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Effectiveness of surgical decompression in the treatment of a calcifying cystic odontogenic tumor

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ABSTRACT

The calcifying odontogenic cystic tumor (CCOT) is a benign lesion of odontogenic origin characterized by an ameloblastoma-like epithelium with ghost cells that may calcify. Despite broadly considered as a cyst, some investigators prefer to classify it as a neoplasm. Clinically, it occurs predominantly during the third decade of life. No difference in gender prevalence has been observed nor predilection of the lesion between maxilla and mandible. The most affected region extends from the incisor tooth to bicuspids. The classic treatment of the lesion is full excision, although a different approach may be determined by the possible association with another odontogenic tumor. Depending on the tumor size and the vicinity with important structures, decompression may be undertaken before its complete removal. The present report describes a case of CCOT with large proportions, located at the right maxilla and extending to the maxillary sinus, nasal cavity, and orbital floor. The treatment option was surgical decompression as the initial procedure, with satisfactory outcome. After partial remission, the lesion was fully removed, and the post-operative follow-up was uneventful.

Keywords
Odontogenic Tumors; Decompression, Surgical; Curettage.

INTRODUCTION

The calcifying cystic odontogenic tumor (CCOT), previously known as calcifying odontogenic cyst (COC or Gorlin cyst) is currently classified as a benign cystic neoplasm of odontogenic origin, characterized by ameloblastoma-like epithelium with ghost cells that may calcify.[1]

This lesion has been differentiated from the calcifying epithelial odontogenic tumor (Pindborg tumor) and therefore is recognized as a distinct entity.[2] The odontogenic nature was suspected due to the following factors: 1) the presence of lesions associated with a developing or unerupted tooth; 2) histology exhibiting a basal cell layer consisting of ameloblastic cells and the production or induction of dentinoid material; and 3) lesion development close to dental structures.[2] Some researchers believe the neoplasm

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develops from the reduced enamel epithelium or from dental lamina remnants from the top of the dental follicle of an unerupted tooth.[3]

Clinically, the CCOT presents expansive behavior together with gingival and mucosal tenderness, although it remains asymptomatic unless secondary infection ensues.[2,4,5] The maxilla and the mandible are similarly affected, generally extending from the central incisor to the premolars teeth.[1,3,5,6] No gender predominance is observed[1,4,5] and patients may be affected between the first and the eighth decades of life. However, the lesion predominates between the second and the third decades of life, possibly associated with an odontoma.[1,3-6]

Imaging examination usually shows a unilocular radiolucency surrounded by a well-defined cortical margin. A varied quantity of calcified material may be present inside the lesion. Approximately one-third of the cases are associated with an unerupted tooth.[1,3,5]

Histological findings are characterized by a thick cystic wall with the basal cell layer showing cuboidal or columnar format. Some cells may resemble ameloblasts, due to the inverted and polarized nuclei away from the basal membrane[3]. Suprabasal overlying epithelial cells are loosely arranged, similar to the stellate reticulum of the enamel organ.[5,6] Isolated or clustered "ghost cells" in the epithelial lining are consistently found and may form large sheets. The ghost cells may show intracellular calcification. Nevertheless, these cells are not pathognomonic since they are frequently present in other tumors as well.[4] Cytokeratin, bcl-2 and Mel-CAM (CD146) expression are some immunohistochemical markers that were described in CCOT.[7]

CCOT may share histologic similarity with other odontogenic tumors like complex odontoma, ameloblastoma, odontoameloblastoma, ameloblastic fibroma, and ameloblastic fibro-odontoma.[5] Despite this similarity, CCOT may be a *de novo* lesion or it evolves from a pre-existing one, what may cause controversy regarding the tumor's clinical behavior and the treatment of choice. Therefore, CCOT has been classified as a distinct entity only if the lesion architecture presents a lumen with epithelial lining.[6]

The solid variants of CCOT (ghost cell odontogenic tumor and dentinogenic ghost cell tumor) are substantially more aggressive, probably because of the lumen invasion by keratinized ghost cells and/or giant cells, besides the frequent presence of calcification and dysplastic dentinoid material.[5,6,8] The malignant variant is rare.[5,6]

The objective of the present case report is to demonstrate the surgical decompression efficacy of a large CCOT located in the right maxilla extending to the maxillary sinus, nasal cavity, orbital floor and pterygopalatine fossa. In these specific cases, the excision in a one-step surgery could jeopardize important structures.

CASE REPORT

A 34-year-old male patient was referred to the Oral and Maxillofacial Surgery Department for the treatment of a calcifying odontogenic cyst that was localized in the right maxilla. He complained of a slow, progressive and painless growing lesion during the last 3 years. Physical examination revealed a bulging and hard mass, slightly effacing the right nasogenian fold. Intraoral examinations disclosed the expansion of the vestibular and palatal cortical bones (Figure 1).

The computerized tomography (CT) showed a large expansive lesion, with soft tissue attenuation, occupying the totality of the right maxillary sinus, reabsorbing the sinusal walls and obliterating the maxillary sinus ostium (Figure 2).

Due to the lesion size and its proximity to the lateral wall of the nasal cavity, orbital floor, and pterygopalatine fossa, it was was first decompressed under local anesthesia, what also rendered specimens for histological analysis. The cystic cavity was opened

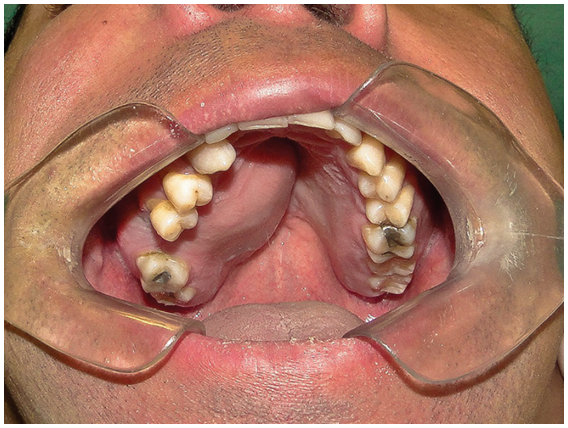

Figure 1. Intraoral aspect with accentuated expansion of vestibular and palatal cortical bone and displacement of adjacent teeth.

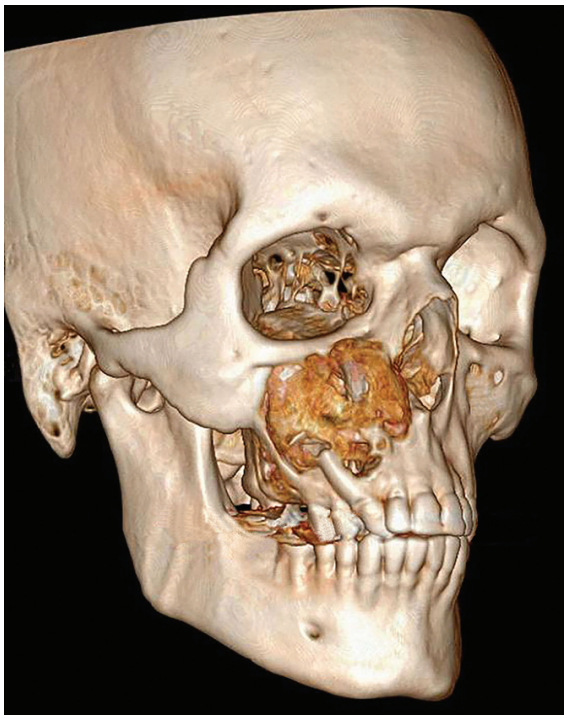

Figure 2. 3D reconstruction of CT of the face showing the bone expansion.

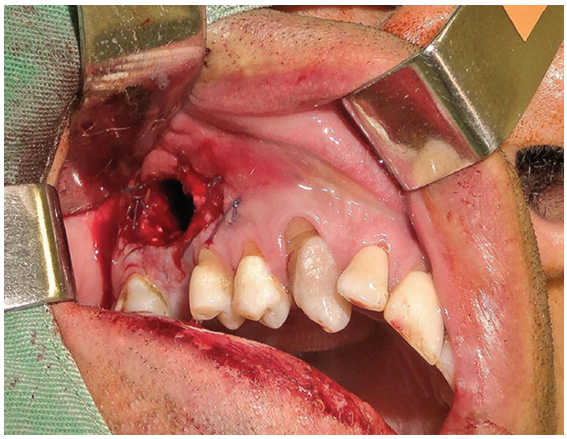

Figure 3. Immediate post-operatory after surgical decompression.

towards the oral cavity, and was left open until the second surgery, which was accomplished 9 months later (Figure 3).

Clinically, 3 months after the decompression, one was noticed partial regression of the lesion (Figures 4A-B).

CT scan 5 months after the decompression, also demonstrated satisfactory regression of the lesion (Figures 5A-C). A new surgical approach under general anesthesia was undertaken, allowing total excision. In this procedure, the maxillary sinus ostium could be maintained, keeping adequate sinus drainage.

The anterior wall of the right maxillary sinus and the former decompression orifice were exposed followed by an ostectomy to enlarge the access to the cyst contents.

The whole lesion was more easily excised due to a defined cleavage plan between the tumor and the remaining bone. Additional curettage was performed. Dental elements 12 and 13 were removed as no surrounding bone was present. With the aim of maintaining normal sinus drainage, a perforation was made to communicate the maxillary sinus to the nasal cavity through which a fine silicone probe was inserted and maintained by being stitched to the right nostril for 20 days (Figure 6).

The gross view of the specimen post-surgical excision is demonstrated in Figure 7.

Histological examination of the surgical specimen showed a fibrous cystic capsule with epithelial lining showing columnar basal cells, resembling ameloblasts, with somewhat loose upper layers containing isolated and clustered ghost cells, consistent with the diagnosis of CCOT (Figure 8).

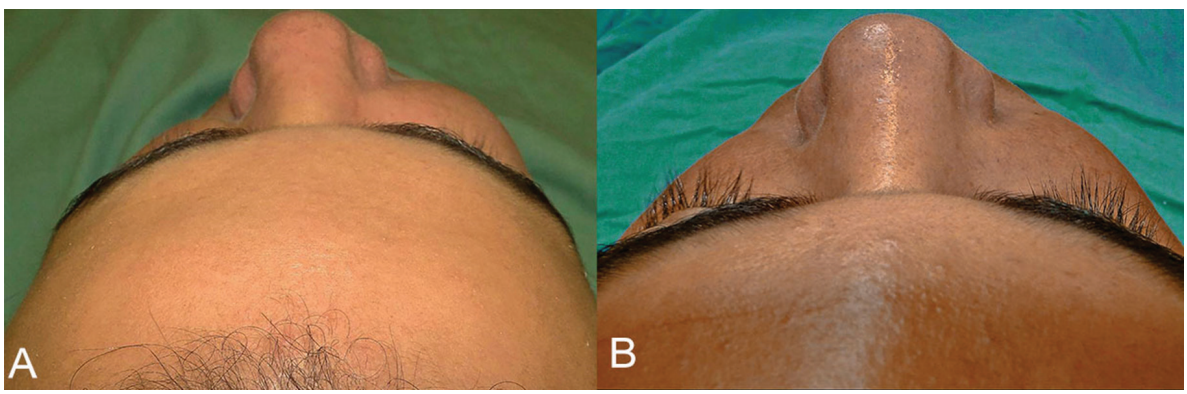

Figure 4. Clinical aspect in craniocaudal view. **A** – At clinical presentation; **B** – Three months after the decompression.

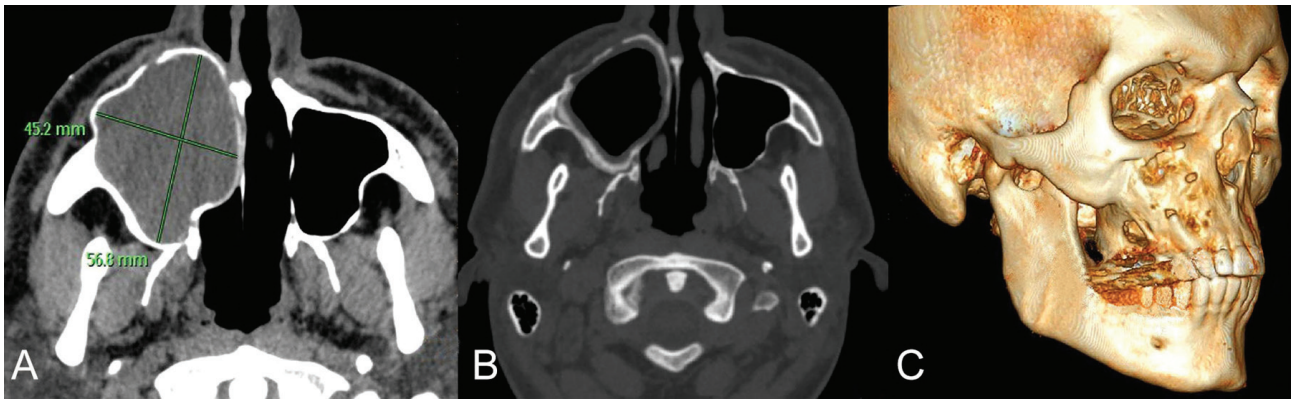

Figure 5. A – Facial CT - axial view before the decompression; **B** – Facial CT - axial view 5 months after the decompression. Note the bone formation circumscribing the lesion; **C** – Facial CT - 3D reconstruction 5 months after the decompression.

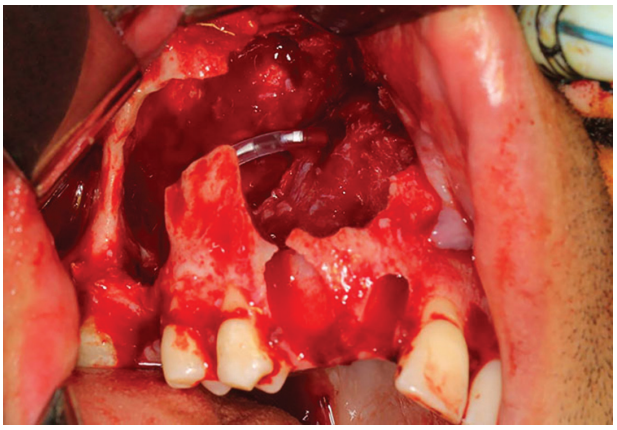

Figure 6. Intraoperative aspect after total removal of the CCOT. The silicone probe was left in place to maintain the sinus drain and promote further re-epithelization of the orifice to the nasal cavity.

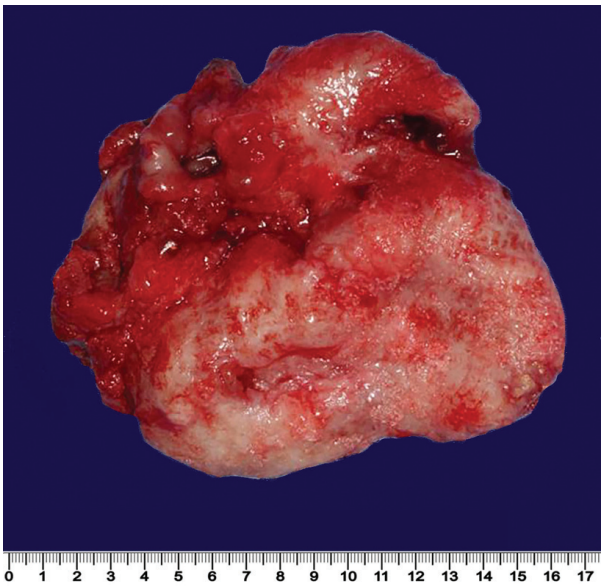

Figure 7. Gross view of the enucleated specimen.

The postoperative period was uneventful. Thirteen months after the full excision, the patient was completely recovered with no clinical signs of recurrence. At intraoral examination, there was still a cortical expansion of vestibular and palatal aspects of the right maxilla, but in a smaller size (Figure 9).

CT scans obtained 13 months after surgery showed bone formation inside the maxillary sinus as well as a radiolucent margin, suggesting the proliferative potential of healthy remaining sinus lining. Long-term annual clinical and imaging controls are expected in order to detect eventual recurrences (Figure 10).

DISCUSSION

Although the CCOT's epithelial cells may share similar features with ameloblastoma and other locally aggressive[6] neoplasms, the lesion has a cystic nature.[9] Depending on the quantity of ghost cells[8] and the epithelial lining proliferation within the lesion cavity, tumoral aggressive behavior[2] and recurrences are to be more expected.[4-6] A multicystic variant may also occur, showing similar CCOT architecture, but on histology, this variant is surrounded by multiple satellite cysts that are enclosed in the epithelial lining.[4,5,8]

Due to the variety of classifications of the present lesion, to comprehend it as a single entity is controversial.[1,4] Therefore, both clinical and histological features, and the association with another neoplasm are taken into account in a more precise method of elucidation.[4,5,9-11]

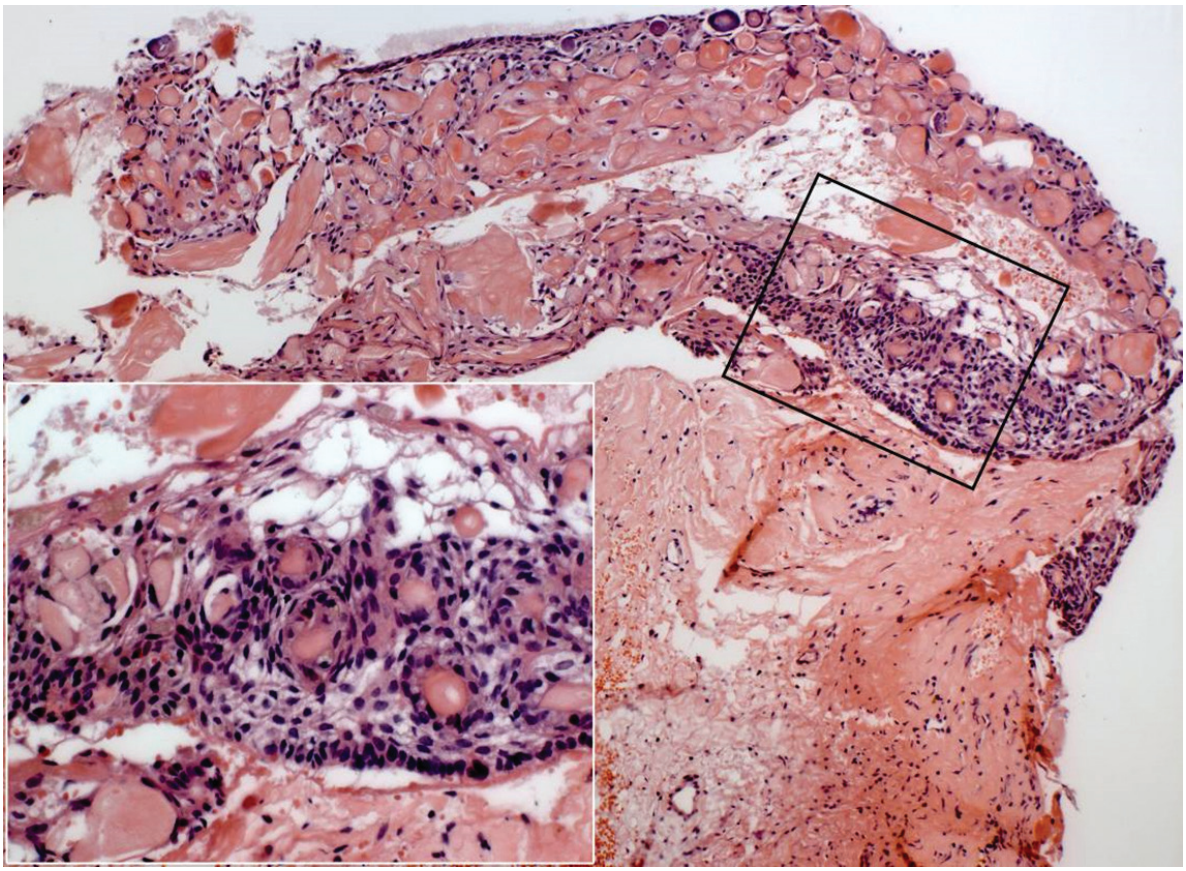

Figure 8. Photomicrographs showing cyst epithelial lining with a columnar basal layer and isolated or clustered eosinophilic ghost cells of different sizes (HE, 100x). Insert shows these features in detail (HE, 400x).

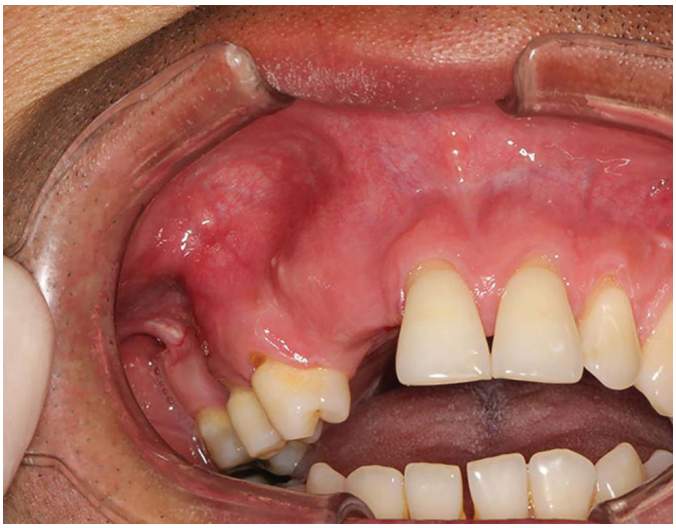

Figure 9. Intraoral examination thirteen months after excision.

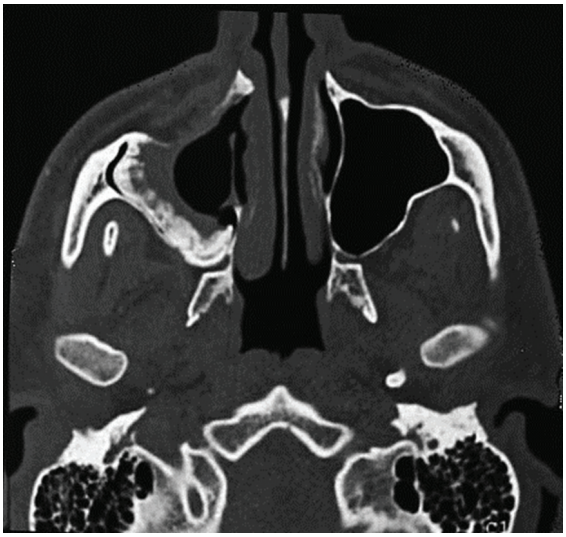

Figure 10. Axial CT view of 2 years and 1 month post-operatory follow-up. There is a radiolucent margin lining the sinus and the presence of air inside it.

The usual and current treatment of the CCOT is the one-step total excision.[2,4] In the present case, one was chosen to proceed with decompression initially, taking into account the large size of the lesion and the involvement of neighboring structures, similar to the management of the unicystic ameloblastoma.[12] Other more aggressive lesions, like keratocystic odontogenic tumor, have a satisfactory response to this initial conservative approach.[12-14] The effectiveness of the surgical decompression is supported by the theory that

with the reduction of the inner cystic pressure the lesion progression will be hampered, thereby promoting its regression.[14] Although no contraindication for surgical decompression has been reported; the patient may complains of the unpleasant smelling even after thorough hygienic maneuvers with repeated saline irrigation of the open cavity. With this first conservative approach total regression may take years to occur.[13] Meticulous post-decompression follow-up is highly advisable, based on radiographic patterns and clinical behavior, since the CCOT epithelium lining may present an over proliferation in response to this surgical procedure[2,13,15] and the tumor may change from an expansive lesion to an invasive one.[13]

A thorough histological examination including the whole specimen is mandatory, as associated neoplasia may be present. In the case reported herein, a marked inflammatory infiltration was observed throughout the capsule. This is an uncommon finding unless the ghost cells are exposed to connective tissue, thereby triggering a foreign body reaction.[7] We believe that this inflammatory reaction was due to the free contact of the oral cavity with the cystic lesion.

FINAL CONSIDERATIONS

The histological findings of CCOT may induce several diagnostic controversies, mainly due to the possibility of the presence of associated neoplasms, implying in different therapeutic approaches.

In the case of large-size and non-solid CCOTs surgical decompression may be a therapeutic option to allow tumoral regression before second and definitive surgical procedure aiming complete excision. This two-step approach prevents injury to adjacent structures and facilitates local healing. Long-term clinical and imaging follow-up is highly advisable due to the possibility of recurrence.

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