

## Case Report

### Ameloblastic Fibroma: Report of 3 Cases and Literature Review

Shalini Gupta, Ankita Tandon, Divya Mehrotra, OP Gupta

#### Abstract

Ameloblastic fibroma is a rare odontogenic tumour of the jaw, in which both the epithelial and ectomesenchymal elements are neoplastic and is commonly seen in the first 2 decades of life. The common clinical manifestation is a slow-growing swelling. Ameloblastic fibroma is most common in adolescents and young adults, and is generally found in the mandible. We report three cases of Ameloblastic fibroma which had appeared at different anatomic sites and had caused marked facial disfigurement. The controversy regarding its management needs to be emphasized due to the malignant transformation reported. Therefore detailed biological behaviour along with the current surgical pathologic issues of Ameloblastic fibroma would be discussed in the subsequent presentation.

**Keywords:** Jaw Neoplasms; Dental Tissue Neoplasms; Odontogenic tumor; Ameloblastic Fibro-odontoma; Ameloblastic Fibro-dentinoma; Ameloblastic fibroma.

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#### Introduction

Ameloblastic fibroma (AF) is a relatively uncommon neoplasm of odontogenic origin comprising about 1.5-4.5% of all odontogenic tumors. Since its description by Kruse in 1891, there has been much debate and confusion regarding its nature and biological behavior. In the earlier years of the last century, AF has been recorded under different headings, with the most common one being the ameloblastoma (Adamantinoma). In 1946, Thoma and Goldman were the first to classify this tumor as a separate entity.<sup>1</sup> AF is defined by WHO as “consisting of odontogenic ectomesenchyme resembling the dental papilla and epithelial strands and nests resembling dental lamina and enamel organ. No dental hard tissues are present.” If the lesion has dentinoid tissue without or with enamel formation, it could be termed as ameloblastic-fibro-dentinoma (AFD) or ameloblastic-fibro-odontoma (AFO), respectively. This group of lesions is also sometimes referred to as mixed odontogenic tumors that histologically resemble various stages of tooth formation.<sup>1</sup>

Clinically the tumor grows slowly and painlessly, expanding the jaw. Radiographically, it appears as a unilocular/multilocular area of radiolucency with a smooth outline. Histologically, the tumor consists of strands and groups of epithelial cells in a connective tissue background and does not invade bone.<sup>2</sup> The

opinion that AF exhibits somewhat slow clinical growth, is well encapsulated and shows an innocuous benign behavior was supported by most authors, thus conservative treatment in keeping with the behavior of AF was recommended. On the other hand, some authors believed that AF is more aggressive than it had been thought and a more radical therapy is needed on the basis of reviewing recurrent or malignantly transformed cases in the literature.<sup>1</sup> Hence, three cases of AF are presented throwing light on the characteristic features as well the treatments adopted.

#### Case reports

**Case 1:** A 12 year old male patient visited our outpatient department with chief complaint of difficulty in swallowing and speech for last 1 year. On intra oral examination, an ulceroproliferative growth roughly 2.5 cm in greatest dimensions was appreciated posterior to maxillary left first molar extending to both palatal and buccal sides (Fig 1A). The growth was erythematous, showed irregular surface and appeared to interfere with local functions. No relevant medical history was associated. On radiographic examination, a well defined unilocular radiolucency was found encircling impacted maxillary left second molar with resorption of roots of maxillary left first molar (Fig 1B). The excision of entire pedunculated mass was performed along with removal of impacted maxillary left first molar.

**Case 2:** An 18 year old male patient with the chief complaint of right facial asymmetry since 2 years visited our outpatient department. On examination, an expansile mass of right mandibular body and ramus (Fig 2A) was noticed with mobility of associated teeth. Medical and dental histories were non contributory. On radiographic examination, a multilocular radiolucent lesion of right mandibular body was appreciated extending across the midline upto left mandibular first pre molar (Fig 2B). The CT findings revealed soft heterogenous predominantly hyperdense tissue attenuation extending in body, ramus and condyle of mandible on right side causing destruction of the same. The lesion infiltrated base and lateral border of tongue on right side. The interface of the lesion was ill defined with masseter muscle (Fig 2C). Mandibular resection, from ipsilateral condylar neck to contralateral canine, was performed with primary reconstruction using titanium reconstruction plate. The remaining condylar stump was fixed with two screws, while three screws were fixed in the body region.

**Case 3:** A 16 year old male patient visited our outpatient department with the chief complaint of pain in left lower posterior teeth for last 1.5 years. On examination, impacted mandibular left second molar was associated with a radiolucent lesion encircling the crown of mandibular left third molar and numerous intervening septae (Fig 3A). There was extensive cortical plate expansion. Partial mandibular resection was performed with immediate reconstruction using titanium reconstruction plate and screws.

Histopathologically, the lesions showed strands, cords and islands of odontogenic epithelium in a primitive connective tissue stroma. The epithelial islands and cords were characterized by peripheral columnar/cuboidal hyperchromatic cells and were frequently only two cell layers thick. The mesenchymal component consisted of evenly distributed plump ovoid and stellate cells in a loose myxoid to predominantly eosinophilic matrix resembling the primitive dental papilla. Mitosis was not a feature. No hard tissue structures were detected (Fig 2D & 3B). The patients were under regular follow up protocol to keep a check on recurrences and the results have been functionally and esthetically pleasant.

## Discussion

Ameloblastic fibroma is a rare mixed odontogenic tumor that usually occurs in young patients, the youngest patient reported is a 7 week old infant.<sup>3</sup> It has been diagnosed at a mean age of 15 years<sup>3</sup> with males being more affected than females.<sup>4,5</sup> It can appear either in the maxilla or mandible, with the posterior region of the mandible as its most common anatomic site.<sup>6,5</sup> Our cases presented clinically in both the jaws. The patients usually present with a hard swelling, but intraoral ulceration, pain, tenderness, or drainage may also be observed.<sup>7</sup> The lesion may affect the normal eruption of teeth in the area. An impacted tooth may be associated with the tumor in approximately three quarters of the cases<sup>5</sup> and the finding was concomitant in our cases.

The biological behaviour of AF shares the nature between hamartoma and true neoplasm.<sup>7</sup> Cahn and Blum postulated that an AF could develop eventually into an odontoma if the lesion had been allowed to remain. They believed that the only difference between the AF and the odontoma is the patient's age and the degree of maturation of the lesions. This would imply that all AF, AFD and AFO merely represent various stages of the same lesion, and will mature over time resulting in ultimately the formation of an odontoma. However, this "continuum concept" has not been widely accepted<sup>1</sup> as recent evidence suggests that AF exhibits a high recurrent rate and, in some cases, malignant transformation.<sup>7</sup> We have followed our cases for almost 2 years and have found no recurrence.

Radiographically, as in our cases ameloblastic fibromas are unilocular lesions, occasionally multilocular when larger, with smooth well-demarcated borders. These lesions are frequently associated with unerupted teeth, so may initially be interpreted as dentigerous cysts.<sup>5</sup> Microscopically, AF comprises strands and islands of an odontogenic epithelium in a loose and primitive connective tissue stroma, characteristic of dental papilla (embryonic dental pulp). The odontogenic epithelial cells are similar to those of ameloblastoma. Tiny islands resembling the follicular stage of the developing enamel organ may be observed. Some recurrent cases developed dentin formation with or without enamel structures, and subsequently differentiate over time into odontoma.<sup>7</sup>

Mitoses should not be a feature of ameloblastic fibroma. The presence of mitosis should expand the differential diagnosis to include malignant entities, to include ameloblastic fibrosarcoma.<sup>5</sup>



Figure 1A: Clinical photograph showing ulceroproliferative growth



Figure 1B: Orthopantomograph showing unilocular radiolucency encircling impacted maxillary left second molar.



Figure 2A: Clinical photograph showing expansile mass related to right mandibular body.



Figure 2B: Orthopantomograph showing extensive radiolucency.

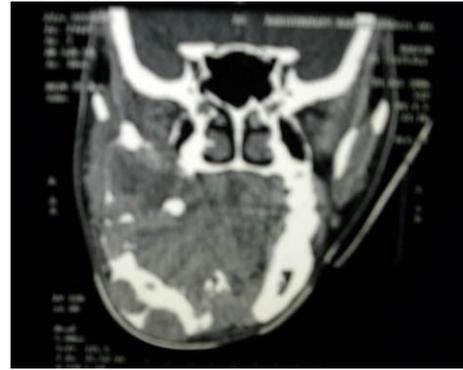


Figure 2C: CT scan showing an expansile growth involving the right mandibular area.



Figure 3A: Orthopantomograph showing unilocular radiolucency encircling mandibular left third molar crown.

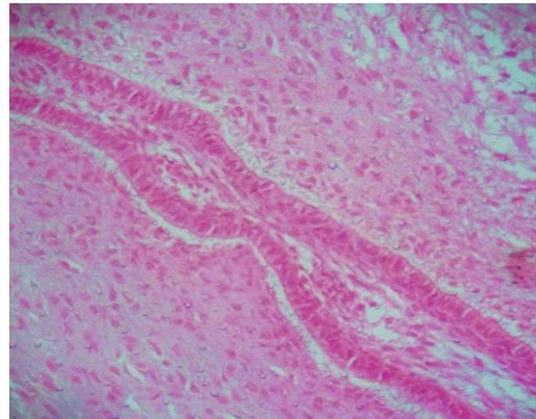


Figure 2D: Photomicrograph showing cords of odontogenic epithelium in a primitive connective tissue stroma.

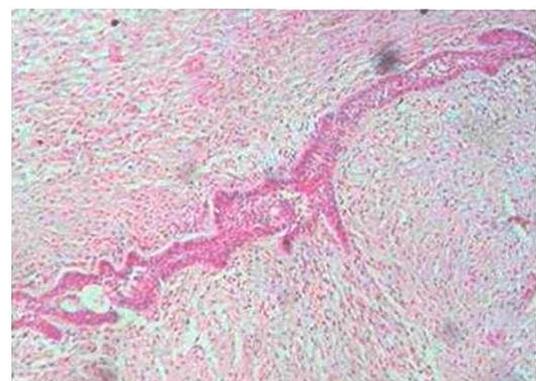


Figure 3B: The Photomicrograph shows epithelium in the primitive ectomesenchyme.

In cases undergoing malignant transformation, there are unequivocal changes in the mesenchymal component, and the odontogenic epithelium is completely disappeared as malignant transformation of AF was found to be associated with oncogenic aberrations in tumour-related genes. Mesenchymal proliferation within the tumour resulting in a loss of an epithelial component, is a usual presentation of sarcomatous changes of AF.<sup>7</sup>

Treatment for ameloblastic fibroma is described as conservative excision. Philipsen et al. proposed that the innocuous behavior of the lesion does not justify aggressive initial treatment but rather meticulous surgical enucleation with close clinical follow-up.<sup>3</sup> In general, a conservative approach, such as enucleation with curettage of the surrounding bone, should be applied for young patients. The neoplastic nature of AF is often suggested by the fact that some of these tumors could recur following surgery and that malignant transformation from a pre-existing AF has been reported.<sup>8</sup> While uncommon, the possibility of malignant transformation of ameloblastic fibroma into ameloblastic fibrosarcoma is well documented<sup>7</sup> and the recurrence rate of AF found by Trodahl *et al.* was 43.5%; on the other hand by Zallen *et al.* it was 18.3% after reviewing the literature with 85 cases of AF.<sup>8</sup> An aggressive surgical treatment is therefore suggested by some authors because of the possibility of malignant transformation of an Ameloblastic fibroma to an ameloblastic fibrosarcoma. No matter what the reason of recurrence is, a long term follow-up is necessary.<sup>8,9</sup>

Various immunohistochemical markers of intra and extracellular proteins have also been studied in AF and other benign odontogenic mixed tumors.<sup>10</sup> In Ameloblastic Fibroma, odontogenic epithelial cells are fully positive for cytokeratin detected by antibody KL-1. On the other hand, only immature dental papilla-like mesenchymal tissue, especially around the dental lamina like odontogenic epithelium, are positive for tenascin. Positive vimentin staining can be observed in some areas of immature dental papilla like cells as well as the basement membrane of odontogenic epithelium. These findings are very well suggestive of the fact that Ameloblastic fibroma develops at an early stage of tooth formation.<sup>1</sup> Some authors suggested the use of proliferating

indices, such as AgNOR, PCNA and Ki-67, to institute an appropriate treatment for each patient, and possibly, to suggest the malignant development.<sup>7</sup> Among these, PCNA is a nuclear protein which is associated with the S phase of DNA synthesis in association with cell proliferation. MIB-1 reacts with Ki-67 nuclear antigen associated with cell proliferation and, has been found throughout the cell cycle excepting for G<sub>0</sub>-phase.<sup>1</sup>

### Conclusion

Evaluation of the growth potential in AF and related lesions could therefore be of help in understanding tumor aggressiveness and in selecting appropriate surgical procedures.

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