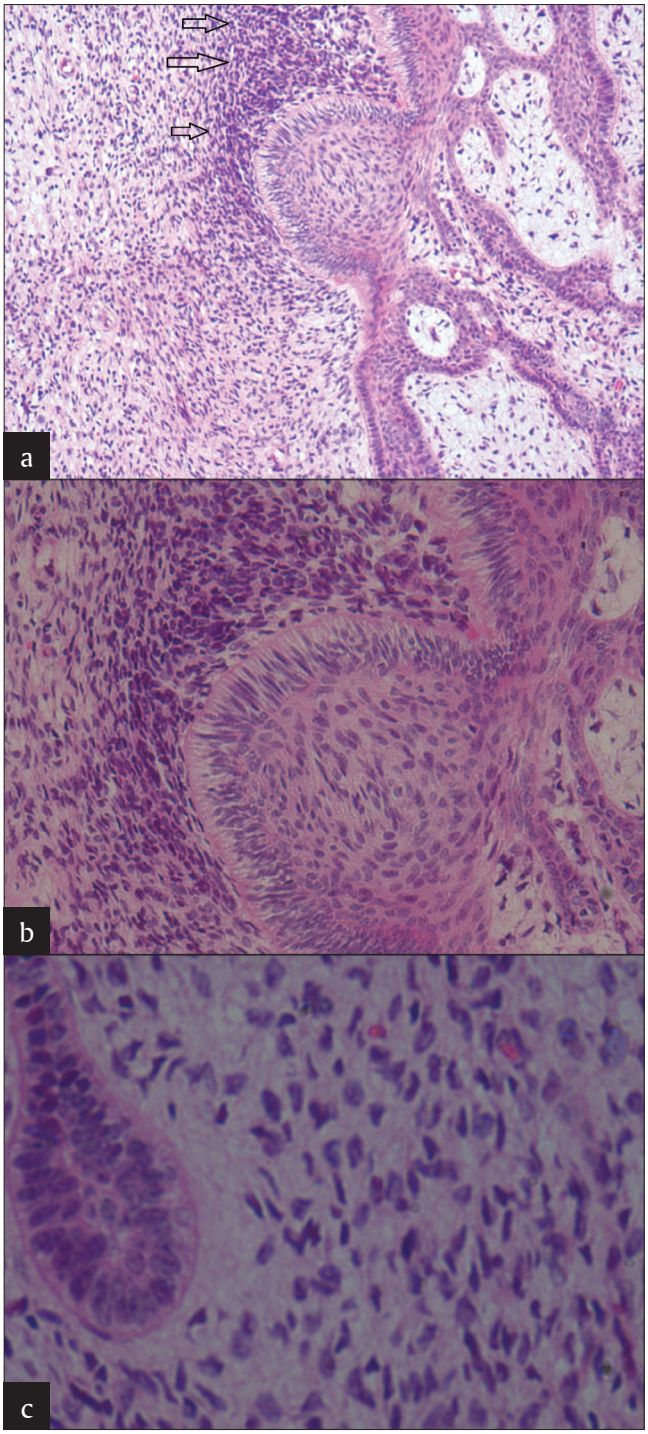

Figure 5: (a) Condensation of ectomesenchymal tissue juxtaepithelialy with marked increase in cellularity – ×100 (b) ×200 – ectomesenchymal cells exhibiting mild pleomorphism (c) Pleomorphic ectomesenchmal cells – ×400

and radiographic presentation of the lesion, a diagnosis of aggressive atypical AFD was made.

Depending on the histological diagnosis and aggressiveness of the lesion, a radical resection was planned in order to prevent further chances of recurrence and malignant transformation that are usually seen with recurrence of the lesion following inadequate removal. Reconstruction of the mandible was performed following resection [Figure 6]. The patient is kept under regular follow-up.

Discussion

Odontogenic tumors are a heterogeneous group of diseases ranging from hamartomas to benign and malignant neoplasms with metastatic potential. Odontogenic tumors arise from the odontogenic epithelium, ectomesenchyme and mesenchyme.[6]

There is still considerable confusion and discussion in the literature concerning the interrelationship of the group of odontogenic lesions consisting of odontogenic epithelium with odontogenic ectomesenchyme with or without dental hard tissue formation, sometimes referred to as the "mixed odontogenic tumors." In general, this term applies to AF, AFD and AFO. Further, of similar interest is the relationship, if any, between the above three lesions and the fully calcified or mineralized odontogenic lesions, the odontomas. Reichart *et al.* attempted to review and update the literature related to these lesions.[5]

The differential diagnosis of a radiolucency containing various amounts of radioopaque foci includes AFD, AFO and other mixed radiolucent/radiopaque lesions, such as calcifying epithelial odontogenic tumor, calcifying odontogenic cyst, adenomatoid odontogenic tumor and immature stage of odontoma. According to the WHO, AFO is included, along with AF and AFD, in the group of tumors of odontogenic epithelium with odontogenic ectomesenchyme, with or without dental hard tissue formation. The histological features of AF include odontogenic ectomesenchyme that resembles the dental papilla and strands or islands of odontogenic epithelium that resemble the dental lamina and enamel organ. If there is dentin formation, the lesion should be diagnosed as AFD; if there is also enamel formation, it should be diagnosed as AFO.[6]

In some of the cases previously diagnosed as AFD, dentine matrix or dentinoid tissue is an area of hyalinization around the epithelial component, and some workers have suggested that hyalinized material may not represent dentine formation. While in some other cases, hyalinised area containing entrapped cells has been considered as an abortive dentine or dentinoid tissue because of its proximity to the odontogenic epithelial. The margin of such a zone often shows radially arranged coarse fiber bundles with elongated cells, and these are rarely found tubular structures. It is important to realize that these AFD exhibit a different biological behavior than AF.[7]

Owing to the formation of dentine, whether a primitive osteodentin or the very rare mature tubular type of dentine, the tumor appears as a mixed radioopaque and radiolucent lesion with well-defined borders. If an embedded tooth is involved, the tumor is often closely associated with the crown.[5]

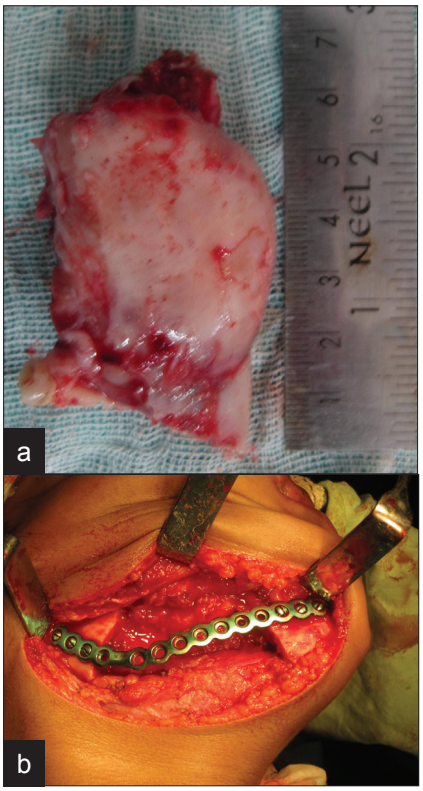

Figure 6: (a) Surgically excised tumor mass (segmental mandibulectomy) (b) Post resection reconstruction of the mandible using titanium plates

When the histogenesis of these mixed odontogenic tumors are considered, some controversy surrounds the relationship between AF, AFD, AFO and odontoma. Some consider them as separate entities. Others regard them as chronological stages in a continuum beginning from AF at one extreme and odontoma at the other extreme with ameloblastic fibroodontoma as well as AFD in an intermediate stage.[7]

In support of this latter proposition, Slootweg[8] analyzed 33 mixed odontogenic tumors and found that the mean age of the patients with ameloblastic fibroodontoma was lower than that of the patients with AF. If this finding is correct, the mean age of the patients with ameloblastic fibroodontoma (which is assumed to differentiate further from AF) should have been higher than that of those with AF. From this, the author concluded that AF represents a separate specific neoplastic entity that does not transit into a more differentiated odontogenic lesion. The age of our patients provides further evidence that contradicts this conclusion as our patient is an extremely young child.

Philipsen *et al*. have suggested that AF and AFD occur in two variants (with indistinguishable histology). The first is a neoplastic lesion, which if left *in situ* does not appear to mature further. The second variant is a hamartomatous (non-neoplastic) lesion that appears to be able to differentiate into

an ameloblastic fibroodontoma and mature further into a complex odontoma.[5]

On the other hand, the compound odontoma is considered as a separate entity resulting from a locally hyperactivity of the dental lamina. This idea has recently been incorporated into the suggested modifications for the 1992 edition of the WHO histological typing of odontogenic tumors.[9]

Clinically and pathologically, AFO and AFD are almost same. But, in the revised WHO classification of odontogenic tumors, both tumors have been considered as distinct entities. Presence of tooth germ elements, e.g. enamel and dentin in combination, or only dentin in isolation, help in the differentiation of the two lesions.[10]

Because of the presence of dentin only, and complete absence of enamel even in multiple sections, the present case was considered as ameloblastic fibrodentinoma.

It is important to note that in the revised WHO classification of odontogenic tumors, AFD and dentinoma are used synonymously. However, there are histological differences between several cases reported previously as dentinoma and AFD. Dentinoma has been defined as a very rare odontogenic neoplasm composed of odontogenic epithelium and immature connective tissue and characterized by the formation of dysplastic dentin. It is divided into two types on the stage of the development, immature and mature type. The immature variant is histologically composed of strands and cords of odontogenic epithelium without an enamel organ-like structure. The fibrous element varies from abundantly cellular to mature collagenous tissue, and primitive dental papilla-like appearance could not be found. Both epithelial and fibrous elements may therefore represent those of odontogenic fibroma with or without dentin formation. However, in the present case reported, the connective tissue typically resembled the ectomesenchyme and thus the diagnosis of AFD was made.[7]

The tendency of the tumor to recur and to undergo malignant transformation also denotes its neoplastic character. The malignant counterparts of AF, AFD and AFO are ameloblastic fibrosarcoma, ameloblastic fibrodentinosarcoma and ameloblastic fibroodontosarcoma, respectively. The 2005 WHO classification of odontogenic sarcomas presented two entities: ameloblastic fibrosarcoma (AFS) and ameloblastic fibrodentinosarcoma (AFDS) and/or fibroodontosarcoma (AFOS).[7,11]

In the malignant counterpart, it is only the mesenchymal component that undergoes malignant transformation while the epithelial component does not show any chance of cancer. Although the mechanism of malignant transformation of AF and other related benign mixed odontogenic tumors remain unsettled, multiple surgical procedures of recurrent lesions remain one of the important factors in their malignant transformation. Metastasis is not common and fatal cases have usually been associated with uncontrollable local infiltration following numerous recurrence.[7]

Therefore, in place of planning a conservative treatment, a radical resection should be performed especially if the lesion is aggressive and presents with erosion and perforation of the cortical plate.

When reviewing the literature on AFDS/AFOS, 15 cases have been reported in the English language literature up to 2009. The age range is 12–83 years, with a peak in the third decade. Clinically, AFDS/AFOS presented as a painful swelling and intraosseous mass, while AFs/AFDs are usually slow growing and painless lesions.

Ameloblastic fibroodontomas are regarded as possible precursor lesions. The WHO distinguishes odontogenic sarcoma devoid of dental hard tissue (AFS) from those displaying focal evidence of dentinoid (AFDS) or dentinoid plus enameloid (AFOS), but the WHO panel acknowledges that presence or absence of dental hard tissue in an odontogenic sarcoma is of no prognostic significance.[12]

Usually, AFD radiographically presents as a well-defined radiolucency, with little dense opacity. However, a case with an irregular border and expansion and perforation of the cortexes should be interpreted with caution and the possibility of malignant odontogenic tumor should be suspected.

The case presented here also showed an aggressive growth pattern and the histological study revealed few areas showing increased cellularity and cellular pleomorphism, raising a doubt toward the malignant transformation of the lesion. Therefore, a radical resection followed by resection was the treatment of choice. The patient is kept under regular follow-up in order to rule out any recurrence.

Conclusion

Considerably more information on the AFD has to be collected in order to better understand the lesion, its biological behavior, risk of malignant transformation and relationship to the other AF-related lesion. A proper clinical, radiological and histological corelation should be established before considering a lesion as a benign or a malignant variant, and the treatment should be planned accordingly. The diagnosed and treated cases should be kept under regular follow-up in order to rule out any evidence of recurrence and malignant transformation.

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How to cite this article: Giraddi GB, Garg V. Aggressive atypical ameloblastic fibrodentinoma: Report of a case. Contemp Clin Dent 2012;3:97-102.

Source of Support: Nil. **Conflict of Interest:** None declared.