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Ameloblastic Fibrossarcoma In The Mandible: A Case Report

Fibrossarcoma Ameloblástico Na Mandíbula: Um Relato de Caso

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ABSTRACT
Ameloblastic fibrosarcoma is a rare malignant neoplasm, commonly found in the mandible and with higher prevalence in men. Extensive expansion and increased volume are the main clinical characteristics observed. **Objective**: To report a case of ameloblastic fibrosarcoma in a 73-year-old female patient, presenting with swelling in the chin area and deformity on the right side of the mandible. **Methodology**: To confirm the diagnosis, two incisional biopsies and radiographic examinations were performed. **Results**: The first histopathological examination showed proliferation of stroma and odontogenic epithelium with benign neoplasia, and negative immunohistochemical staining for Ki-67. The second examination revealed an ameloblastic appearance, confirming the diagnosis of ameloblastic fibrosarcoma. Treatment consisted solely of complete resection of the mandible. **Conclusion**: The rarity of this tumor underscores the importance of disseminating the clinical, radiographic, and histological findings of this case. Accurate diagnosis is crucial for early and conservative treatment, determining factors for patient cure and survival.,

Keywords: Odontogenic tumors, fibrosarcoma, malignant neoplasms, mandible, case report.

RESUMO
O fibrossarcoma ameloblástico é uma neoplasia maligna rara, comum em mandíbula e com maior prevalência em homens. Extensa expansão e aumento de volume são as principais características clínicas encontradas. **Objetivo**: relatar um caso de fibrossarcoma ameloblástico em uma paciente do sexo feminino, 73 anos, com abaulamento na região do mento e deformação do lado direito da mandíbula. **Metodologia**: para confirmar o diagnóstico, foram realizadas duas biópsias incisionais e exames radiográficos. **Resultados**: o primeiro exame histopatológico apresentou proliferação de estroma e epitélio odontogênico com neoplasia benigna, e exame imuno-histoquímico negativo para Ki-67. O segundo exame demonstrou aspecto ameloblástico, confirmando o diagnóstico de fibrossarcoma ameloblástico. Para o tratamento, optou-se apenas pela ressecção completa da mandíbula. **Conclusão**: a raridade deste tumor explica a importância da divulgação dos achados clínicos, radiográficos e histológicos deste caso. O correto diagnóstico é primordial para o tratamento precoce e conservador, fatores determinantes para cura e sobrevida dos pacientes.

Palavras-chave: Tumores odontogênicos, fibrossarcoma, neoplasias malignas, mandíbula, relato de caso.

INTRODUCTION

Ameloblastic fibrosarcoma (AFS) is characterized as a malignant tumor primarily found in the mandible, predominantly affecting men in their third and fourth decades of life[1]. Considered extremely rare, as of 2023, only 107 cases had been reported in the literature[2].

Previously, ameloblastic fibrosarcoma was termed ameloblastic fibroma (AF). However, in 2022, the WHO described ameloblastic fibrosarcoma as a distinct lesion that may develop with ameloblastic fibroma as its initial stage, or independently[2,3].

The main characteristic of the lesion is intraoral and extraoral swelling due to tumor expansion. Additionally, patients may experience occlusal pain, facial pain, dysphagia, paresthesia, dysesthesia, and ulcers. Radiographically, it appears as a uni- or multilocular cystic lesion with poorly defined borders, significant bone expansion, and destruction. Therefore, an incisional biopsy is recommended to confirm the diagnosis, along with histological, radiographic, and clinical data. Furthermore, immunohistochemical marker tests are indicated, with Ki-67 being the primary diagnostic test[3,4,5].

The histological features of ameloblastic fibrosarcoma exhibit structural similarities to those of ameloblastic fibroma. However, ameloblastic fibrosarcoma includes a benign epithelial tissue component alongside malignant mesenchymal tissue. Moreover, this tumor lacks sarcomatous content, unlike ameloblastic fibroma[4,6,7].

Due to its aggressiveness, conducting an epidemiological study of this rare tumor is essential to improve diagnostic precision. This enables healthcare professionals to plan treatments that optimize clinical outcomes and enhance patients' quality of life[6]. Additionally, raising awareness through campaigns about this tumor is crucial. Since it primarily develops due to genetic factors, regular consultations with physicians and dentists can increase the chances of early detection[7].

Based on the above, this research aims to report a clinical case of Ameloblastic Fibrosarcoma located in the mandible, diagnosed at the dental clinic of Faculdade de Ilhéus – Ilhéus/BA, in a 73-year-old female patient.

METHODOLOGY

This report was approved by the Research Ethics Committee of the Federal Institute of Education, Science, and Technology of Bahia - IF BAIANO, under protocol number 69006723.4.0000.0249. All ethical principles of honesty, reliability, objectivity, impartiality, care, respect, truthfulness, and responsibility were adhered to.

A 73-year-old retired white female patient attended the Clinic of Semiology and Diagnosis II at Faculdade de Ilhéus in April 2019, presenting with a swelling in the anterior region of the mandible, specifically at the chin, without pain symptoms (Fig. 1).

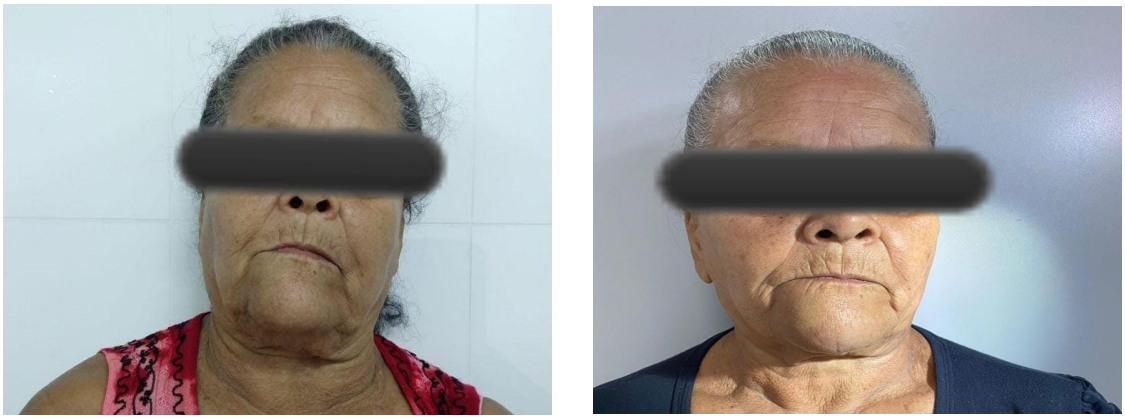

Figure 1 - Physical appearance of the patient in 2019 and 2021
Source: author

During the medical history, it was noted that the patient was hypertensive and taking medication for control. On extraoral examination, facial deformation on the right side of the mandible was observed, with no changes in lymph nodes. On intraoral examination, there was swelling in the vestibular region, involving from the left lower second premolar (tooth 35) to the right lower second premolar (tooth 45) (Fig. 2).

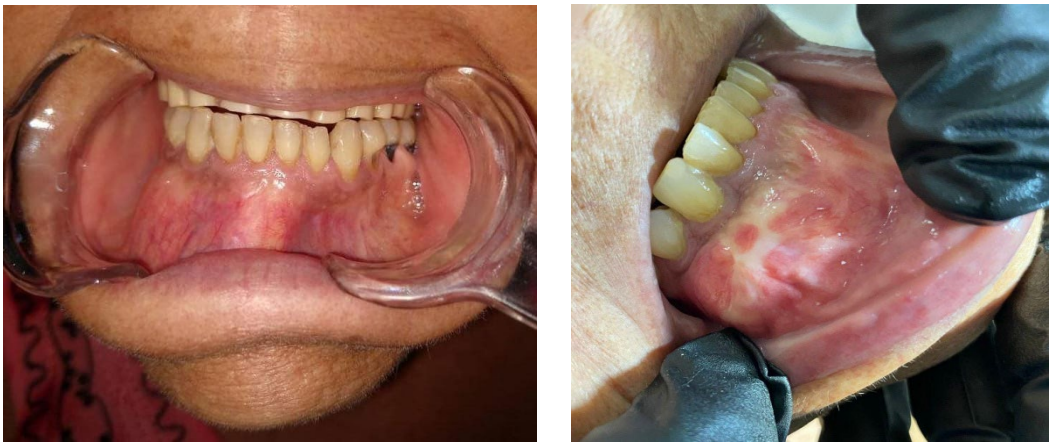

Figure 2 - Comparison of the clinical appearance of ameloblastic fibrosarcoma in 2019 and 2021
Source: author

Additional exams were requested, including panoramic radiography, cone beam computed tomography (CBCT), and laboratory tests.

The panoramic radiograph revealed an extensive radiolucent lesion with imprecise borders in the mental region. There was moderate alveolar bone loss (3 to 4 mm of resorption), osteosclerosis adjacent to the root apex of tooth 36, and the lesion likely extended from tooth 35 to 45. There was no root resorption in the involved teeth, but there was displacement of the roots of teeth 42 and 43, and slight expansion of the inferior mandibular cortex (Fig. 3).

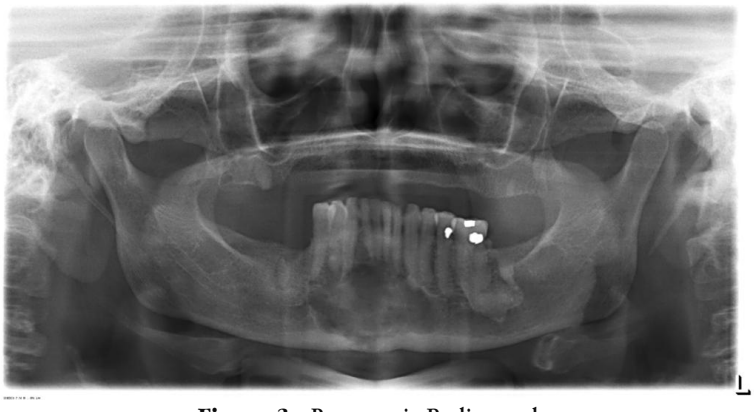

Figure 3 - Panoramic Radiography
Source: author

The computed tomography of the mandible revealed an extensive hypodense lesion with vestibular bone expansion and thinning of the lingual cortex, with cortical breakthrough, extending from the root of tooth 35 to the root of tooth 45, and with intact roots. There was

displacement of the roots of teeth 42 and 43, osteosclerosis in the region corresponding to tooth 37, residual root of tooth 46, periapical changes, and multiple missing teeth (Fig. 4).

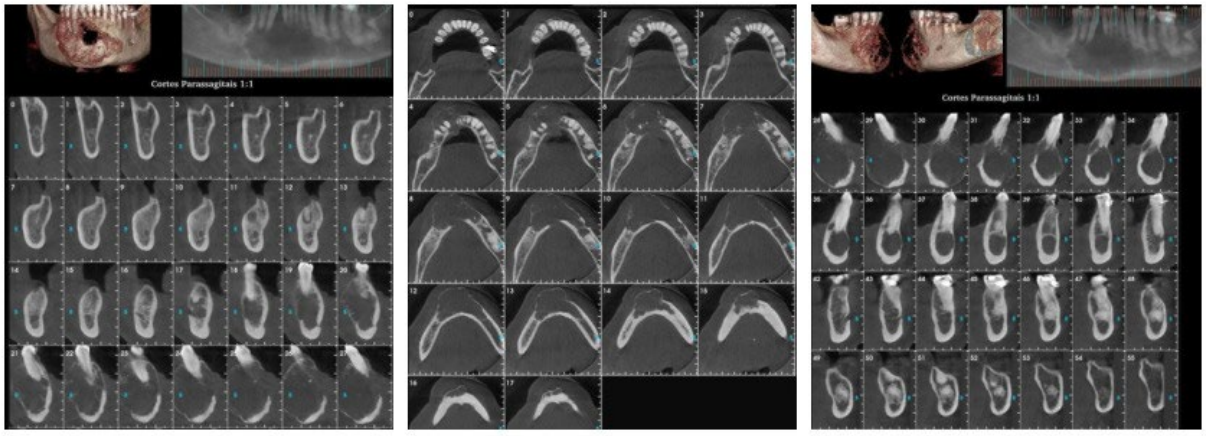

Figure 4. Computed tomography
Source: author

The patient was anesthetized with infiltrative anesthesia, followed by an aspiration biopsy using a 25-gauge needle, which yielded a negative result. Subsequently, an incisional biopsy of the lesion was performed using a no. 15c blade. The specimen was fixed in 10% buffered formalin and sent to the Pathology Institute of Ilhéus for further examination.

In the histopathological examination, fragments of connective tissue stroma were observed interspersed with proliferation of spindle-shaped cells and clusters of epithelial cells. The result indicated proliferation of odontogenic stroma and epithelium (Fig. 5).

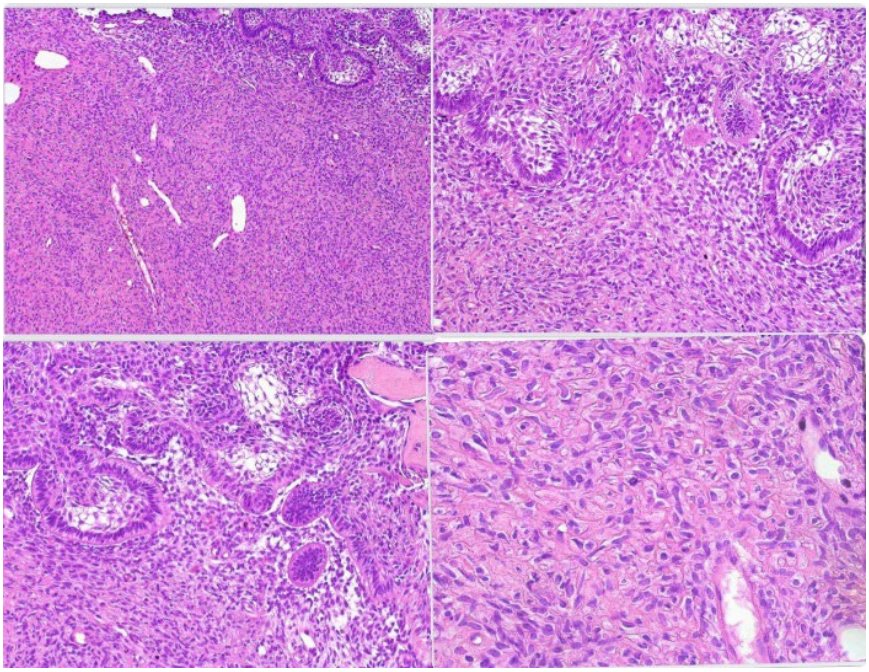

Figure 5 - Histopathological characteristics of ameloblastic fibrosarcoma
Source: author

In the absence of a conclusive diagnosis, the paraffin block and histological slide were sent to another laboratory at SLMandic University in Campinas, SP. The result indicated a benign odontogenic neoplasm, with a negative immunohistochemical test for Ki-67.

Com the end of the semester in 2019, the patient's treatment resumed in February 2020. However, due to the COVID-19 pandemic and suspension of classes, it was not possible to continue treatment. In September 2021, the patient returned to the clinic and reported weight loss and was diagnosed with osteopenia through bone densitometry. On intraoral evaluation, an increase in the size of the lesion was observed. A new aspiration biopsy using a 25-gauge needle yielded a negative result once again. Subsequently, a new incisional biopsy was performed using a no. 15c blade. During the surgical procedure, rupture of the vestibular bone cortex was observed, and the lesion was found to be in contact with soft tissue. The histopathological specimen was placed in a container with 10% formalin and sent to the pathology laboratory at the School of Dentistry, UNICAMP.

No descriptive report, an odontogenic neoplasm predominantly of ectomesenchymal origin was observed, with some islands of ameloblastic epithelial appearance. There was noted presence of hypercellularity, nuclear pleomorphism, and mitotic figures in the ectomesenchymal portion. Immunohistochemical staining for Ki-67 showed low activity.

Clinical, histopathological, and radiographic findings indicated the diagnosis of Ameloblastic Fibrosarcoma; however, suggesting that the final diagnosis should be made upon analysis of the surgical specimen.

The patient was referred through the Unified Health System (SUS) to Aristides Maltez Hospital in Salvador (BA) for consultation with a head and neck surgeon. She underwent complete resection of the mandible and is currently under follow-up, without the need for chemotherapy or radiotherapy.

DISCUSSION

Odontogenic fibrosarcoma or ameloblastic fibrosarcoma is a rare tumor classified within the group of malignant odontogenic tumors. In 70% of cases, ameloblastic fibroma (AF) serves as the initial stage of this tumor. However, this lesion can develop without presenting AF as the initial stage. This tumor represents only 2% of odontogenic tumors, underscoring its rarity[1,5].

The first description of this tumor occurred in 1887, where authors described five other types of tumors of the maxilla[12]. Since then, ameloblastic fibrosarcoma (AFS) has been known by various names, including dentinosarcoma ameloblástico (when dentin deposition is identified) and odontosarcoma ameloblástico (when both dentin and enamel deposition are identified)[2,3]. However, in 2022, the World Health Organization (WHO) defined AFS as a unique tumor distinct from these previous classifications.

It is a tumor that mainly affects men between the second and third decades of life, with a mean age of 26 years, and the posterior region of the mandible being the primary site for the lesion. However, new reports indicate a significant number of AFS cases in the maxilla, particularly in the palate and premolar regions[1,9].

The most predominant clinical features of AFS include intraoral and extraoral swelling, occlusal pain, facial pain, and dysphagia. Paresthesia, dysesthesia, and ulcers may also be present. The extensive expansion of the lesion through the mandible or maxilla demonstrates an important clinical characteristic of the condition. Therefore, an incisional biopsy is recommended for confirming the diagnosis[3,5]. In this case study, the patient was an elderly woman in her eighth decade of life, with the condition affecting her mandible. She presented with swelling extending from the left premolar to the right premolar region of the vestibule. Following the COVID-19 pandemic, the patient returned for diagnostic closure and exhibited, in addition to the aforementioned characteristics, weight loss, osteopenia, and an increase in the size of the lesion.

Histologically, fibroma ameloblastic and ameloblastic fibrosarcoma share similar

structures. They are odontogenic tumors of ectomesenchymal origin, resembling dental papilla, with epithelial tissue and islands similar to enamel organ and dental lamina, but without hard tissue formation[6]. The main difference between FA and FAS lies in the mesenchymal cells, with FA having a sarcomatous content[4]. Additionally, FAS presents benign epithelial tissue alongside malignant mesenchymal tissue, making FAS a malignant version of FA, where the tumor loses its epithelial tissue during its progression[6,7]. Another histological characteristic of FAS that allows differentiation from other malignant tumors is the hypercellular connective tissue with polygonal and spindle-shaped stromal cells, along with mitotic cells, peripheral cuboidal and columnar cells with hyperchromatic nuclei[10]. Additionally, the epithelium shows ameloblastic islands, and the malignant component may exhibit hypercellularity, pleomorphism, and mitotic figures[11].

Although rare, hormonal changes during pregnancy can increase the chances of developing both malignant and benign tumors, with lymphoma, leukemia, and melanoma being more common. However, a study conducted in 2013, focusing on a pregnant Nigerian woman diagnosed with FAS, reported the disease's progression from a pre-existing ameloblastic fibroma[2].

For a correct diagnostic closure, the use of immunohistochemical marker tests is recommended, with the presence of Ki-67 being the main marker for diagnosis. Other markers can be used, such as p53, Bcl-2, VEGF-A, genetic polymorphisms, and microRNA 99[3,4,13,14].

Two histopathological examinations were performed for diagnosis in this case. The first examination revealed fragments of connective stroma, clusters of epithelial cells, and proliferation of fusiform cells. It yielded a result of stromal and odontogenic epithelial proliferation, but was inconclusive. Therefore, it was sent to another laboratory where it was diagnosed as a benign odontogenic neoplasm. The immunohistochemical examination for Ki-67 resulted negative. In the second histopathological examination, an ectomesenchymal odontogenic neoplasm was observed, with islands of ameloblastic epithelium, nuclear pleomorphism, hypercellularity, and mitosis in the ectomesenchymal portion. Additionally, Ki-67 staining was low. These characteristics indicate that the ameloblastic fibrosarcoma in question did not evolve from an ameloblastic fibroma. Its development was unique, displaying both malignant and benign features.

In radiographic images, ameloblastic fibrosarcoma presents as a uni- or multilocular cystic lesion with poorly defined borders and significant expansion[5,14,15]. Root resorption (bone destruction) is also notably present[6,16]. Through computed tomography (CT), a soft tissue mass causing bone expansion and cortical bone perforation can be observed[7]. For this patient, panoramic radiography and computed tomography were requested. Panoramic radiography revealed root resorption with extensive, radiolucent, and poorly defined borders, as described in the literature. Computed tomography showed an extensive hypodense lesion with bone expansion and cortical breakthrough, consistent with literature findings. Additionally, osteosclerosis and periapical changes can be observed.

Studies demonstrate that the average time for the evolution of an ameloblastic fibroma into an ameloblastic fibrosarcoma, without oncological treatment, is 55 months, with high rates of disease recurrence[6]. Although it is a malignant tumor, it does not exhibit metastatic characteristics or frequent recurrence, and has a good prognosis. However, it carries a 20% risk of mortality due to the aggressiveness of the lesion.[10].

Segmental resection, also known as surgical excision with wide clear margins, where the entire extent of the lesion is removed, has emerged as a more relevant and recommended approach for treating AFS, with a 70.5% disease regression rate. Long-term patient follow-up is crucial for effective treatment, although this disease does not typically recur. However, chemotherapy or radiotherapy, or a combination of both, can also be employed, with radiotherapy being the preferred choice in these cases.

Studies demonstrate that radiotherapy is indicated to prevent recurrence, progression of the lesion, or local invasion. Soltany (2020) reported the use of doxorubicin and olaratumab

as adjuvant therapies in treating this tumor. However, there is insufficient evidence to guarantee the success of these adjuvant therapies, necessitating further long-term analysis. In this case, the patient underwent complete mandibular resection without the need for chemotherapy or radiotherapy. She is currently under medical follow-up to prevent disease recurrence.

Although this study enabled the diagnosis and treatment, it is essential to highlight the limitations and challenges associated with this lesion. Many cases present ameloblastic fibroma as the initial stage of AFS; however, the transformation process is not fully elucidated in the literature. The expression of Ki-67, a marker of tumor proliferation and supportive in cancer diagnosis, may also be inconclusive in cases of AFS. Age and gender vary significantly, although there is a higher prevalence in young men. Therefore, the scarcity of conclusive information underscores the importance of further studies on this tumor. Understanding all clinical implications of this lesion allows for more accurate diagnosis and appropriate treatment.

CONCLUSION

Ameloblastic fibrosarcoma is a rare lesion found predominantly in the jaw, especially the maxilla. It affects both men and women of all age groups, with a higher incidence observed in men in their third decade of life.

Diagnosis is typically confirmed through a combination of histopathological, radiographic, and clinical data. Treatment primarily involves complete surgical resection of the lesion in most cases.

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