Oral Pathology | Clinical and Laboratorial Research in Dentistry

Ameloblastic fibroma: a case report

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ABSTRACT | Ameloblastic fibroma is a rare benign odontogenic tumor in which both epithelial and ectomesenchymal components are neoplastic. A 24-year-old male patient was referred to the Stomatology Department with difficulty to chew and swelling in the right posterior region of the mandible. The panoramic radiograph showed a well-circumscribed, unilocular radiolucent lesion with partially radiopaque borders involving first and second unerupted molars. Computed tomography imaging presented a hypodense image with well-delimited isodense content, bulging and rupture of cortical bones. The patient underwent an incisional biopsy. Microscopically, the lesion was composed of many mesenchymal tissue cells in strand form, arranged in cords, islands and nests of odontogenic epithelium; the diagnostic was ameloblastic fibroma. The patient was referred to the hospital for enucleation and curettage of the lesion and extraction of the associated teeth. After 8 months of follow-up, no recurrence was observed. This case emphasizes the importance of differential diagnosis, anatomopathological exam, and both clinical and imaging follow-up, since this kind of tumor can recur and progress to malignancy.

DESCRIPTORS | Odontogenic Tumors; Oral Pathology; Ameloblastic Fibroma.

RESUMO | **Fibroma ameloblástico: um estudo de caso** • O fibroma ameloblástico é um tumor odontogênico benigno raro no qual os componentes epiteliais e ectomesenquimais são neoplásicos. Paciente de 24 anos de idade foi encaminhado à clínica de Estomatologia devido à dificuldade de mastigar e edema na região posterior direita da mandíbula. A radiografia panorâmica evidenciou uma lesão radiolúcida unilocular, circunscrita, com bordas parcialmente radiopacas envolvendo o primeiro e segundo molar não irrompidos. A tomografia computadorizada apresentou imagem hipodensa, com conteúdo isodenso, bem delimitada, com abaulamento e rompimento das corticais ósseas. O paciente foi submetido a uma biópsia incisional. Microscopicamente, a lesão foi composta por tecido mesenquimal rico em células, formando cordões, ilhas e ninhos de epitélio odontogênico, cujo diagnóstico foi de fibroma ameloblástico. O paciente foi encaminhado ao hospital para enucleação e curetagem da lesão com extração dos dentes associados. Após 8 meses de acompanhamento, não se observou recorrência. Este caso enfatiza a importância do diagnóstico diferencial, exame anatomopatológico, e acompanhamento clínico e radiográfico, uma vez que este tumor pode recidivar e evoluir para malignidade.

DESCRITORES | Tumores Odontogênicos; Patologia Oral; Fibroma Ameloblástico.

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INTRODUCTION

Ameloblastic fibroma (AF) is a rare benign odontogenic tumor, originating from the odontogenic epithelium and odontogenic mesenchyme,[1] and it is classified as a true mixed tumor.[2]

According to Barnes et al.,[2] mixed odontogenic tumors include: ameloblastic fibrodentinoma (AFD), ameloblastic fibro-odontoma (AFO), odontoma complex and compound, odontoameloblastoma, calcifying cystic odontogenic tumor, dentinogenic ghost cell tumor, and ameloblastic fibroma. To some authors, mixed odontogenic tumors are different developmental stages of the same lesion.[1]

The incidence of odontogenic tumors in a study by Nalabolu et al. was 2.17% of a total 7,400 oral biopsies. The AF corresponded to 0.6% of all odontogenic tumors.[3] The mean age was 14.8 years (ranging from 7 weeks to 62 years).[2] AF occurs more frequently in the mandible and the posterior region is more affected than the anterior region.[1,4]

Clinical and radiographic features of odontogenic tumors, as well as their prognosis and malignant transformation are conflicting.[1] The radiographic features include well-defined, uni- or multilocular radiolucency, and, in most cases, a radiopaque boundary.[2,4]

This case report describes the case of a young man affected by mandibular AF, associated with first and second molars on the right side.

CASE REPORT

A 24-year old male was referred to the Stomatology Department of the School of Dentistry, University of São Paulo, complaining of difficulty chewing and a progressive, asymptomatic increase in the size of his right mandible, which he noticed about 15 days before examination. The patient had no relevant medical history.

Extraoral examination revealed facial asymmetry, bulging of the right lower third of the face, intact skin, no palpable lymphonodes, and no paresthesia.

The intraoral examination revealed a tumor in the right mandible, with an ulcerated surface, a reddish color, well-defined borders, and measuring approximately 3 cm. Absence of the second premolar and the first and second molars was noted in the region of the tumor.

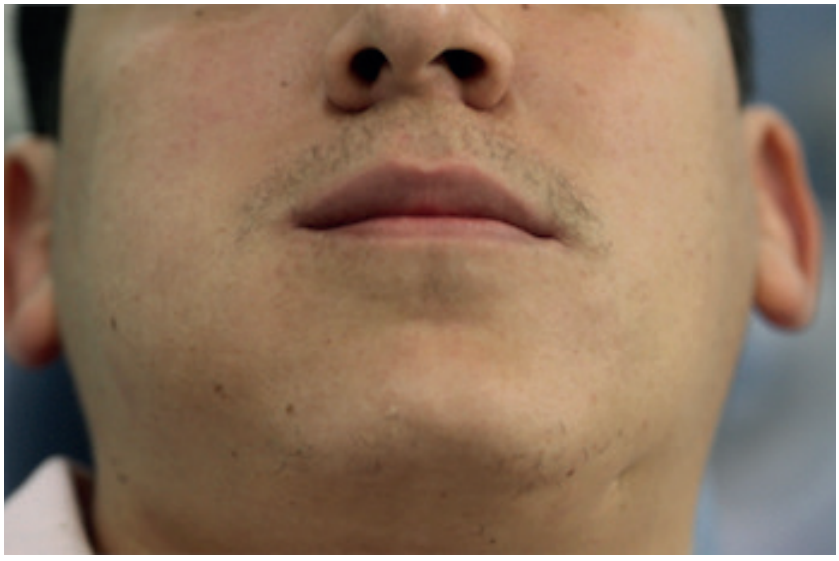

Figure 1 | Extraoral examination revealed facial asymmetry with bulging of the lower third of the face and intact skin, on the right side.

A panoramic radiograph (PR) revealed a unilocular, radiolucent lesion with a partially defined radiopaque boundary, associated to non-erupted first and second molars displaced towards the base of the mandible. Helicoidal computed tomography (HCT) soft window image revealed a hypodense image with isodense content, and cortical bulging with rupture of alveolar crest.

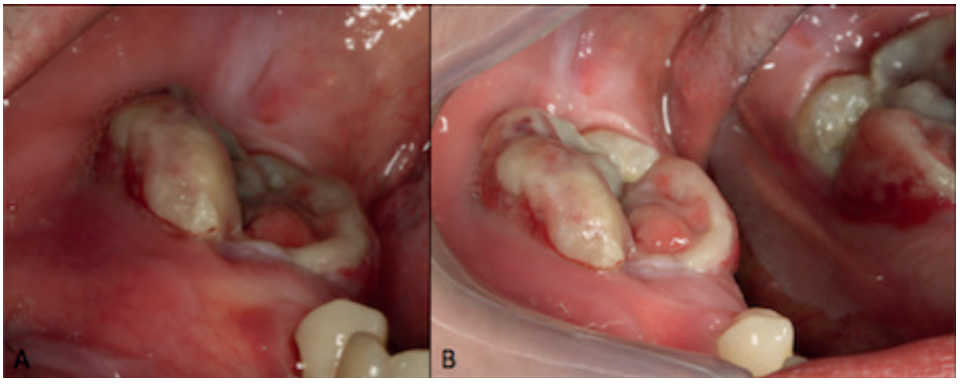

Figure 2 | **A,B**: An ulcerated tumor due to chewing, affecting the posterior right mandible and causing expansion of the cortical bone.

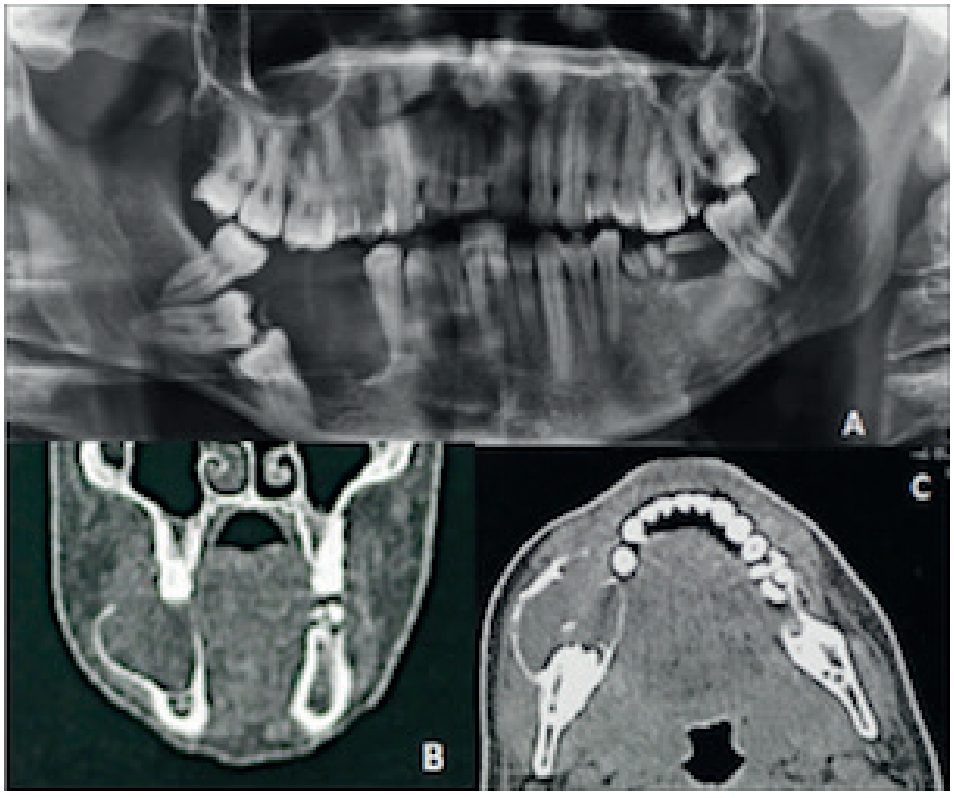

Figure 3 | **A**: A panoramic radiograph shows a well-delimited radiolucent lesion with partially radiopaque borders. **B, C**: HCT coronal and axial view of tissues shows a well-delimited, unilocular, hypodense lesion with isodense content, with cortical expansion and rupture, affecting the posterior right mandible.

INVESTIGATION, HISTOPATHOLOGY AND TREATMENT

The patient was submitted to an incisional biopsy under local anesthesia, and the tissue was sent for histopathological analysis. Microscopically, the tumor consisted of odontogenic epithelium lying in mesenchymal tissue resembling embryonic tooth pulp. The odontogenic epithelium consisted of short and long narrow cords or islands, usually two cells thick, with cuboidal or columnar cells sometimes in anastomosing arrangement. The final histopathological diagnosis was ameloblastic fibroma.

Therefore, surgery was indicated and performed under general anesthesia, with curettage of the lesion and tooth extraction (Figure 5). AF diagnosis was confirmed. A helicoidal tomography was performed 8 months after surgery (Figure 6). The patient has been followed-up with no evidence of recurrence, and has been asymptomatic ever since (Figure 7).

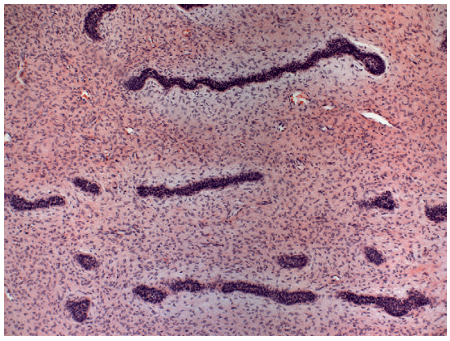

Figure 4 | Benign neoplasm consisting of mesenchymal tissue associated with odontogenic epithelium arranged in short and long, narrow cords or islands (H&E 200x).

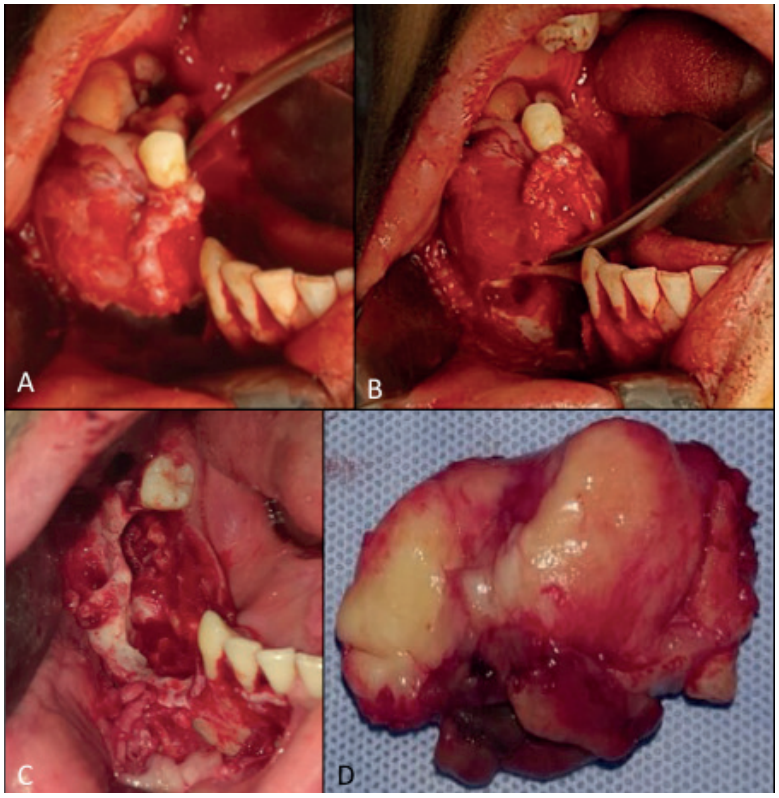

Figure 5 | **A-C**: Trans-surgical procedure: enucleation with curettage of the surrounding bone and removal of the affected tooth. **D**: An extracted specimen.

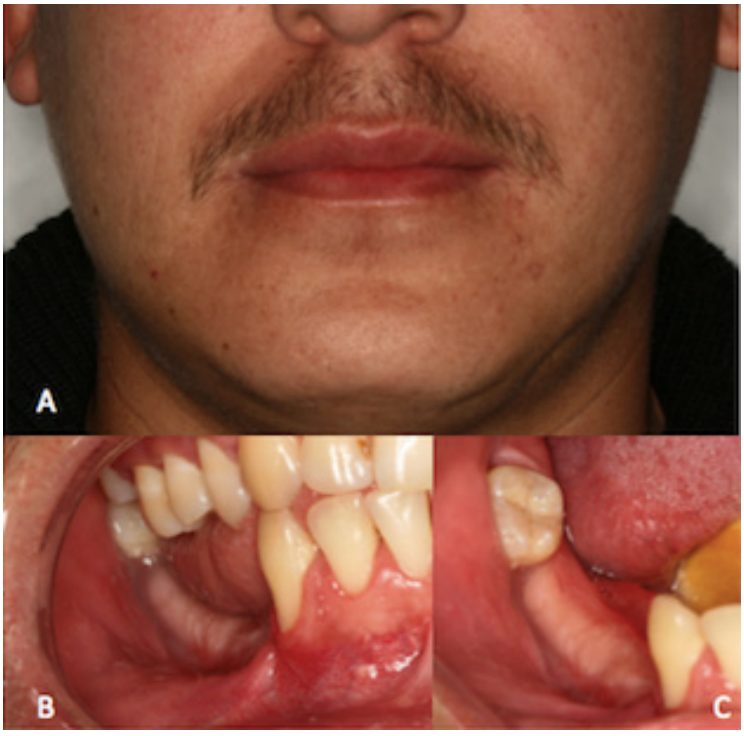

Figure 6 | **A**: Five months after surgery, the patient presented symmetry. **B, C**: Intraoral examination revealed normal alveolar ridge and intact surface.

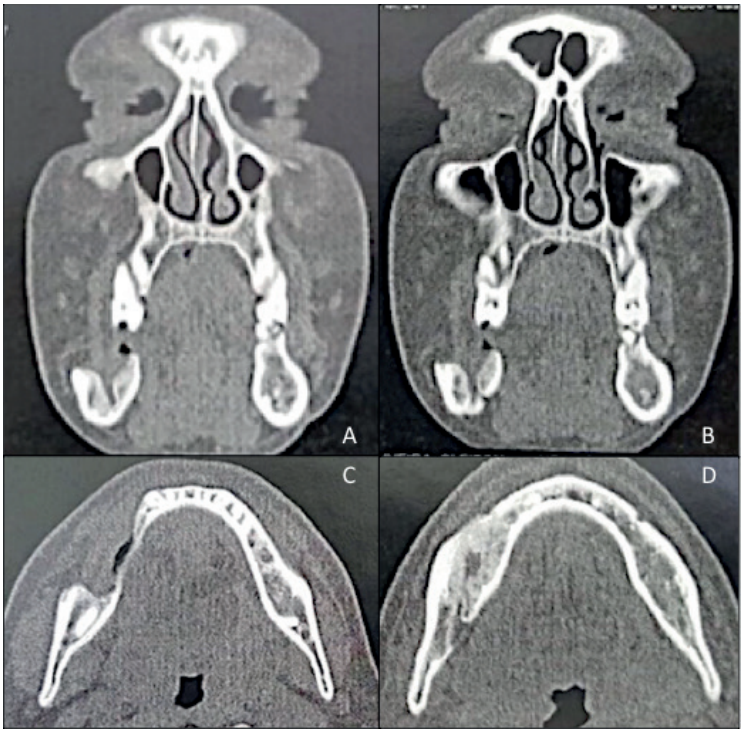

Figure 7 | **A, B**: HCT coronal view shows an area of bone defect from surgery, with no evidence of lesion. **C, D**: HCT axial view shows a hyperdense area, suggesting a process of bone repair in the right mandible.

DISCUSSION

Ameloblastic fibroma of the jaw is a benign, relatively rare, mixed odontogenic tumor, whose epithelial and mesenchymal components are neoplastic.[2,4] This tumor is usually diagnosed in the first and second decades of life (72.4%), when odontogenesis is complete (80% of cases), and affects mainly the mandible.[1,4] In this case, the lesion was diagnosed in the third decade of life, and occurred in the posterior region of the mandible. However, some cases of AF in the maxilla have also been reported.[1,5]

AF does not have a specific sign or symptom, and it is often observed in a routine radiograph, in the form of cysts and other odontogenic tumors.[2] In this case, the patient never complained about the absence of his right lower molars. His chief complaint was just difficulty chewing due to the large mass of tissue in this region.

Most cases of AF present painless swelling, or are discovered due to disturbances of tooth eruption. Radiographically, the tumor presents a well-demarcated radiolucency, often associated with a malpositioned tooth.[2] In addition, a multilocular pattern often characterizes larger tumors (75% of the cases), and a unilocular pattern is more common in smaller lesions (up to 4 cm),[6] as was this case. Differential diagnosis of AF lesions must be made, distinguishing ameloblastoma, odontogenic myxoma, dentigerous cysts, odontogenic keratocysts, central giant cell granuloma, and histocytosis.[7]

Histological examination of AF showed strands, cords, and islands of odontogenic epithelium in a primitive connective tissue stroma closely resembling the dental papilla. No hard tooth structures were detected in any of the primary tumors.[4] Tumors with AF histomorphology may form dysplastic dentin; in this case, they are called ameloblastic fibrodentinoma.[2] Some authors state that AF is a separate, specific neoplastic entity that does not develop into a more differentiated odontogenic tumor.[2]

Another study asserts that there are two variants of ameloblastic fibroma: neoplastic and hamartomatous. Lesions in patients aged >22 years are considered true neoplasms, whereas those occurring in younger patients may be either true neoplasms or odontomas. Asymptomatic, small unilocular lesions with no or minimal bone expansion in young individuals are likely to be developing odontomas, whereas large, expansive lesions with bone destruction are neoplasms. Since the histopathology of these two variants is indistinguishable, they may be distinguished by clinical and radiological features.[1] It is important to emphasize that this case was classified as a neoplasm.

The ectomesenchymal component of AF presents relatively scanty stellate reticulum in smaller follicles, compared to ameloblastoma. These histological characteristics help distinguish the pathologies.[7]

AF may rarely present ghost cell differentiation and calcification in the epithelial component. Recently, a study addressing an association between AF and calcifying odontogenic cyst (COC) with ghost cell differentiation was published. Interestingly, all cases of COC with ghost cell differentiation were observed in the epithelial lining. AF components existed in the cystic wall and some solid areas. Luo et al. described a case of AF that had ghost cell differentiation and calcifications in some of the neoplastic epithelial islands, but did not present any histological characteristics of COC in the cystic wall. Most lesions were cystic-solid, comprised of odontogenic epithelial strands, islands, and ectomesenchymal myxoid component. The behavior of ghost cell differentiation in AF remains unclear.[8]

The epithelial component of AF consists of branching and anastomosing epithelial strands of different size, which form knots. Mitotic figures both in epithelial and mesenchymal components may occur; if present, they should raise concern about the benign nature of the case.[8] AF may rarely progress to malignancy (ameloblastic fibrosarcoma – AFS).

In this case, AFS presents a benign epithelial and a malignant ectomesenchymal component.[2,9] An overexpression of Ki-67 immunoexpression (a proliferative nuclear cell marker) and Bcl-2 proteins in AFS, in association with histopathological features, may be useful markers to identify malignancy.[9]

Regarding the nature and biological behavior of tumors, the treatment for AF in 90% of the cases initially consists of a conservative surgical approach, and in 10% of the cases, radical surgery.[4] The most appropriate treatment for AF is still unclear. In this case, a conservative technique was proposed, especially because the patient was young, as corroborated by the literature. The patient's follow-up time is short to detect recurrences, however, a regular follow-up is maintained.

CONCLUSION

Although rare, ameloblastic fibroma may recur and progress to malignancy. The objective of this case report was to highlight the challenge involved in its correct diagnosis and treatment, considering AF lesion behavior, and emphasizes the importance of clinical and radiographic follow-up, which should be for long periods for early detection of possible recurrences.

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