

## Calcifying cystic odontogenic tumor of maxilla: A case report

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### Abstract:

The calcifying cystic odontogenic tumor (CCOT) is an uncommon pathology, representing about 2% of all odontogenic cysts and tumors. Generally, is presented as slow growth, asymptomatic, without predilection of gender or location. The most common radiographic presentation is radiolucent image with radiopaque areas, especially in the lesion periphery, unilocular, expansive and well- defined margin. The purpose of this report is to describe the case of a 16-year-old boy with a painless and progressive increase of volume on the face, which started 7 months ago. In oroscopy, we observed an expansive increase of volume in right maxilla, extending from premolars to molars on the same side. In computed tomography, it was possible to observe an unilocular, expansive image in right maxilla. After incisional biopsy, the histopathological analysis evidenced epithelial neoplasia of odontogenic origin that presents cystic portions and solid portions. In the cystic portions, there is an epithelial lining which basal cells are cuboidal or cylindrical arrangement in palisade, presence of ghost cells and dystrophic mineralization, confirming the diagnosis of calcifying cystic odontogenic tumor. Excision and curettage were performed under general anesthesia. The excised tumor was analyzed, confirming the diagnosis of CCOT and the patient has been in clinical and radiographic follow- up for a period of 5 years, without signs of relapses.

**Keywords:** Odontogenic Tumors; Maxilla; Pathology, Oral.

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## INTRODUCTION

In 1962, Gorlin et al.<sup>1</sup> described a new cystic pathology, with varied clinical and histological presentation, derived from odontogenic epithelial remains. Posteriorly, in 2005, the World Health Organization classified it as calcifying cystic odontogenic tumor (CCOT) due to its neoplastic behavior<sup>2</sup>.

Although odontogenic tumors are part of the day-to-day services of oral diagnosis, corresponding to about 3.5% of all final diagnoses<sup>3</sup>, CCOT is an uncommon pathology, accounting for about 2% of all odontogenic cysts and tumors. Usually has intraosseous origin, however, there is the extraosseous variation that is less common<sup>4</sup>. Occurs in maxilla with the same frequency of the jaw, with no predilection for gender, however, there is a higher incidence in young adults between the 3<sup>rd</sup> and 4<sup>th</sup> decades of life<sup>5-7</sup>.

The most common radiographic presentation is a radiolucent image with radiopaque areas, especially in the periphery of the lesion, unilocular, expansive and well-defined margins. Root resorption and teeth divergence are observed frequently<sup>8</sup>.

Histologically, the CCOT presents like a cystic cavity with covered with non-keratinized stratified squamous epithelium of varying thickness, composed of basal layer of columnar arrangement in palisade, similar to ameloblasts. It is possible to observe layers of eosinophilic cells, also called “ghost cells”, besides acellular calcified matrix dentinoid or bone<sup>4,9</sup>.

The treatment consists of the surgical removal and curettage of the bone walls. Due to the potential for relapse, follow-up should be careful<sup>5</sup>. The malignant transformation of CCOT is controversial. In literature, it is possible to observe reports of development of odontogenic carcinoma of ghost cells. Therefore, although rare, the transformation may occur. Therefore, the postoperative follow-up of these patients is required<sup>10</sup>.

## CASE REPORT

A 16 year- old boy, sought treatment with complaint of an increase of volume, painless and progressive on the face in the last 7 months. At the extraoral physical examination is possible to observe facial asymmetry, with slight nasolabial groove erasure, on the right side (Figure 1). In oroscopy, an increase of volume in right maxilla was noted (Figure 2).



**Figure 1.** Extraoral aspect with slight nasolabial groove erasure, on the right side.

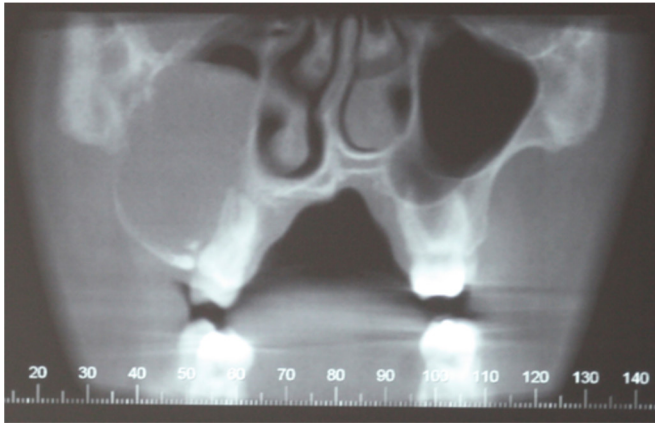


**Figure 2.** Intraoral aspect evidencing increase of volume by vestibular in maxilla, in the region of right superior premolars and molars.

Computed tomography was requested, which shown a homogeneous expansive hypodense lesion in right maxilla (Figures 3 and 4). With diagnostic hypotheses of keratocystic odontogenic tumor and calcifying epithelial cyst, an incisional biopsy with anatomopathological result of an odontogenic cystic calcifying tumor was performed.

In view of this result, conservative surgery for tumor excision with curettage was proposed and performed (Figure 5). The excised tumor was analyzed histopathologically, an analysis that revealed epithelial neoplasia of odontogenic origin that presents cystic portions and solid portions. In cystic portions, there is an epithelial lining which basal cells are cuboidal or cylindrical arrangement in palisade and superficial polyhedral or fusiform cells arranged in a looser arrangement.

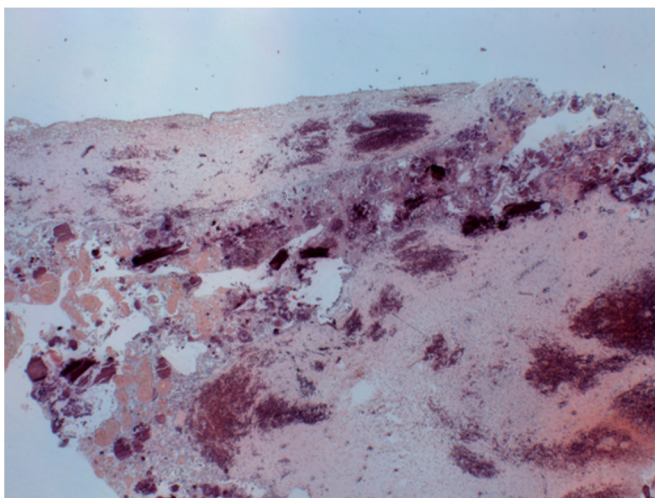
In this arrangement are seen several ghost cells which, when fused, form areas of eosinophilic



**Figure 3.** CT, coronal section, showing a hypodense lesion in right maxilla with expansion and loss of continuity of the vestibular cortex of maxillary sinus.



**Figure 4.** CT, sagittal section, where it is possible to observe the lesion extending posteriorly to the posterior wall of the maxillary sinus.



**Figure 5.** Intraoperative aspect of the pathological cavity after excision of the neoplasia.

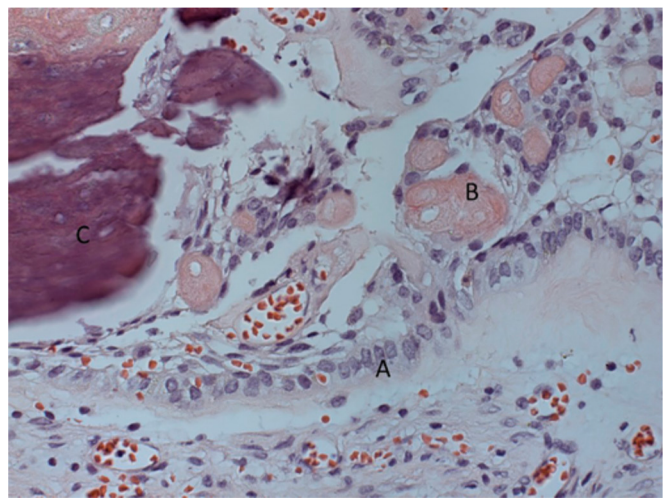
and acellular material. Several foci of dystrophic mineralization are also part of the lesion. Capsule composed of dense connective tissue with moderate chronic inflammatory infiltrate (Figures 6, 7 and 8). Foci of interstitial hemorrhage completed the analyzed picture, confirming the diagnosis of CCOT.

## DISCUSSION

The CCOT, although it can occur in any age, has a preference for the 2<sup>nd</sup> and 3<sup>rd</sup> decades of life, with no predilection of gender<sup>11</sup>. In this way, the case presented here is in agreement with most of the literature.

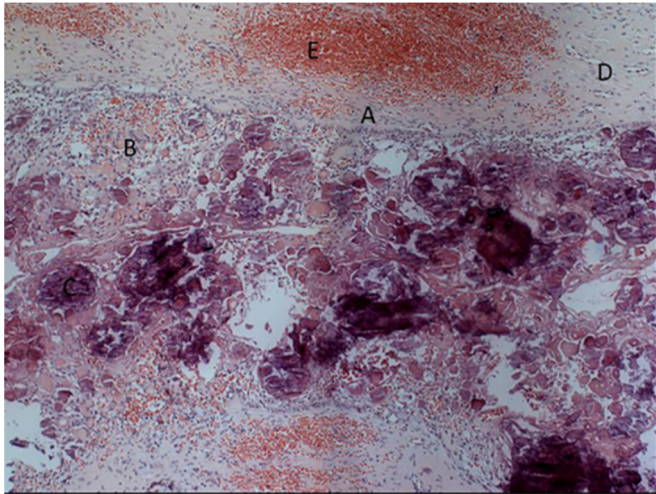


**Figure 6.** H&E 2.5X: Epithelial neoplasia of odontogenic origin that presents cystic portions and solid portions. Several foci of dystrophic mineralization are also part of the lesion. The capsule is made of dense connective tissue with moderate chronic inflammatory infiltrate. Foci of interstitial hemorrhage may also be observed.



**Figure 7.** H&E 40X: (A) Basal cells in palisade. (B) Ghost cells. (C) Foci of dystrophic calcification.





**Figure 8.** H&E 10X: (A) Basal cells in palisade. (B) Ghost cells with nuclei absence. (C) Foci of calcification. (D) Cystic capsule. (E) Foci of interstitial hemorrhage.

In general, this neoplasia is slow growing and asymptomatic<sup>4,7</sup>, and the jaw is as affected as the maxilla, what changes are the signs and the possible symptoms in each locality. Mandibular lesions usually do not result in any symptoms. However, maxillary lesions, when expansive towards the nasal fossa can lead to decreased air perfusion in the nostrils or even epistaxis<sup>5</sup>. In this report, despite the maxillary involvement, he did not present any nasal complaints, perhaps because the lesion does not move importantly the walls of the nasal cavity.

Our case presents with an unilocular lesion with well- defined limits. This is the most common radiographic manifestation in the cases reported<sup>5,6,8,11,12</sup>, however it is possible to occur with multiloculated image<sup>9</sup>.

Due to the varied clinical / radiographic picture, the CCOT has several differential diagnoses, varying according to the presence to a greater or less degree of radiopacities into the lesions. Predominantly radiolucent images should be confronted with a dentigerous cyst, keratocystic odontogenic tumor, adenomatoid odontogenic tumor, and others shown radiopacities images in the interior can be confused with calcifying epithelial odontogenic tumor, ossifying fibroma and even odontoma<sup>13</sup>.

The histopathological aspect of CCOT consists of a fibrous capsule with odontogenic epithelial lining. In addition, a microscopic characteristic of this pathology is the presence of cells with absent nuclei, called “ghost cells”. Various presence of dentinoid or bone matrix can also be visualized<sup>10,11</sup>. In our case, it was possible

to observe a neoplasia, sometimes solid or cystic, with palisade epithelial lining. The “ghost cells” could be seen permeating the entire lesion. In certain localities they were grouped forming large areas of eosinophilic and acellular material. Although the literature says that it is possible to observe mineralized material similar to dentin<sup>14</sup>, in our case, the mineral product was in form of a dystrophic mineralization.

About the treatment, the CCOT can resort to surgical enucleation, however, this does not occur in the vast majority of cases. According to Stoelting<sup>15</sup>, the surgical planning for aggressive cystic lesions should take into account some aspects: presentation (uni or multilocular), buccal and lingual expansion and location. Due to low recurrence reported in the literature, conservative surgical treatment has been proposed with safety<sup>5,6,11</sup>.

Despite this, it has been suggested that the frequency of relapses for this neoplasia should be reviewed, because the articles present sequences of small case reports and with small postoperative follow-up<sup>15</sup>. In addition, there is the rare but reported possibility of malignant transformation to odontogenic carcinoma of ghost cells. Therefore, the consensus of the literature requires the careful monitoring of these patients over a long period of time, to follow up any possible recurrence or transformation<sup>10</sup>. In the present case, the patient was evaluated through physical and imaging exams for 5 years. Where bone neoformation was observed in absence of any tumor recurrence.

## CONCLUSION

We report a case of calcifying cystic odontogenic tumor, which, although it is an odontogenic neoplasia, can be treated conservatively in most cases. New studies with larger series of cases are necessary to determine more precisely the frequency of recurrences and possible malignant transformations.

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