CASE REPORT

Ameloblastic fibroma: an uncommon entity

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SUMMARY

Ameloblastic fibroma is an uncommon mixed odontogenic tumour, which is often confused with ameloblastoma. It exhibits both epithelial and mesenchymal components with absence of any calcified dental structure. This paper presents two cases of this rare entity with detailed review of literature.

BACKGROUND

Ameloblastic fibroma (AF) is known as a tumour of odontogenic origin characterised by the proliferation of both epithelial and mesenchymal odontogenic tissues without the formation of hard tissue structures.¹ It is relatively uncommon, representing only 2.5% of all odontogenic tumours.² The epithelial (ameloblastic) component consists of islands and cords of odontogenic epithelium that often resemble the dental lamina of early tooth development. There is often sufficient differentiation of the peripheral cells of these islands for recognition of characteristic high cuboidal or columnar ameloblasts. Not infrequently, the central portions of the islands resemble the stellate reticulum of the embryonic enamel organ. The mesenchymal component closely resembles the fibromyxoid tissue of the dental papilla, that is, the portion of the developing tooth bud that underlies the 'cap' or 'bell' of the enamel organ. Both epithelial and mesenchymal components appear to be involved in the proliferation, and for that reason the tumour has traditionally been classified as a member of the mixed (ectodermal and mesodermal) odontogenic tumour group.³

The WHO classification defines this lesion as: "The ameloblastic fibroma is a rare benign neoplasm in which both the odontogenic epithelial and ectomesenchymal components are neoplastic".¹ The tumour was first described as AF by Kruse in 1891.² Gorlin *et al*⁴ classified AF as epithelial odontogenic tumour with induction and in 1961, the latter author reviewed 35 documented cases in

their series and found two recurrences. AF was classified as a benign neoplasm in the first WHO Histological Typing of Odontogenic Tumors, 1971.⁵ A total of 24 cases from the files of Armed Forces Institute of Pathology were reviewed by Trodahl JN in 1972.³ In 1992, WHO classified this tumour as a true mixed tumour.⁵

CASE PRESENTATION**Case 1**

A 12-year-old boy reported to the dental clinic with swelling on the left maxillary region. Clinical examination revealed smooth swelling obliterating the gingivobuccal sulcus from the canine to the first molar region (figure 1). On palpation the swelling was non-tender. The lesion was surgically excised and was submitted for histopathological examination.

Case 2

A 10-year-old boy reported to the dental clinic with a swelling in the right mandibular region. On clinical examination facial asymmetry was obvious and the swelling was seen in the area of right body and angle of the mandible. Intraorally the occlusion was altered. The swelling was firm, non-tender and obliterated the mucogingival fold. The overlying mucosa was normal in colour. Surgically excision was performed and tissue was submitted for histopathological examination (figure 2).

INVESTIGATIONS

Biopsy specimens from both the cases were submitted for histopathological examination.

Case 1

Microscopically the H&E stained sections revealed neoplastic tissue characterised by islands of odontogenic epithelial cells arranged in strands, cords and nests. The epithelial cells were cuboidal to columnar and well differentiated. The islands of

Figure 1 Swelling in the maxillary left region.

Figure 2 Gross specimen.

To cite: Vij R, Vij H. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-010279

epithelium were embedded in an immature connective tissue stroma (figure 3).

Case 2

Microscopy revealed the classical picture of AF similar to that seen in case 1 (figure 4).

DIFFERENTIAL DIAGNOSIS

The clinical presentation of this lesion is exemplary and does not cause any dilemma in diagnosis. If the presentation is not typical then other entities, which need to be differentiated, can be odontogenic myxoma, dentigerous cyst, odontogenic keratocyst, central giant cell granuloma and histiocytosis.

TREATMENT

Since this pathology lacks invasiveness it is not aggressively treated. Very rarely recurrences have been reported. Thus both of the cases under discussion were treated conservatively.

OUTCOME AND FOLLOW-UP

Both the patients were asked to come for regular follow-up examination at an interval of 3 months for the first year and 6 months interval thereafter. The first patient came for first two follow-up examinations. There were no complications after an uneventful healing, but the patient was then lost for further follow-up. The second patient did not come for any of the follow-up appointments.

DISCUSSION

AF is a neoplasm of childhood/adolescence.⁶ The age at time of diagnosis falls within the first two decades, as was observed in both the cases on hand. In a survey of cases from the Armed Forces Institute of Pathology by Trodahl³ age range of 1.5–41 years and mean age of 15.5 years was reported. Adebayo *et al*⁷ reported a mean age of 23 years and an age range of 15–58 years.

AF has been known to affect men more commonly than women in many of the reports available. Both the cases currently under discussion also were seen in men. In study performed in the Armed Forces Institute of Pathology by Trodahl³ male to female ratio of 2:1 was reported. In study published by Adebayo *et al*⁷ equal predilections for men and women were reported.

The majority of AFs are found in the posterior region of the mandible as also observed in the second case reported in the present paper. AF seems to occur exclusively as a central or

Figure 4 Odontogenic island with peripheral columnar cells (H&E, ×400).

intraosseous tumour. Adebayo *et al*⁷ reported six cases in the maxilla, two in the posterior aspect of the mandible and two in the anterior mandible, out of 10 cases.

AF is a painless, slow growing and expanding tumour as were both the cases reported in the current article. AF exhibits somewhat slower clinical growth than the simple ameloblastoma and does not tend to infiltrate among trabeculae of bone. Instead it enlarges gradually so that the periphery of the lesion often remains smooth. It will frequently cause no symptoms on the part of the patient and the most frequent presenting symptom is swelling. Pain, tenderness or mild swelling of the jaw may induce the patient to refer a dentist.⁸

The tumour appears as a well-defined, unilocular or multilocular radiolucency, in most cases exhibiting a sclerotic radiopaque boundary. Whereas a multilocular appearance is often seen in larger tumours and is found in about 75% of the cases, a unilocular pattern characterises the smaller lesions. When associated with an unerupted tooth, the tumour may radiographically easily mimic a dentigerous cyst.⁶

Microscopically, the epithelial tumour component is characterised by proliferating islands, cords and strands of odontogenic epithelium. The strands often resemble the dental lamina of early tooth development with a double or triple layer of cuboidal cells. The islands, however, often show a peripheral row of high cuboidal or columnar, ameloblast-like cells. The central portions of the islands may resemble the stellate reticulum of the embryonic enamel organ. The amount and density of the epithelial component may vary considerably from area to area within the same tumour.⁶ These characteristic features were noted in both the cases presented (figures 3 and 4). Cystic degeneration within the epithelium is not a common finding. The ectomesenchymal tumour component is that of an embryonic, cell-rich mesenchyme mimicking the dental papilla or primitive pulp tissue. The cells are rounded or angular and fibroblast-like, and there are very few and delicate collagen fibrils. These morphological characteristics are of great importance when differentiating this tumour from the ameloblastoma in which the stroma is a mature, fibrous connective tissue. The degree of cellularity varies within one and the same tumour and between tumours. Some areas of the ectomesenchymal tumour tissue may show a loose myxomatous structure with a weakly positive metachromatic intercellular substance. Although a cell-free zone and/or a zone of hyalinisation may be found at the epithelial–connective tissue interface, induction phenomena, resulting in dental hard tissue formation, do not as per

Figure 3 Odontogenic islands in immature stroma (H&E, ×100).

definition occur in AFs. A definite capsule is considered an unusual feature of the AF. Melanin granules within the epithelial tumour component have been found in some cases of AFs.^{9 10}

The ultrastructure of AF has been examined by Farman *et al.*¹¹ In comparing the follicular ameloblastoma with the AF, the authors found that changes in the basal lamina region in both tumours are consistent with attempted inductive stimulation, possibly very similar to that during normal odontogenesis. Both tumours revealed differing degrees of thickening of the lamina densa by a granulofilamentous material. In no case was this granulofilamentous zone wide enough to account for the hyaline cell-free bands seen in light microscopy. The ultrastructural changes described by Farman *et al.*¹¹ are very similar to those reported by van Wyk and van der Vyver¹⁰ who described the epithelial connective tissue interface in a case of AF with initial dentinoid formation (early ameloblastic fibrodentinoma). Where dentinoid material, consisting of dense collagen bundles, was evident, the interface showed, in some areas, an interrupted basal lamina coated with a dense 'brush border' of aperiodic fibrils and the epithelial cells were cylindrical, with their nuclei situated proximally.

A malignant counterpart of the AF has been described, according to a review by Muller *et al.*¹² in as many as 51 cases. Yet another case was added very recently.¹³ The ameloblastic fibrosarcoma, the term that is applied for this rare tumour, is characterised by a malignant transformation of the ectomesenchymal component and not the odontogenic epithelium. The average age at the time of diagnosis for ameloblastic fibrosarcoma is 27.5 years as opposed to 14.8 years for the AF. This age difference supports a step-wise progression of a benign to a malignant tumour as opposed to a *de novo* malignancy.¹²

The treatment of AF has been somewhat more conservative than that of the simple ameloblastoma, since it does appear to infiltrate bone as actively or as widely as the ameloblastoma. It also tends to separate from the bone more readily. Though considered to have little tendency for recurrence, variable recurrence rates have been reported by various authors.⁸

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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Learning points

- Ameloblastic fibroma (AF) should always be kept in mind in younger age group patients presenting with orofacial swellings.
- Though AF is a tumour, it can be successfully treated with a conservative approach.
- Not every pathology needs aggressive treatment.
- Histologically the ectomesenchymal component of AF along with the relatively scanty stellate reticulum in smaller follicles as compared with ameloblastoma helps in differentiating these pathosis from each other.

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