Revista Brasileira de Odontologia
Brazilian Journal of Dentistry
Associação Brasileira de Odontologia - Seção Rio de Janeiro
DOI: http://dx.doi.org/10.18363/rbo.v77.2020.e1891

Conservative Approach of an Unicystic Ameloblastoma on a Young Patient - Case Report

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• Conflicts of interest: none declared.

Abstract

Objective: the aim of this study was to report the success of the conservative treatment on an unicystic ameloblastoma on a young patient. **Case report:** a 12-year young male, presented for consultation at the Stomatology clinic with the chief complaint of facial asymmetry and limited mouth opening. At the extraoral clinical examination, it could be observed the presence of facial swelling on the left side. At intraoral examination, swelling and erased vestibule in the left posterior region of the mandible was observed. The panoramic radiography showed an unilocular, radiolucent osteolytic lesion that involved the mandible body, angle and ramus and caused the upper displacement of tooth 38. Cone-beam computed tomography showed expansion of the buccal and lingual bone. In view of these findings, the diagnostic hypotheses were: Dentigerous Cyst; Unicystic Ameloblastoma and Ameloblastic Fibroma. An incisional biopsy was performed, which established the definitive diagnosis of unicystic ameloblastoma. The patient underwent marsupialization and seven months later, enucleation. After a 7-year clinical follow-up, the patient remains free of the disease. **Conclusion:** conservative surgical treatment of unicystic ameloblastoma may be indicated in young patients, in order to avoid a mutilating procedure.

Keywords: Odontogenic Tumors; Ameloblastoma; Conservative Treatment.

Introduction

Unicystic Ameloblastoma (UA) is a distinguishable entity of Ameloblastomas, accounting for a third or less of all cases. This variant is clinically distinct - as it shows a slight preference for patients around age 13 (range 5-16 years) - and prognostically, as less recurrence rates are observed within this tumor when treated conservatively.[1,2,3,4,5] Therefore, a therapeutic dilemma remains on whether UA should be treated as the conventional Ameloblastoma or in a less-invasive way, once that most patients are at a younger age and radical surgery could result in teeth loss, bone growth alterations and significant emotional alterations.

Therefore, we report a case of UA affecting a 12-year-old young male patient, treated conservatively and a 7-year follow-up.

Case Report

A 12-year-old young patient was referred from an orthodontist that had already started the orthodontic treatment for evaluation of a 2-month lasting facial swelling on the left side, restricted mouth opening and a radiolucent lesion located on the left mandible. Medical history was non-contributory. Extraoral clinical examination showed a left-sided unilateral large swelling - referring tenderness on palpation - on the left lower and middle third region of the face, extending from the corner of the mouth to the angle molar ramus region of the mandible and continuing to the coronoid process resulting in facial asymmetry (Figure 1A, 1B, 1C). Intraoral clinical examination revealed the presence of mandibular swelling covered by normal oral mucosa and erasure of the vestibule in the left posterior region and also, mobility and displacement of the first and second left mandibular molars (Figure 1D).

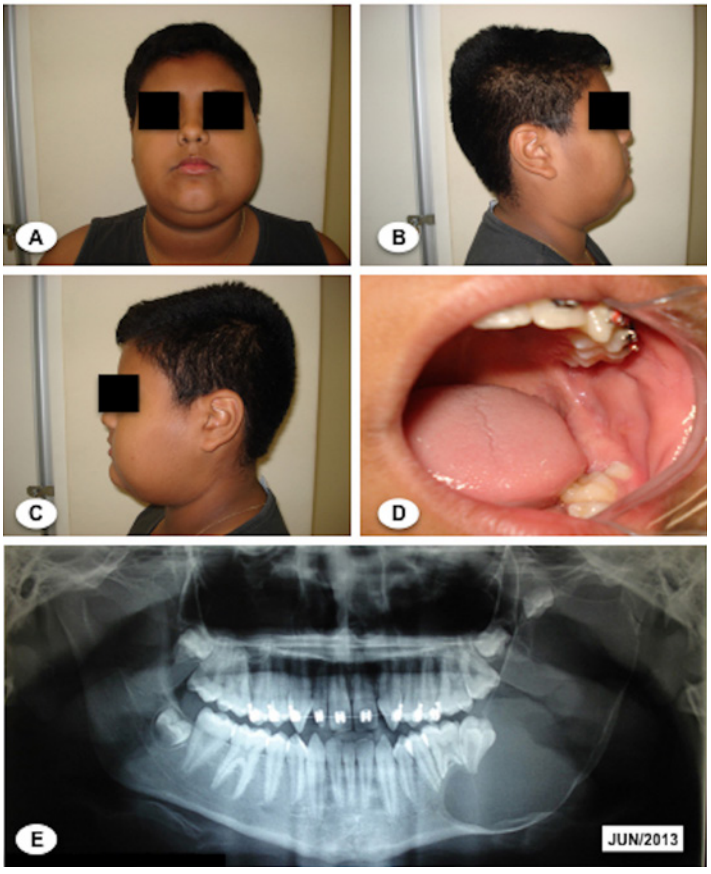

Figure 1. Initial clinical and radiographic findings. A. Frontal view - A large left-sided facial asymmetry may be observed. B. Right profile view showing no alterations. C. Left profile view - facial swelling extending from the corner of the mouth to the angle molar ramus region of the mandible and continuing to the coronoid process resulting in facial asymmetry. D. Intraoral aspect showing a left posterior mandibular swelling covered by normal mucosa; teeth #36 and #37 displacement. E. A large unilocular radiolucent lesion in association with tooth #38, causing root resorption of the first and second left mandibular molars could be observed on the initial panoramic radiography.

On the initial panoramic radiography, a large, well-delimited and expansive, unilocular lesion associated with impacted tooth #38 - measuring 8.5 cm in its largest diameter - could be observed extending from the first left mandibular teeth to the coronoid process, resulting on root resorption of teeth #36 and #37 (Figure 1E). A cone-beam computed tomographic exam was requested and the expansion of buccal and lingual corticals of the body, angle and mandibular ramus could be observed (Figure 2). Differential clinical diagnosis was dentigerous cyst, unicystic ameloblastoma and ameloblastic-fibroma. The cavity was punctured prior to the incisional biopsy and marsupialization - both performed under local anesthesia (Figure 3A).

The specimen was immersed in 10% formaldehyde and sent for histological analysis. Hematoxylin and eosin stained histological sections showed a cystic lining with ameloblast-like basilar cells with no evidence of stromal invasion. (Figure 3B). Due to the clinical, radiological, and histological findings the UA diagnosis was established. Patient was submitted to the oral surgeon to enucleate the lesion after a 7-month marsupialization period, when the lesion presented smaller than initially (Figure 3C). Patient recovery was uneventful (Figure 4A) and a 2-year radiographic follow-up showed osseous neoformation and repair in the area (Figure 4B). At a 3-year follow-up, there were no radiographic signs of the disease (Figure 4C). The patient remains in clinical follow-up and there are no signs or symptons of local recurrence after seven-years.

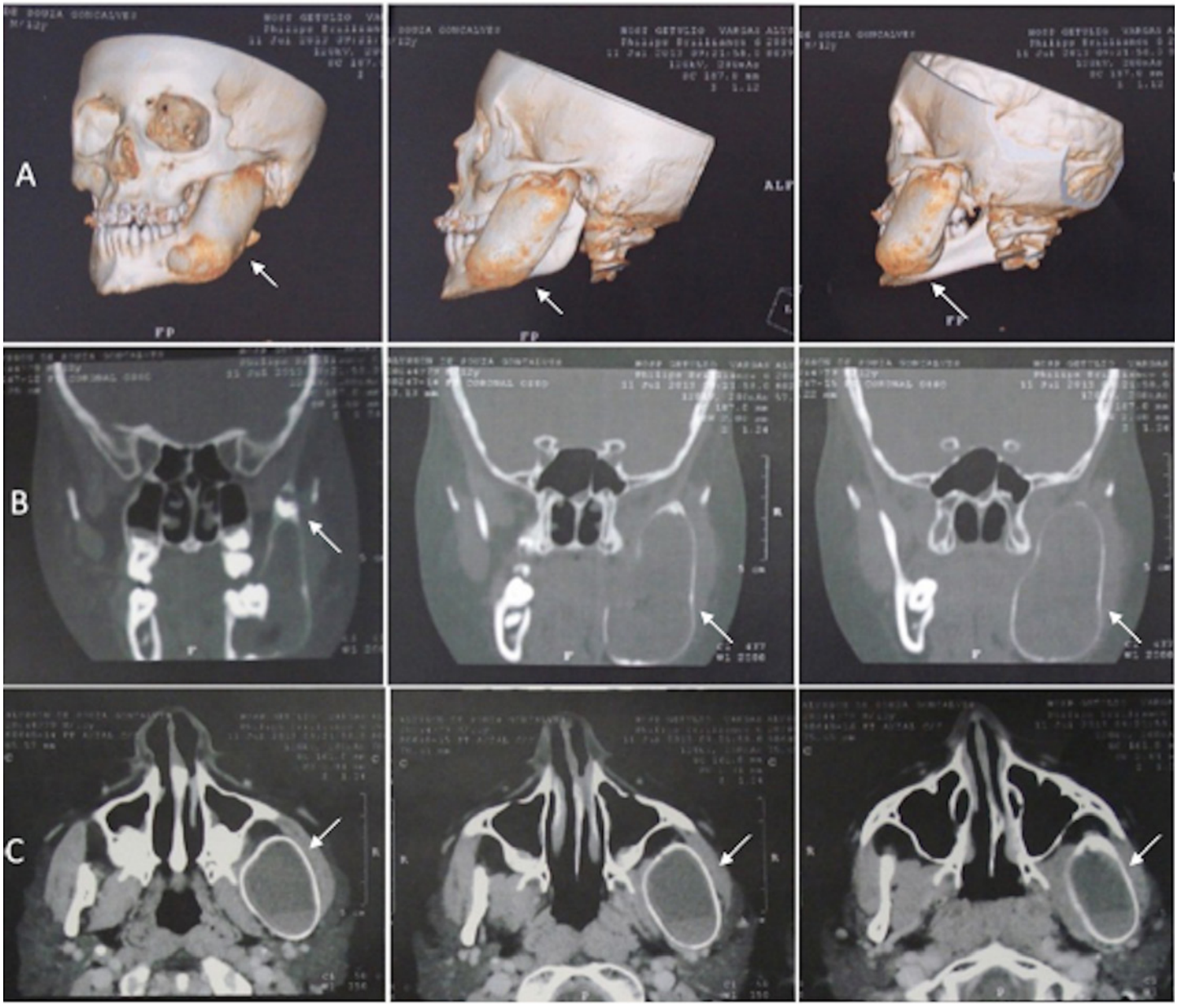

Figure 2. Cone-beam computed tomographic exam. A. 3D Reconstruction; arrow pointing out to the lesion. B. Coronal section; arrow pointing out to a well-circumscribed hypodense lesion with no cortical rupture and tooth #38 displaced all the way to the coronoid process. C. Axial section; arrow pointing out to the lesion, suggestive of a cystic lesion with no cortical rupture.

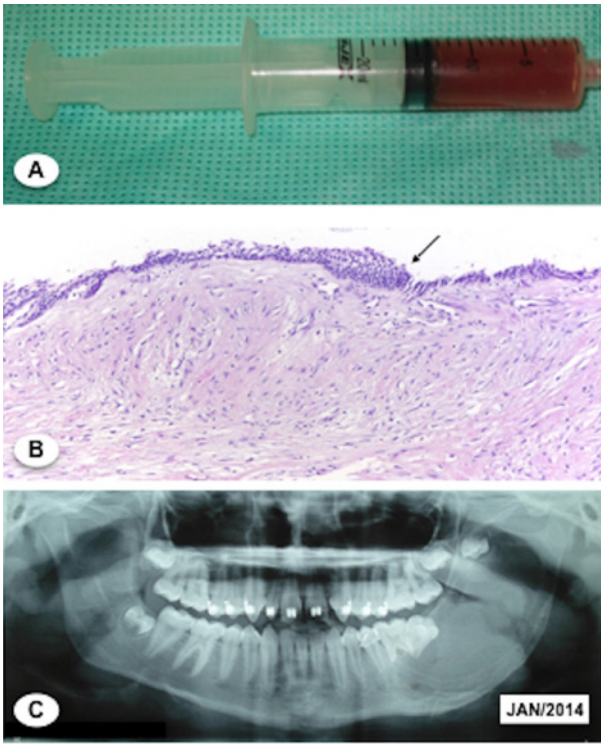

Figure 3. Biopsy and marsupialization findings. A. The puncture exam collected blood from the cavity. B. Histological features of the lesion showing a cystic lining with ameloblast-like basilar cells with no stromal invasion (Hematoxylin and eosin). C. Panoramic radiography after a 7-month marsupialization period prior to enucleation, showed a decrease in the lesion size.

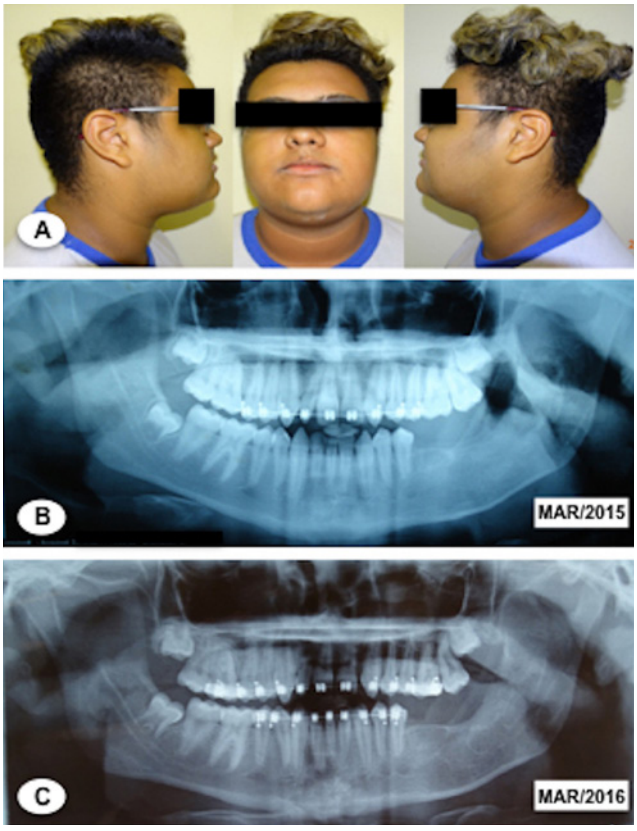

Figure 4.Clinical and radiographic follow-up. A. Extraoral clinical aspect showing no asymmetry. B. 2-year radiographic follow-up showing osseous neoformation and repair. C. A 3-year radiographic follow-up showed no signs of the disease.

Discussion

UA is a variant of the Ameloblastoma that shows some distinct characteristics such as its radiographic and histological patterns, lower rates of local recurrence and higher success when treated conservatively once compared with the conventional ameloblastoma[1-3]. This condition has equal predilection for both male and female and most patients are diagnosed in the second decade of life (mean age of 13 years; range 5-16 years). The most common anatomical location for UA is the posterior mandible and it presents as a well-defined solitary unilocular radiolucency that can cause tooth root resorption, teeth displacement and cortical destruction.[4,5,6] The present case, an UA affecting a 12-year old male patient located in the left posterior mandible that caused the first left lower molar and the second lower left molar roots resorption and the lower left third molar displacement is in accordance with most findings reported in literature.

Ameloblastoma and its variants can only be diagnosed after histopathological examination. The incisional biopsy remains the gold standard for its diagnosis. This diagnostic method is important once dealing with a radiolucent lesion on posterior mandible region - especially when associated with an impacted tooth as in the present case - to excluded others odontogenic tumor/cyst such as the dentigerous cyst, odontogenic keratocyst and even conventional ameloblastoma.[7,8]

Unicystic ameloblastoma can be classified into three different histological types (Figure 5) - the luminal subtype in which the ameloblast-like cells, stellate-/reticulum-like cells, and vacuolated cells are confined to the epithelium line; Intraluminal subtype is characterized by the proliferation of tumoral nodules into the cyst lumen and finally, the mural subtype in which the ameloblastoma epithelium invades the connective tissue wall.[9,10] Differently from the conventional ameloblastoma - which its histological subtypes do not change its treatment - the UA histopathological classification has an important role on its therapeutic decision, recurrence rate and prognosis whereas the luminal and intraluminal subtype show good prognosis and rarely recur and the mural shows characteristics similar to the conventional ameloblastoma.[9] In our case, as histopathological exam did not show mural invasion, although recurrence was possible, we opted to treat conservatively, because of the morbidity that radical surgery could cause on a young male.

Despite the differences between therapeutic modalities - conservative vs radical - of each UA subtypes, its incidence, prognosis and recurrence rate may vary from each series of reported cases. Siriwarden *et al.*[11] reported that 63% of the UA cases presented as the luminal ou intraluminal subtype and recurrence was only seen in 3.5% for all of the cases whereas *P*hilipsen and Reichart[5] reported a recurrence rate of 10.7% for the luminal and intraluminal subtypes. A systematic review

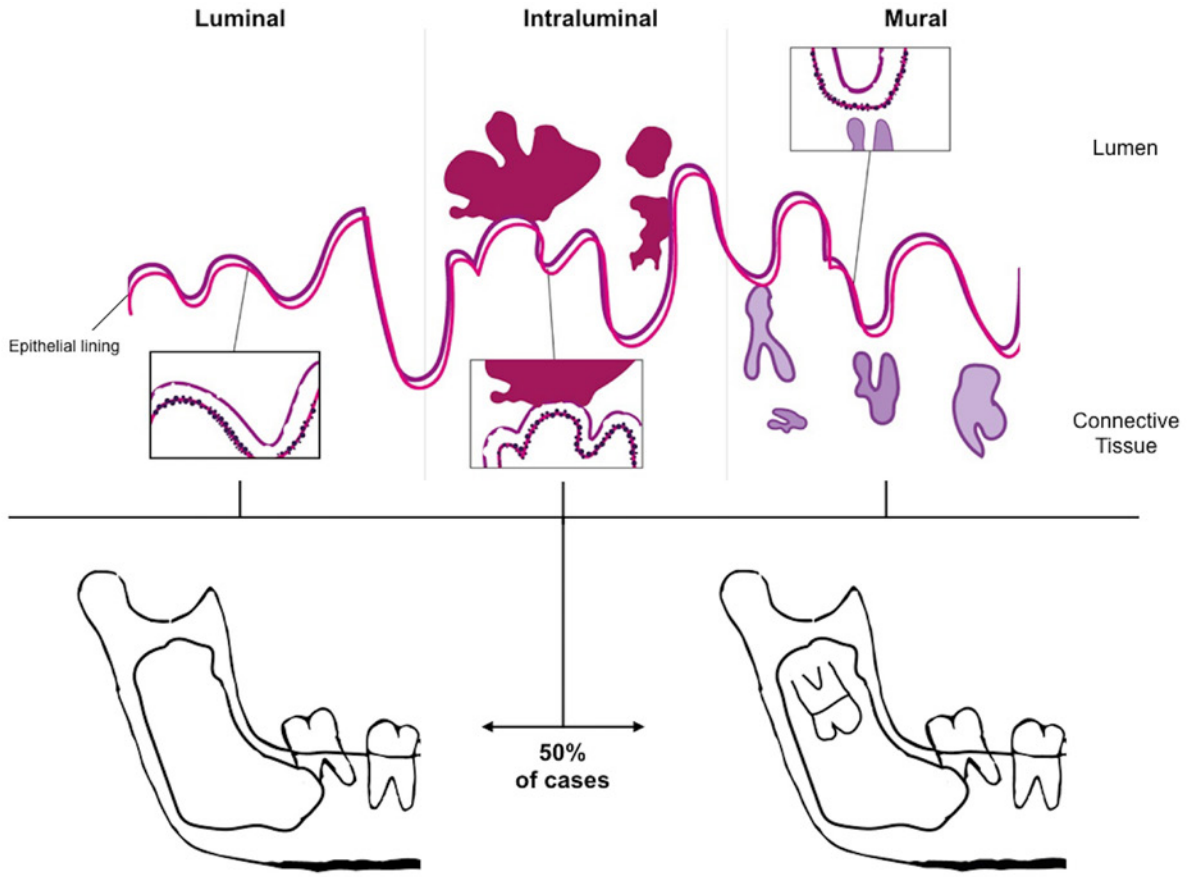

Figure 5. Unicystic Ameloblastoma histological subtypes: The luminal subtype in which the ameloblast-like cells, stellate-/reticulum-like cells, and vacuolated cells are confined to the epithelium line; Intraluminal subtype is characterized by the proliferation of tumoral nodules into the cyst lumen and the mural subtype in which the ameloblastoma epithelium invades the connective tissue wall. Despite the histological differences, UA radiographic aspect is the same for all variants - it presents as an unilocular radiolucent lesion that can cause tooth displacement and root resorption, furthermore, in 50% of the cases the lesion is in association with an impacted tooth.

by Lau and Samman[3] showed that radical surgery resulted in the lowest recurrence rate (3.6%) between all treatment options. On a systematic review and meta-analysis reported by Hendra *et al.*[12] the recurrence rate were of 3% and 21% after radical and conservative treatment, respectively. It is important to emphasize that most of UA affects pediatric to young patients and that radical surgery can cause several morbidities other than functional, like sociological and psychological. Furthermore, treating younger patients is always a challenge due to their morphology and physiology - continuing facial growth, more trabecular bone, increased bone turnover and the presence of unerupted teeth bud.[13,14] Therefore, conservative approaches when treating UA affecting the youngest should be considered at first.

In the present case, although the possibility of recurrence, we opted to conduct a conservative treatment in order to avoid several sequels on a young male. This choice was made based on the fact that we were dealing with an UA and not the conventional one - it was the luminal subtype which is described with lower recurrence rates and better prognosis when treated conservatively and as there were no symptoms or risk to neuro-vascular structures. In this case, the use of marsupialization prior to the enucleation was essential to its success.

Marsupialization is a procedure for the treatment of large cystic lesions and can be used to relieve symptoms and promote a more conservative enucleation afterwards by decompressing it.[15,16] Furthermore, it lowers the intracystic hydrostatic pressure mechanism which results on the reduce of invasiveness and bone resorption (elevated hydrostatic pressure can upregulate MMP-2 and MMP-9 expression via Wnt/β-catenin pathway).[17] Recurrence rate and the unpredictability of this technique stand for its contraindications. Despite its promising results, some studies suggest that marsupialization may not be indicated as an initial treatment when, specifically, three factors are involved - root resorption, perforation of the cortical bone or when dealing with the mural histopathological subtype.[18] In our case, even though one of these factors were present, we understood that this practice could generate more benefits to the patient. Therefore, as the lesion decreased size, bone remodeling and osteogenesis were observed and permitted an easier enucleation 7-months after the procedure.

Conclusion

Unicystic Ameloblastoma (UA) is a distinguishable entity of ameloblastoma that shows preference for younger patients, better prognosis and lower recurrence rate when treated conservatively. Therefore, the conservative management should be considered first once dealing with this unusual entity in order to cause less-morbidity.

Acknowledgment

We would like to thank Professor Fábio Ramôa Pires - Oral Pathology Laboratory of State University of Rio de Janeiro - for the histopathological examination performed for this case.

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Submitted: 03/08/2020 / Accepted for publication: 10/08/2020

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