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Large calcifying odontogenic cyst in the posterior maxilla

Extenso cisto odontogênico calcificante em maxila posterior

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ABSTRACT

The occurrence of calcifying odontogenic cyst (COC) in the posterior maxillary region is uncommon, with few cases described in literature. We report a case of a 13-year-old female patient presenting a large lesion in the left maxilla (> 7.5 cm). Panoramic radiograph showed a well-delimited radiolucent unilocular lesion extending from the posterior left maxilla to the maxillary sinus. The patient was submitted to decompression followed by conservative surgical removal of the lesion. Histological analysis of the surgical specimen confirmed the diagnosis of COC. After one year, no local recurrence has been observed and the patient remains under follow-up on a regular basis.

Key words: calcifying odontogenic cyst; decompression; maxilla; maxillary sinus; conservative treatment.

RESUMO

É incomum a ocorrência de cisto odontogênico calcificante (COC) em região posterior de maxila; na literatura, há poucos relatos descritos. Reportamos o caso de uma paciente, 13 anos, que apresentou uma extensa lesão na maxila esquerda (> 7,5 cm). A radiografia panorâmica exibiu lesão radiolúcida unilocular bem delimitada, extendendo-se da maxila posterior esquerda ao seio maxilar. A paciente foi submetida à descompressão, seguida de remoção cirúrgica conservadora da lesão. A análise histológica do espécime cirúrgico confirmou o diagnóstico de COC. Após um ano, nenhuma recorrência foi observada. A paciente permanece em acompanhamento regularmente.

Unitermos: *cisto odontogênico calcificante; descompressão; maxila; seio maxilar; tratamento conservador.*

RESUMEN

La aparición de un quiste odontogénico calcificante (QOC) en la región posterior de la maxila es infrecuente; hay pocos informes descritos en la literatura. Presentamos el caso de una paciente de 13 años que presentó una lesión extensa en la maxila izquierda (> 7,5 cm). La radiografía panorámica mostró una lesión radiolúcida unilocular bien delimitada, que se extendía desde el maxilar posterior izquierdo hasta el seno maxilar. La paciente fue sometida a descompresión, seguida de de la extirpación quirúrgica conservadora de la lesión. El análisis histológico de la pieza quirúrgica confirmó el diagnóstico de QOC. Después de un año, no se observaron recurrencias. La paciente permanece en seguimiento regular.

Palabras clave: *quite odontogénico calcificante; descompresión; maxila; seno maxilar; tratamiento conservador.*

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INTRODUCTION

The calcifying odontogenic cyst (COC) is an uncommon developmental cyst that accounts for less than 1% of all odontogenic cysts and tumors[1]. The World Health Organization (WHO) defines COC as a cyst lined by ameloblastoma-like epithelium containing focal accumulations of ghost cells, dentinoid material and calcifications[2]. In addition to COC, which is considered a non-neoplastic cystic condition, dentinogenic ghost cell tumor (a benign neoplasm) (DGCT) and ghost cell odontogenic carcinoma (a malignant neoplasm) (GCOC) complete the typical triad of ghost cell-containing odontogenic conditions[3, 4]. It is accepted that COC arises from remnants of the dental lamina[2] and several studies have studied the immunoexpression of cytokeratins and the presence of Wnt/β-catenin signaling pathway proteins in COC, suggesting their importance in the development and progression of these lesions[5, 6].

COC may appear clinically as an intraosseous or an extraosseous/peripheral lesion[2]. The most common presentation is a slow-growing swelling in the anterior region of the maxillary bones and jaw, affecting young adults in the third or fourth decades of life[2, 4]. The COC presents as well-defined unilocular or multilocular radiolucencies associated with the variable presence of radiopaque foci[7-9]. They can be associated with unerupted teeth, mostly the upper canines, and can produce root resorption in the adjacent teeth[10, 11]. Most COCs measure from 2 to 4 cm at the time of diagnosis, but some cases can reach larger proportions[12].

COC can be diagnosed in association with other odontogenic cysts and tumors, mostly odontomas[8]. Despite its growth potential, COC treatment is based on a conservative surgical approach, and marsupialization/decompression is sometimes indicated for extensive lesions[1]. Recurrence after conservative management of COC is low[1]. The aim of this paper is to report the case of a large maxillary COC with emphasis on its clinical, pathological and treatment features.

CASE REPORT

A 13-year-old girl was referred for evaluation of a swelling in the left maxilla associated with teeth mobility in the area lasting six months. An extraoral examination revealed a swelling in the infraorbital, paranasal, and left zygomatic regions. Intraoral examination revealed a swelling in the upper left vestibule extending from the first premolar to the third molar associated with mobility on teeth 25, 26 and 27. Medical history revealed no abnormalities.

Panoramic radiograph showed a well-defined unilocular radiolucent area in the left maxilla extending from the right central incisor to the left third molar (**Figure 1A**). Root resorption in the adjacent teeth was observed, as well as several radiopaque foci inside of the lesion. Computed tomography (CT) scans showed swelling and perforation of both buccal and palatal cortical bones (**Figure 1B** and **C**). The lesion occupied almost the entire left maxillary sinus and it was in close proximity to the orbit floor, measuring 7.5 cm in its largest diameter. Clinical diagnosis included ameloblastoma, odontogenic keratocyst, and COC. An incisional biopsy was performed. As the intraoperative features were compatible with a cystic lesion, a decompression with a tube was performed (**Figure 2A**). Histological analysis of the hematoxylin and eosin (HE)-stained slides revealed a cystic cavity lined by an epithelium composed of a basal layer of polarized columnar cells and a superficial area resembling the stellate reticulum. In addition, ghost cells and calcifications were present in the epithelium, rendering the diagnosis of COC.

The patient underwent monthly routine clinical follow-up and was submitted to a conservative surgical removal of the lesion under general anesthesia after six months (**Figure 2B** and **C**). The procedure included a vertical para-papillary incision

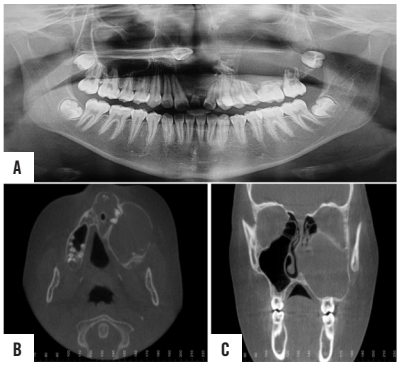

FIGURE 1 – *A) panoramic radiograph showing an extensive radiolucency extending from the upper right central incisor to the upper left third molar. Note the presence of radiopaque foci and root resorption of the teeth closely associated with the lesion; B) axial and C) CBCT images showing the large dimensions of the lesion*

CBCT: coronal cone-beam computed tomography.

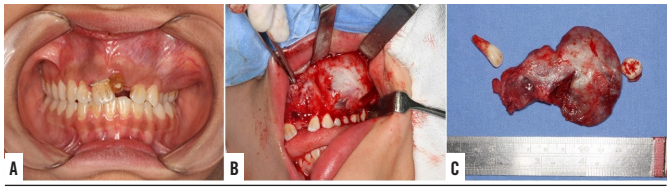

FIGURE 2 – *A) initial surgical approach for cyst decompression. Note the vestibular expansion associated with the lesion; B) conservative surgical removal of the lesion showing buccal expansion; C) surgical specimen associated with the upper right central incisor and third mola*

between teeth 11 and 22 and a sulcular incision of tooth 11 to 27 to access the lesion. After the excision was completed, curettage and peripheral osteotomy were performed in the entire bone cavity to remove any epithelial remnants and to reduce the risk of recurrence. The surgical specimen confirmed the presence of a cystic cavity and histological analysis confirmed the diagnosis of COC (**Figure 3A** and **B**). Panoramic view and CT scans revealed bone formation throughout the area (**Figure 4A** and **B**). The patient remains in clinical follow-up for one year with no signs of recurrence (**Figure 4C**).

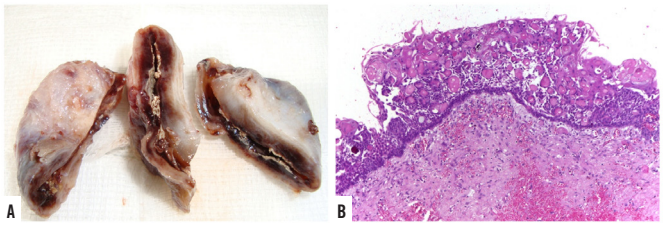

FIGURE 3 – *A) detail of the surgical specimen showing the cyst cavity; B) HE stained histological section showing the cyst lining composed by basal cells, areas resembling stellate reticulum, ghost cells and calcifications (HE, 100× magnification)*

HE: hematoxylin and eosin.

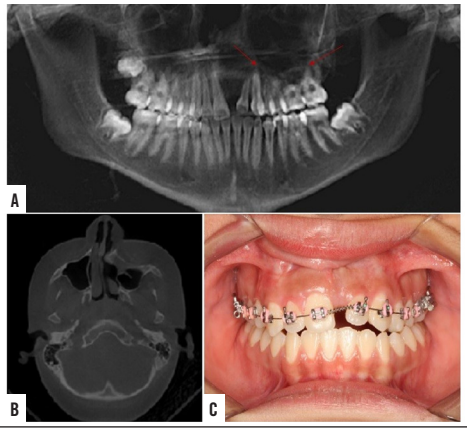

FIGURE 4 – *A) panoramic view; B) axial CBCT images taken one year after surgery showing evidence of bone formation in the region (red arrows); C) intraoral appearance one year after surgery*

CBCT: coronal cone-beam computed tomography.

DISCUSSION

COCs are uncommon, distinctive, and characterized as benign cystic odontogenic lesions with a predilection for the anterior region of the jaws. It may involve bone or extraosseous/peripheral tissues[2]. COC was first described by Gorlin in 1962 as a cystic lesion, but it shares many clinical, radiological and histological characteristics with its solid counterpart, the DGCT[3, 13, 14]. Extensive maxillary COC (> 4 cm) in the posterior region are rare, with only a few cases reported in the literature (**Table**).

In a study of 215 cases of COC by Buchner (1991)[12], the size of the lesion was known in 58 cases, ranging from 0.5 to 12 cm. Of these, almost 60% were sized between 2 and 3.9 cm, and the average recorded size was 3.3 cm. The average age at the time of presentation of extensive COC cases collected from the literature (Table), was 31 ± 17.2 years (ranging 13 to 62 years), without any gender predilection. The occurrence of these lesions in the posterior maxillary region is generally low, representing from 2% to 11.7% of all cases[12]. Extensive COC can be associated with increased volume, malocclusion, painful sensation, aesthetic deformity, and risk of compromising adjacent anatomical structures[15-17]. Likewise, in a multicenter study of 268 cases of COC, Arruda *et al*. (2018)[1] found that the size of the lesion was larger among symptomatic lesions. In these cases, the most common radiographic findings are bone expansion, that may or may not be accompanied by cortical erosion, tooth displacement, root resorption, and occasionally, the involvement of the maxillary sinus[7, 10, 17, 18]. Similarly, the lesion described in our report extended throughout the maxillary sinus and occupied part of the floor of the left orbit, causing increased volume in the maxilla.

The distinctive histology of COC is characterized by an ameloblastoma-like epithelium lining a cystic cavity, associated

TABLE – Summary of extensive (larger than 4 cm) calcifying odontogenic cysts in the posterior maxillary region (premolar to molar) reported in the English language literature

Reference	Sex	Age (years)	Side	Size (cm)*	Association	Treatment
Tanimoto *et al*. (1988)	M	17	R	> 4**	Odontoma	Enucleation
Tanimoto *et al*. (1988)	F	14	R	> 4**	Odontoma	Enucleation
Rushton *et al*. (1997)	M	51	L	> 4**	NA	NA
Yoshiura *et al*. (1998)	M	62	L	> 4**	NA	NA
Fregnani *et al*. (2003)	F	43	R	5	NA	Enucleation
Utumi *et al*. (2012)	F	36	L	> 4**	Odontoma	Enucleation
Kim *et al*. (2016)	M	24	L	> 4**	NA	Marsupialization and enucleation
Emam *et al*. (2017)	M	36	R	5	NA	Decompression, enucleation and curettage
Arboleda *et al*. (2018)	F	15	R	> 4**	Dentigerous cyst	Enucleation
Present case	F	13	L	7,5	Odontoma	Decompression, curettage and peripheral ostectomy

*M: male; F: female; R: right; L: left; NA: not available; *largest diameter; > 4**: cut-point estimated of radiographic images in cases without recorded dimensions.*

with the presence of ghost cells and calcifications, and dentinoid formation[2, 14]. COC has been previously classified into different histological variants (simple cyst, proliferative cyst, ameloblastomatous type, and associated with odontoma or other benign odontogenic tumors other than odontoma) but this classification does not seem to have clinical and behavioral significance. Therefore it has not been adopted by the WHO classification[2, 14, 17]. When evaluating the histological variant of 52 cases of COC, Irani *et al*. (2017)[14] observed that the simple cystic and odontoma-associated COC rate was 30.8% for both subtypes, being the most commonly found histological types. As described in Table, odontoma is the most common COC-associated odontogenic tumor in larger lesions.

Enucleation is the treatment of choice for intraosseous COC and recurrences are observed in less than 5% of the cases[1, 12]. This therapeutic modality is seen as an effective technique even for larger lesions, as it avoids unnecessary mutilation and tooth loss[16, 19]. As seen in Table, enucleation was performed in 87.5% (7/8) of the extensive COC included in our review. Arruda *et al*. (2018)[1] found that only 4% of COC reported in the literature were managed through a two-step approach, including decompression or marsupialization followed by enucleation. Particularly, we have shown a conservative two-step approach to COC with good clinical results in the present report. Apart from that, COC rarely undergoes malignant transformation[20], reinforcing the utility of this conservative surgical approach even for large posterior maxillary COC.

In summary, we report a case of an extensive posterior maxillary COC treated conservatively with the improvement of the patient's overall condition. The present case reinforces that decompression should be considered as an option in large lesions in young patients, reducing the morbidity and thus helping the preservation of important anatomical structures.

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