

Ameloblastic Fibroma: A Rare Case Report with 7-Year Follow-Up

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SUMMARY

Introduction Ameloblastic fibroma (AF) is a rare benign odontogenic tumor that usually occurs in the first two decades of life. It affects adolescents and young adults and is found in the mandible and with a higher frequency in the posterior region of this segment. There are rare case reports with a long-term follow-up.

Case Outline We report the case of a 6-year-old boy with extensive ameloblastic fibroma in the mandible. Treatment consisted of enucleation and bone curettage, with the preservation of permanent teeth adjacent to the tumor. Clinical and radiographic follow-up of the patient over a period of 7 years showed no signs of recurrence or malignant transformation.

Conclusion Patients with AF should be under follow-up for prolonged periods of time, even in cases exhibiting a low proliferation index, because of the potential for recurrence and malignant transformation of this tumor.

Keywords: ameloblastic fibroma; odontogenic tumors; jaws; follow-up

INTRODUCTION

Ameloblastic fibroma (AF) is a rare tumor which accounts for 0.9 to 2.4% of all odontogenic tumors involving the jaw bones [1-4]. AF mainly affects adolescents and young adults and has a slight male predominance. Most tumors are found in the mandible, while a higher incidence is observed in the posterior region. Patients usually seek treatment because of a swelling in the jaw bones and delayed tooth eruption, although some cases are diagnosed incidentally during routine radiographic examination [5]. Radiographically, AF appears as a unilocular lesion with well-defined margins. Multilocular lesions are occasionally observed in more extensive cases. The lesions are frequently associated with impacted teeth [6, 7].

Histologically, AF is characterized by epithelial islands and cords whose center resembles the stellate reticulum of the enamel organ. The islands and cords are immersed in ectomesenchyme that mimics the dental papilla. The presence of a capsule is considered an uncommon feature of AF [8].

AF shows a high rate of recurrence [5], while malignant transformation is observed in some cases [9, 10]. These findings highlight the importance of the study of this tumor and the need for long-term follow-up. This study reports a case of extensive AF in the mandible of a child. No signs of recurrence were observed after 7 years of follow-up.

CASE REPORT

A 6-year-old boy was referred to a private clinic in Salvador, Bahia, Brazil, for evaluation of a swelling on the right side of the face. The parents reported no eating difficulties. The medical history revealed that the patient had thalassemia minor, but no clinical signs or symptoms were observed.

Extraoral physical examination showed a hard swelling in the mandibular body and angle on the right side, which caused a mild facial asymmetry. Intraoral examination revealed a swelling of the alveolar ridge in the right posteroinferior region and the absence of teeth in the same region, which led the patient to seek treatment (Figure 1A). Panoramic radiography showed the presence of a well-defined multilocular radiolucent area extending from the right deciduous first molar to the mandibular angle on the same side, causing the retention of teeth 85 and 46 (Figure 1B). The clinical suspicions included dentigerous cysts, keratocystic odontogenic tumor, and ameloblastoma. An incisional biopsy was performed and anatomopathological analysis showed odontogenic epithelial islands and cords amidst a cellular and embryonic stroma resembling the dental papilla. These findings were consistent with the diagnosis of AF. Two weeks later, the patient was submitted to general anesthesia for enucleation of the lesion, extraction of tooth 85, and bone curettage (Figure 1C). Macroscopic inspection of the tu-

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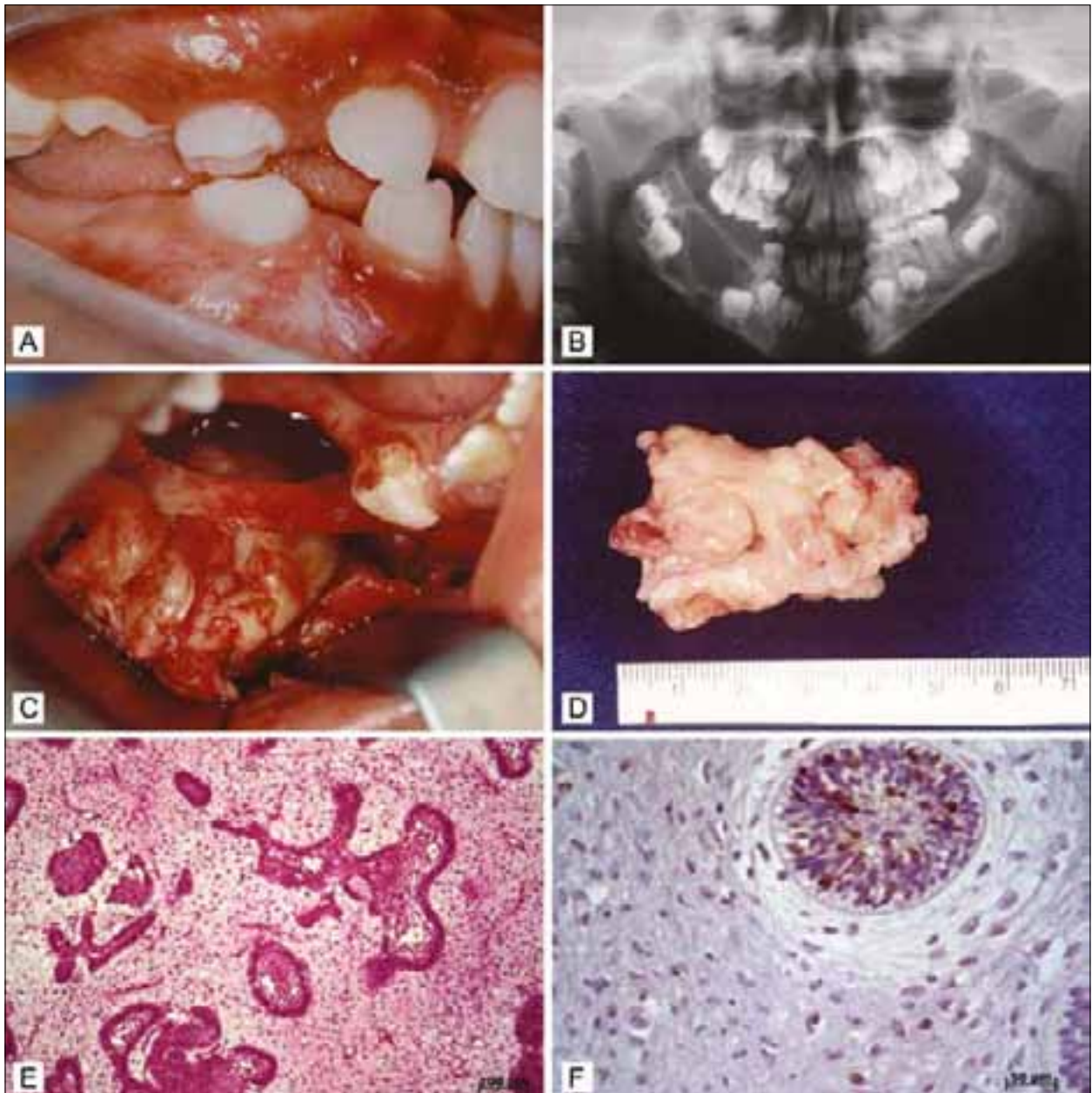


Figure 1. **A.** Intraoral view. Swelling of the alveolar ridge in the right posteroinferior region and absence of teeth. **B.** Panoramic radiograph showing a well-defined multilocular radiolucent lesion at the right side of the mandible. **C.** Intraoperative image obtained during surgical removal of the tumor. **D.** Macroscopic inspection of the tumor specimen measuring 45×40×10 mm. **E.** Presence of odontogenic epithelial islands and cords amidst a cellular and embryonic stroma resembling the dental papilla (HE, 10×). **F.** Immunohistochemical analysis showing the presence of few Ki-67-positive nuclei in the epithelial parenchyma.

mor specimen, which measured 45×40×10 mm, revealed an irregular surface and fibroelastic consistency (Figure 1D). Histopathological analysis confirmed the diagnosis of AF (Figure 1E). Immunohistochemistry detected few Ki-67-positive nuclei, especially in the epithelial parenchyma, indicating a low proliferation index of the tumor (Figure 1F).

No complications were observed during the postoperative period. The patient was followed up over a period of 7 years and no clinical or radiographic signs of recurrence were observed. Control panoramic radiographs, after 2 months, 1 and 5 years, showed the formation of new bone in the right posterior region of the mandibular body and complete eruption of teeth 46 and 47 (Figure 2). Com-

puted tomography scans obtained after 7 years revealed cortical bone integrity, complete formation of medullary bone, and complete eruption of tooth 46 which had been preserved during surgery. In addition, the bone architecture in the right posteroinferior region was preserved and no radiolucency was observed (Figure 3).

DISCUSSION

This study reported a case of AF involving the body and angle of the mandible in a 6-year-old child. Philipsen et al. [11] and Tomich [6] suggested that AF corresponds to a stage of maturation of ameloblastic fibro-odontoma and

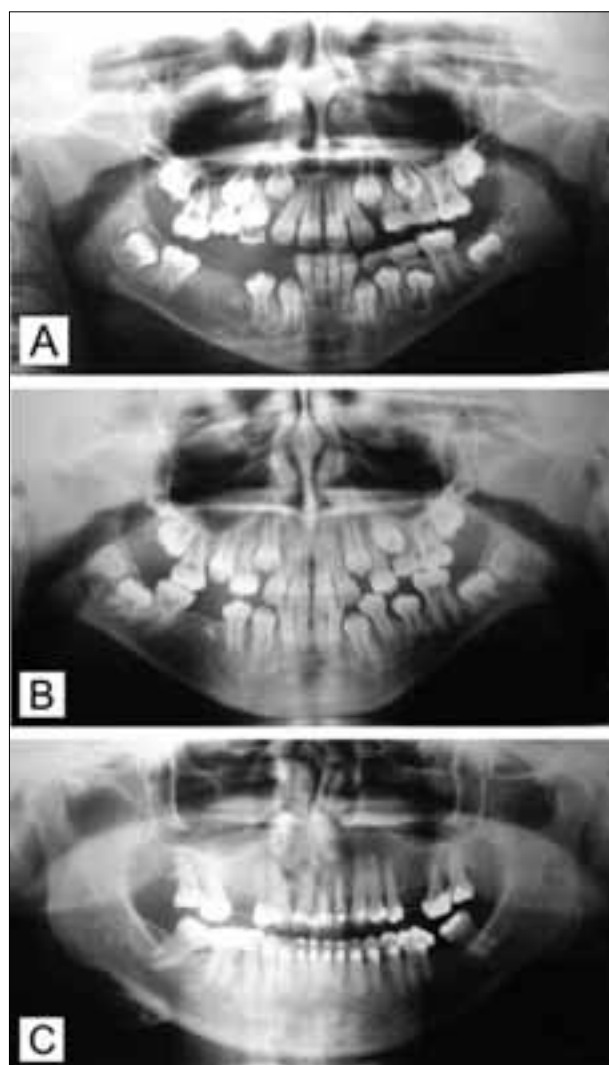


Figure 2. Control panoramic radiographs, after 2 months (A), 1 year (B) and 5 years (C), respectively, showing the formation of new bone in the right posterior region of the mandibular body and complete eruption of teeth 46 and 47.



Figure 3. Control computed tomograph scans obtained after 7 years. Note the integrity of cortical bone, absence of radiolucency, and preservation of bone architecture in the region.

odontoma. This hypothesis has been refuted since recurrent AFs show no evidence of maturation to ameloblastic fibro-odontoma or odontoma. In addition, AFs usually affect patients in the third decade of life when the process of odontogenesis is already completed [12].

Clinically, the patient presented with a mandibular swelling extending from the right deciduous first molar to the mandibular angle on the same side, causing a delay in tooth eruption. According to Kim and Jang [8], this swelling is caused by gradual expansion of the cortical plate during growth of the tumor. In addition, these authors have reported that AF shows slower growth than other odontogenic tumors, such as ameloblastoma, and does not tend to infiltrate bone. However, Pereira et al. [13] reported a case of rapidly growing AF in an 11-month-old infant, which was attributed to the small size of the jaw.

The patient had thalassemia minor, the heterozygous state of a hereditary genetic disease characterized by a reduction in beta globin chain synthesis, causing mild and silent microcytic and hypochromic anemia [14, 15]. There are no reports in the literature correlating this hereditary disease with AF, but this odontogenic tumor has been diagnosed in other diseases such as Goldenhar syndrome [16].

The histopathological features of the present case met the criteria for AF [12, 17]. The lesion consisted of odontogenic epithelial islands and cords amidst a cellular and embryonic stroma resembling the dental papilla. Usubutun et al. [18] also found extensive cystic formation as a result of a degenerative process, but this was not observed in the present case.

According to Cernelio and Vij [19] and Rao et al. [20], AF exhibits a low proliferation rate. This was confirmed in the present study by sparse immunostaining for Ki-67, especially in the parenchymal compartment. Evaluation of the proliferative potential of AF using immunohistochemical markers contributes to the understanding of tumor aggressiveness and to adequate surgical planning [8], as in the present case.

The initial diagnosis of the case included a dentigerous cyst, odontogenic keratocystic tumor and ameloblastoma due to the clinical and radiographic similarities between these lesions. The differential diagnosis of AF should include odontogenic lesions such as the dentigerous cyst, ameloblastoma, myxoma, and other mixed tumors [21, 22]. According to Nelson and Folk [23], despite many similarities, it is essential to differentiate AF from other mixed odontogenic tumors in view of its true neoplastic potential, possibility of recurrence and malignant transformation potential.

The present patient was submitted to conservative treatment considering that the lesion was a primary tumor and to avoid possible esthetic deformities. Consensus exists regarding the preference of conservative treatment of AFs, including enucleation of the tumor and removal of all teeth involving bone curettage [13, 24, 25]. According to these authors, en bloc resection results in significant morbidity and esthetic deformity and should be reserved for extensive tumors invading soft tissue and for recurrent cases.

The present patient was followed up for 7 years and no clinical or radiographic signs of recurrence or malignant transformation were observed after this period, demonstrating the success of treatment. Long-term follow-up of AF as done in the present study is important due to the possibility of recurrence and malignant transformation.

Follow-up of cases for up to 12 years has been reported [8, 13, 21, 25, 26].

In conclusion, patients with AF should be followed up for prolonged periods of time, even cases exhibiting a low proliferation index, because of the potential for recurrence and malignant transformation of this tumor.

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Амелобластни фибром: приказ ретког случаја са седмогодишњим периодом клиничког праћења

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КРАТАК САДРЖАЈ

Увод Амелобластни фибром је редак, бенигни, одонтогени тумор који се обично јавља до двадесете године живота. Обољење погађа адолесценте и младе одрасле особе, а захвата доњу вилицу, и то чешће постериорну регију овог сегмента. Прикази болесника са дугорочним клиничким праћењем су ретки.

Приказ болесника Приказујемо шестогодишњег дечака са екстензивним амелобластним фибромом доње вилице. Болесник је лечен применом хируршке ресекције и киретаже

кости, уз очување сталних зуба у близини тумора. Клиничко и радиолошко испитивање болесника током седмогодишњег периода није показало знаке рецидива или малигне трансформације.

Закључак Болеснике оболеле од амелобластног фиброма потребно је дугорочно надгледати, чак и у случајевима с ниским индексом пролиферације, с обзиром на могућу појаву рецидива и малигну трансформацију овог тумора.

Кључне речи: амелобластни фибром; одонтогени тумори; вилица; клиничко праћење

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