CASE REPORT

CLINICAL PATHOLOGICAL CHARACTERISTICS AND TREATMENT OF UNICYSTIC AMELOBLASTOMA: CASE REPORT

CARACTERÍSTICAS CLÍNICO-PATOLÓGICAS E TRATAMENTO DO AMELOBLASTOMA UNICÍSTICO: RELATO DE CASO

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Resumo

O ameloblastoma é um tumor odontogênico benigno de origem epitelial, localmente invasivo, de curso lento e que apresenta consideráveis taxas de recidivas. É considerado o tumor odontogênico benigno mais relevante clinicamente, sendo atualmente classificado como "ameloblastoma", "ameloblastoma unicístico", "ameloblastoma periférico ou extraósseo" e "ameloblastoma metastatizante". A variante unicística é a segunda mais prevalente, respondendo por aproximadamente 15% de todos os ameloblastomas, mostrando características singulares especialmente em pacientes mais jovens. Este trabalho tem por finalidade aclarar as características desta variante do ameloblastoma a partir do relato de caso do atendimento de um paciente adulto jovem acometido pelo tumor na mandíbula, abordando os aspectos clínicos, imaginológicos, histopatológicos, terapêuticos e prognósticos envolvidos. Neste caso clínico, o tratamento de escolha para o ameloblastoma unicístico foi, inicialmente, a marsupialização da lesão e posteriormente a enucleação com ostectomia periférica e exodontia dos dentes envolvidos. Esta opção de tratamento demonstrou bons resultados, diminuindo os danos ao paciente quando bem indicada, para isso é essencial um diagnóstico precoce e assertivo associando características clínicas e histopatológicas. O paciente segue em proservação cautelosa, visto que o padrão de proliferação do tipo mural requer uma maior atenção dada a sua maior possibilidade de recidiva.

Palavras-chave: Ameloblastoma.Tumores Odontogênicos. Osso e Ossos. Mandíbula.

Abstract

Ameloblastomas are a type of epithelial benign odontogenic tumor which is locally invasive, slow-growing and presents considerable recidive rates. It is considered the most clinically relevant benign odontogenic tumor, currently being classified as "ameloblastoma", "unicystic ameloblastoma", "peripheral or extraosseous ameloblastoma" and "metastatic ameloblastoma". The unicystic type is the second most prevalent ameloblastoma, occurring in approximately 15% of all ameloblastomas, showing specific characteristics, especially in younger patients. This study verifies the characteristics of this ameloblastoma variant by analyzing the case report of the care of a young adult patient affected by jaw tumor, clinical localization, imaginological histopathological, therapeutic and prognostic aspects involved. In this case, the initial treatment was the marsupialization of the lesion, and afterwards the enucleation with peripheral ostectomy and extraction of the affected teeth. This treatment option has shown good results, decreasing the patients damage when properly indicated, for this, an early and assertive diagnosis is essential, associating clinical and histopathological characteristics. The patient is kept under careful follow-up, since the proliferation pattern of mural type requires more attention given its higher chances of recurrence.

Keywords: *Ameloblastoma. Odontogenic Tumors. Bone and Bones. Mandible.*

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INTRODUCTION

Odontogenic tumors are, by definition, neoplasms, often benign, usually resulting from genetic changes during odontogenesis(1). Ameloblastomas are jaw diseases that usually show indolent and quietly progressive behavior, commonly leading to major tissue losses in affected patients. Similar to all diseases, early diagnosis is essential in order for less comorbidity to occur in patients affected by ameloblastoma. In this context, its discovery occurs accidentally in imaging exams or through some noticeable volumetric increase in the jaws, with the dentist being the professional that is most involved in the process. Ameloblastomas represent about 10% of all benign odontogenic tumors (2) and approximately 1% of all cysts and tumors that affect the mandible and maxilla(3). Hinds et al. and Gorlin et al. reported that the component cells of ameloblastomas may arise from the epithelial lining of an odontogenic cyst, dental lamina, enamel organ, stratified squamous epithelium of the oral cavity or displaced epithelial remains, representing tumors in soft tissues without bone involvement(4, 5).

In 2005, ameloblastomas were reclassified by the World Health Organization (WHO) into solid or multicystic ameloblastoma, unicystic ameloblastoma, peripheral or extrosseous ameloblastoma, and desmoplastic ameloblastoma(6). Subsequently, in 2017, the WHO published a reclassification of lesions into the following types: ameloblastoma, unicystic ameloblastoma, peripheral or extraosseous ameloblastoma and metastatic ameloblastoma. In this reclassification, the terms "solid/multicystic" were preferably deprecated in favor of the simple nomenclature "ameloblastoma" or "conventional ameloblastoma". Desmoplastic ameloblastoma has been reclassified as a histological subtype, and no longer as an independent clinical entity as it once was(7, 8). It is also worth mentioning the insertion of the "metastatic ameloblastoma", which despite the suggestion of malignancy in its name, accounts for a conventional ameloblastoma that may occasionally have metastatic spread to distant regions, such as lungs and bones.

Unicystic ameloblastomas particularly occur more frequently in younger patients, with about 50% of all these tumors diagnosed during the second decade of life. Neville et al. report that the average age is 23 years (9). More than 90% of unicystic ameloblastomas affect the mandible, usually in the posterior regions. The lesion is often asymptomatic, although large lesions can cause a painless swelling in the gnathic bones(9). Histologically, unicystic ameloblastoma can present three proliferation patterns: luminal, intraluminal and mural(10). Such subtypes can interfere with biological behavior, treatment and prognosis of the lesion(11). In this context, the luminous and intraluminous variants would respond positively to the treatment of enucleation with peripheral ostectomy, while unicystic ameloblastomas with a mural variant would be better treated in a more aggressive behavior, justifying the possibility of surgical resections with a safety margin.

This study thus presents the case of a patient affected by mandibular unicystic ameloblastoma, highlighting all the clinical, propaedeutic, imaginological, histopathological, therapeutic and prognostic aspects involved.

CASE REPORT

The subject was a PHRS patient, male, leucoderma, 26 years old, born in Carmo do Paranaíba, Minas Gerais, attending the Stomatology Clinic of the School of Dentistry of the Federal University of Uberlândia (FOUFU), referred by the Basic Health Unit of his hometown due to the presence of mandibular lesion on the right side. In the anamnesis, he reported being asymptomatic and also not knowing the time of the disease's evolution. In the medical and dental history, the patient reported frequent gingival bleeding, in addition to routine headaches, a history of asthma/bronchitis and hepatitis.

Upon extraoral physical examination , no significant changes were noted. During intraoral physical examination, a slight bulge was identified in the buccal and lingual re-

gion of the lower molars on the right side, in addition to slight sensitivity to palpation (Figure 1). The patient was carrying a panoramic radiograph showing a single, radiolucent, unilocular lesion, with well-defined edges, extending from the mandibular on the right side involving the lower right second molar (tooth 31) with root resorption and the mesio-angled (tooth 32), up to the uppermost ipsilateral ascending branch, close to the coronoid process (Figure 2).

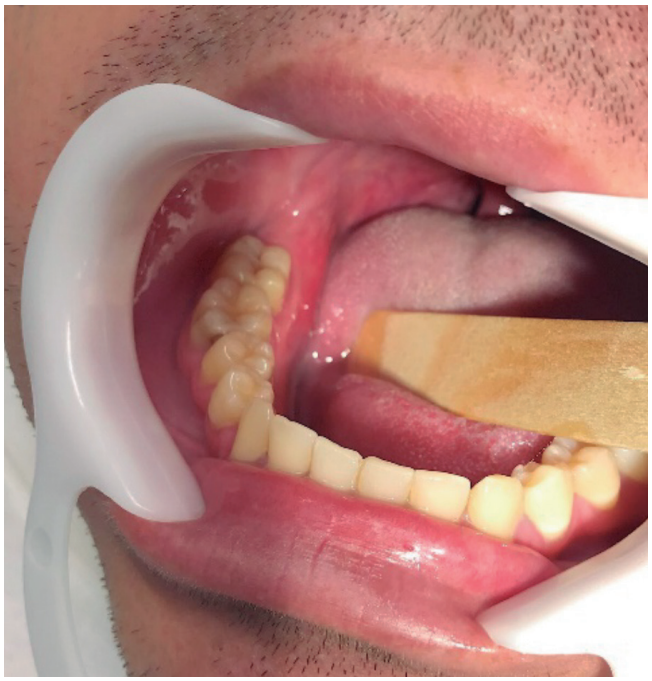


Figure 1 - Initial intraoral photo highlighting slight buccal and lingual bone expansion next to the lower molars on the right side, without tooth 32.

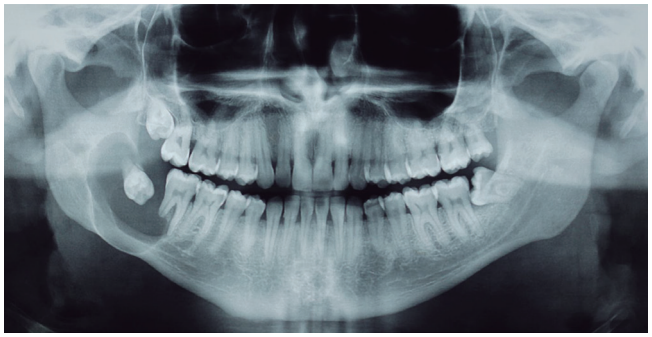


Figure 2 - Initial panoramic radiograph showing extensive intraosseous lesion involving tooth 32 and resorption of the distal root of the tooth 31.

In order to obtain better details of the lesion, Cone-Beam Computed Tomography (CBCT) was performed to visualize the well-defined, hypodense, unilocular lesion, with approximately 3.5 centimeters in its largest diameter (Figure 3).

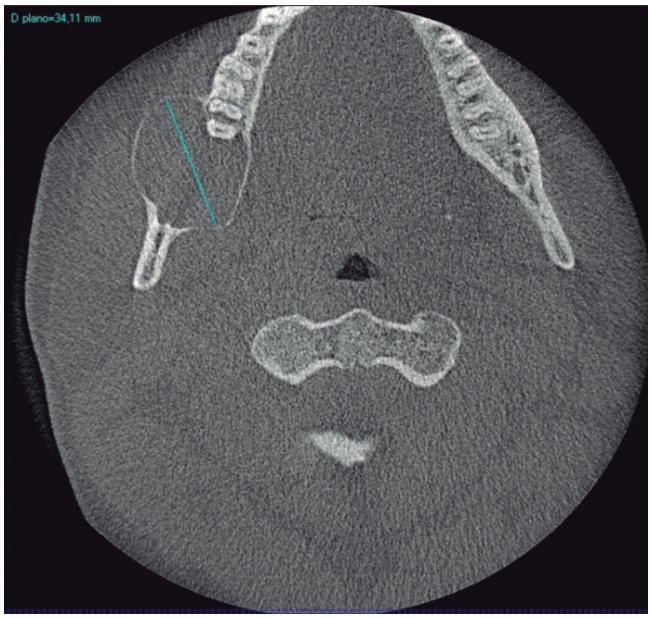


Figure 3 - Axial tomographic section showing a hypodense lesion of approximately 3.5 cm with thinned cortices.

In view of the clinical findings, an aspiration puncture was performed, resulting in positive for yellow-citrus content (Figures 4A and 4B). In the same intervention, an incisional biopsy was performed with the collection of tissue specimen and lesional decompression in order to promote dimensional regression. Thus, the hypotheses of diagnosis suggested were Unicystic Ameloblastoma, Keratocyst and Dentigerous Cyst. The patient was released and instructed to perform oral hygiene with irrigation of the lesion using 0.12% chlorhexidine or saline, in addition to requiring monthly returns for clinical follow-up and quarterly returns for clinical-imaging follow-up. The material obtained was stored in 10% formaldehyde and sent to the FOUFU Oral Pathology Laboratory for anatomopathological examination.

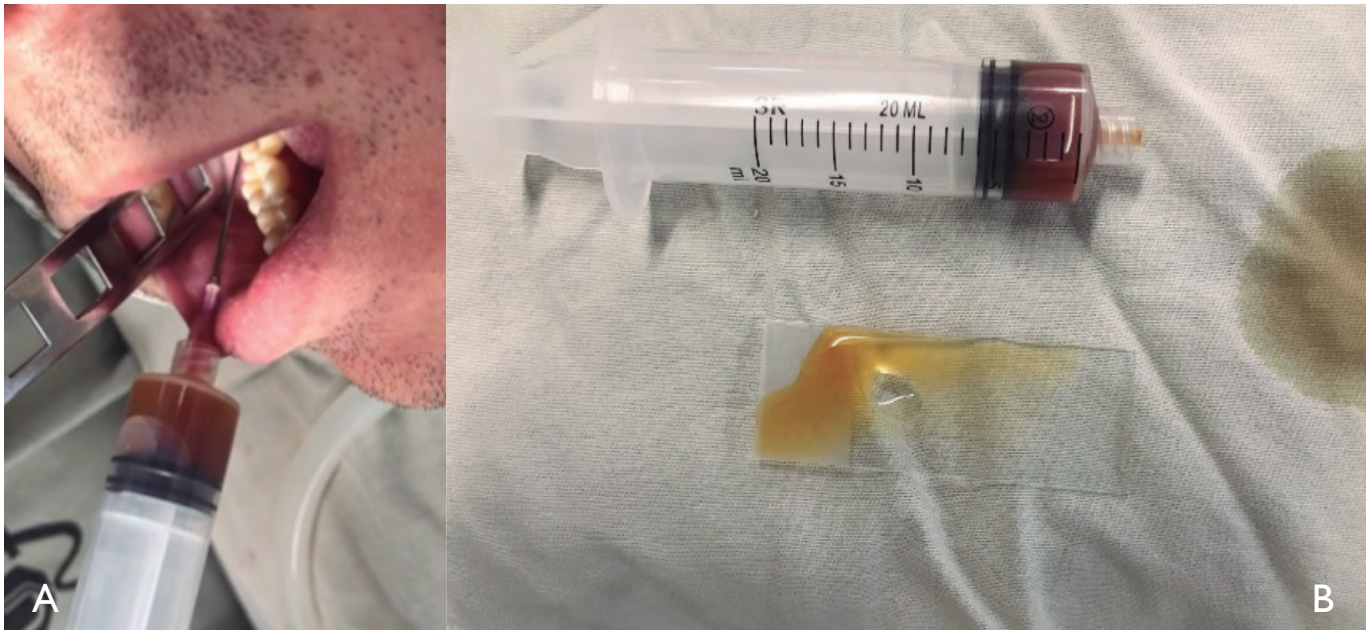


Figure 4 - (A) To the left, the aspiration puncture of the positive lesion for liquid; (B) To the right, the aspirated content with a citrus-yellow appearance.

The histological sections resulting from the incisional biopsy were stained in Hematoxylin-Eosin (HE) and showed fragments of lesions with capsular structure, being partly covered by stratified epithelium, with irregular thickness and flattened cells. In other regions, there was a proliferation of ameloblastic epithelium delimiting follicles or plexiform arrangements, some of which showed adenoid structures. Within these cell masses, the cells were spindle-shaped, stellar, with variations in cell density along the lesional parenchyma. It was found that clear cells predominated throughout the parenchyma. The picture completes the presence of regional mucosa without significant pathological changes (Figures 5A and 5B). These findings confirmed the diagnosis of Ameloblastoma which, when associated with the clinical information obtained, rendered the unicystic variant of the mural type as the best characterized for the case in question.

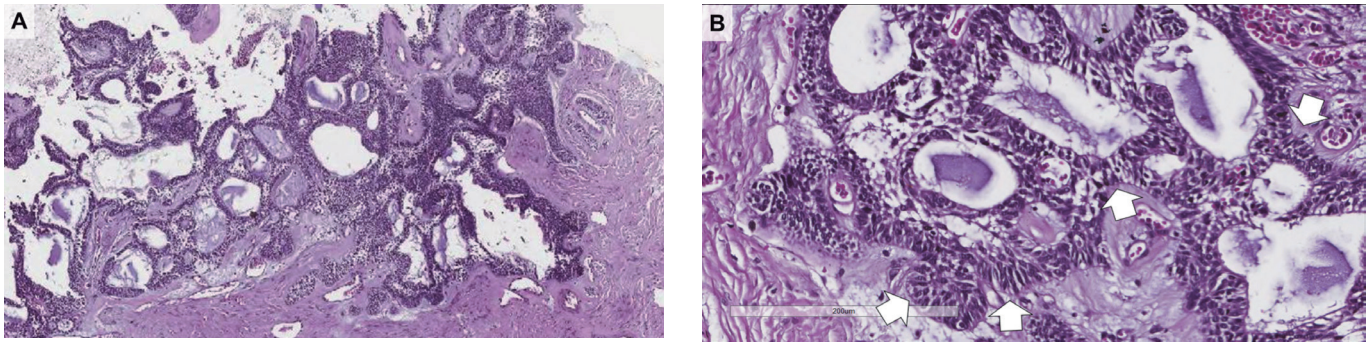
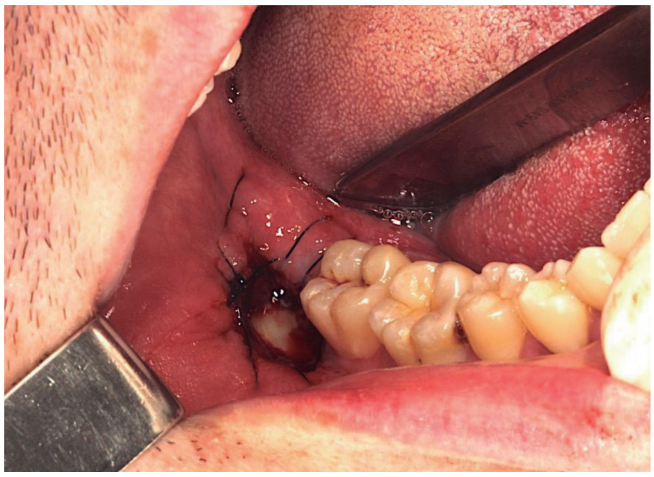


Figure 5 - (A) Photomicrography in lower magnification showing a fragment of a cystic capsule covered by ameloblastic epithelium, which then proliferates forming strands of neoplastic epithelial cells in the connective tissue wall, presenting a predominantly plexiform pattern. Hematoxylin and eosin, original 5X magnification; (B) Photomicrography in greater magnification highlighting the strands of neoplastic odontogenic epithelium, peripherally composed of tall columnar cells, characterized by polarized nucleus and clear cytoplasm, similarly to pre-ameloblasts (arrows). In some areas, these cells form rounded, adenoid-looking structures. In the center, it is possible to observe loosely arranged cells, sometimes spindle-shaped, sometimes star-like, occasionally showing cystoid degeneration. Hematoxylin and eosin, original 20X magnification.

The patient was followed up, showing good general health in return visits. Two months after the decompression surgery, the access made to irrigate the lesion was closed, requiring a new decompression procedure (Figure 6). Four months after initial decompression, the panoramic radiograph showed satisfactory centripetal bone formation with increased thickness of the mandibular basilar. Despite the good evolution, the team considered it prudent to wait approximately 2 more months for the patient to be reevaluated with the possibility of scheduling the final excision of the injury. After approximately 6 months of the first marsupialization, a new panoramic radiograph showed lesion regression with evident bone neoformation, allowing the final and definitive surgery to be conducted. Thus, the patient was then referred to the Buccomaxillofacial Surgery and Traumatology outpatient clinic, where surgery in the operating room was considered, under local anesthesia and conscious sedation, to curett the lesion with vigorous peripheral ostectomy and removal of the affected teeth, 31 and 32 (Figure 7). All tissues removed were again sent to the FOUFU Oral and Maxillofacial Pathology Laboratory.



Figue 6 -New marsupialization of the lesion performed seeking progressive decompression.

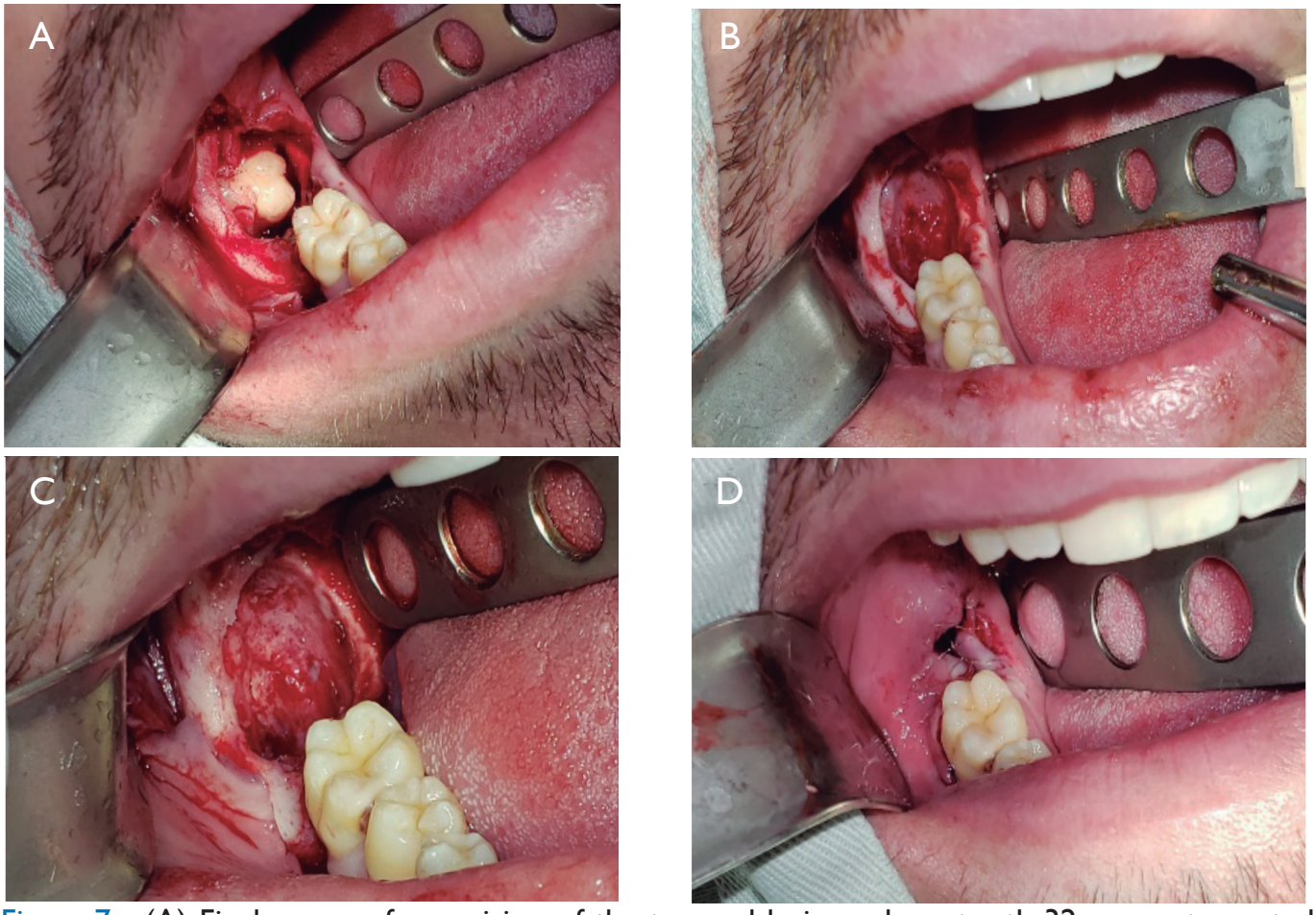


Figure 7 - (A) Final surgery for excision of the tumoral lesion, where tooth 32 was not erupted in the oral cavity, after tooth 31 was extracted; (B) Bone cavity after enucleation of the lesion and removal of tooth 32; (C) Cavity formed after peripheral ostectomy; (D) Immediate post-surgical appearance with positioned suture.

The macroscopy consisted of two molar teeth, with soft tissue adhered to the crowns (restricted to the cemento-enamel junction) and evident cystic space. In addition, fragments of soft tissue with brownish color, rubbery consistency, smooth luminal surface and cupuliform appearance were sent. Then, the microscopy of the final surgery confirmed the diagnosis of a unicystic ameloblastoma with mural variant.

Currently, the patient is in the postoperative period, asymptomatic, and undergoing clinical-imaging follow-up, without any sign of recurrence, as shown in the panoramic radiograph performed three months after the surgery (Figure 8).

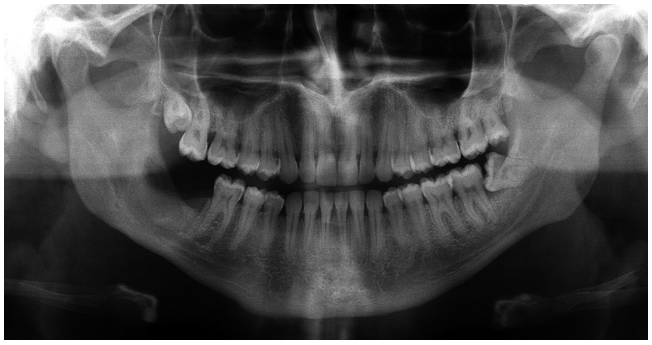


Figure 8 - Panoramic radiograph 3 months after surgery.

DISCUSSION

Several studies reporting cases of ameloblastoma are available in the literature, showing that, according to their behavior, they can be classified as: ameloblastoma, unicystic ameloblastoma and peripheral or extraosseous ameloblastoma(8). Each of them is manifested through their own biological behavior, and must be analyzed separately since they interfere directly with the different considerations regarding therapy and prognosis.

This specific case reports the occurrence of a unicystic ameloblastoma, which differs from multicystic ameloblastoma in that it is less aggressive and presents better clinical behavior (12, 13). In general, out of the ameloblastoma cases, 5 to 15% are unicystic, occurring mainly between the first and third decade of life, affecting the posterior region of the mandible in ninety percent of cases (10). This is in line with the information from the case reported in this study, since the lesion affects the 26-year-old patient and is present in the region of the body and branch of the mandible.

Moreira et al. reported that the most frequent clinical signs related to unicystic ameloblastoma are the swelling of the lesion site and/or the absence of a tooth in the tumor region. They are usually painless and the symptoms are minimal, making it difficult for the patient to notice in the early stages(10). The patient described here was referred by a dentist in his city, due to the discovery of a lesion in a radiographic examination performed for conventional dental treatment.

Radiographically, it appears as a unilocular radiolucent lesion, which in most cases surrounds the crown of a tooth, including asymptomatic lower third molars, presents a clinical and radiographic aspect similar to a cystic lesion, commonly confused with dentigerous cysts (9, 10, 12). The ameloblastoma of the present case shows an extensive unilocular radiolucent image, very well detailed in the axial, coronal and sagittal sections of the computed tomography, involving part of the body and the right mandibular branch extending up to the region of the mandible notch. It is still possible to note the involvement of tooth 32, which is included and mesioangulated, in addition to evident resorption of the distal root of tooth 31, favoring the diagnostic hypothesis of ameloblastic lesion over cystic lesion.

Unicystic ameloblastomas have three histopathological characteristics: luminal, intraluminal and mural(9, 10). Among the histological subtypes, the intraluminal or luminal types can be treated efficiently using conservative surgical techniques. However, the mural subtype, in which neoplastic cells cross the epithelial barrier and are found in the fibrous capsule, presents the possibility of invasion of the adjacent tissues, making the treatment, inevitably, more radical(9, 14). Ackermann and Shear report that the mural subtype shows more aggressive behavior and should be treated in a more comprehensive manner(15). Histopathology in the present case was characterized as a unicystic ameloblastoma of the mural type, which, although it has a better prognosis than the conventional variant, it shows greater capsular

invasiveness, justifying appropriate treatment and preservation.

Treatment for ameloblastomas, in most cases, occurs via radical surgical excisions with a safe margin of healthy bone(9, 16). On the other hand, the literature demonstrates that unicystic ameloblastomas, when receiving conservative treatment, present significant chances of success, despite the likelihood of recurrence of these tumors(12, 17, 18).

More conservative approaches are suggested as treatment for these lesions, such as curettage(12, 19), enucleation and decompression(20), which consists of using installed devices, adjacent to the lesion to make intralesional irrigations, preventing the lesion from growing and encouraging its reduction due to bone neoformation(21). According to this more conservative conduct proposal, the patient in the reported case was initially subjected to puncture, which was positive for fluid, whereafter marsupialization and concomitant incisional biopsy were conducted, ending with the complete enucleation of the lesion and vigorous ostectomy, since it is the mural variant of the lesion. It is worth noting that, in the literature, this variant has the surgical approach advocated by performing enucleation with vigorous peripheral ostectomy or even surgical resection with a safety margin. In order to achieve more tissue preservation and, taking into account the patient's more collaborative character, we opted for the more conservative approach.

Nakamura et al. stresses that the treatment protocol is mainly characterized by marsupialization of cystic tumors to reduce tumor volume and minimize the extent of surgery and treatment planning considering the tumor's growth characteristics. This approach saves patients from extensive resection of the mandible(21). In this case, the option for enucleation was made after approximately six months of marsupialization, since, radiographically, this was the time necessary for there to be considerable peripheral bone neoformation and consequent reduction of the tumor, to the point that the patient was able to undergo the definitive surgical procedure.

Lastly, it is worth noting that regardless of the technique used, clinical and radiographic monitoring of these lesions is essential for at least ten years and, preferably, fifteen years(19). Thus, the patient's preservation should extend, if possible, for at least ten years at the institution.

CONCLUSION

The present report showed a case of unicystic ameloblastoma, highlighting the importance of clinical and pathological correlation for the accurate diagnosis and indication of conservative and efficient treatment, in addition to the definition of the histological type to achieve careful preservation given its higher chances of recurrence.

The authors declare no conflict of interest.

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