Ghost Cell Odontogenic Tumor Associated with Odontoma – Report of Two Rare Cases

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The ghost cell odontogenic tumor (GCOT) is a neoplastic/cystic lesion with a diverse histopathological and clinical behavior. It was formerly known as calcified odontogenic cyst, but in 2005 the World Health Organization categorized this lesion as an odontogenic, benign tumor rather than a cyst; nominating this neoplasm as calcifying cystic odontogenic tumor. A later comprehensive classification named it ghost cell odontogenic tumor, because the most remarkable histopathologic characteristic is the presence of a mass of ghost cells embedded in the epithelium. We report two cases of a rare variant of a ghost cell odontogenic tumor associated with odontoma; to our knowledge, one is the youngest patient (four month old) reported in the English literature.

Keywords: *ghost cell odontogenic tumor, calcified odontogenic cyst, children.*
J Clin Pediatr Dent 36(4): 373–376, 2012

INTRODUCTION

The ghost cell odontogenic tumor (GCOT), also called calcifying cystic odontogenic tumor (CCOT), is a neoplastic/cystic lesion with a diverse histopathological and clinical behavior. This lesion was known as calcified odontogenic cyst. In 1962, Gorlin *et al* recognized the resemblance of this lesion with the cutaneous calcifying epithelioma of Malherbe.[1] In 1981 Praetarious *et al* identified the lesion as a cystic and tumor-like entity.[2] Later in 2005 the WHO classified this lesion as an odontogenic, benign tumor rather than a cyst; nominating the neoplasm as calcifying cystic odontogenic tumor. GCOT, as other odontogenic tumors or cysts has a peripheral counterpart, located in gingiva, in about 25% of the cases. Upon reviewing 122 cases, Ledesma-Montes *et al*[3] named it ghost cell odontogenic tumor and developed a comprehensive and simplified classification

GCOT may occur at any age, however, is most common in the second or third decade of life. To our knowledge, the youngest patient reported up to now, with this tumor, was four years old. There is not gender preference and anterior maxilla is the most common location. Mandibular cases seem to be more often located in the molar area. Early central GCOT may be discovered incidentally as a well defined radiolucent lesion, sometimes associated with the crown of an impacted tooth. However, as they mature, they cause expansion and intralesional calcifications. The radiopacities can be seen as salt and pepper, cloud-like or an eccentric, (crescent-shaped) area. The most common type is the cystic variant. Other types of GCOT are very rare. The most remarkable histopathologic characteristic of GCOT is the presence of a mass of ghost cells embedded in the epithelium.[4]

Case Report 1

Eleven year old female, presented with her mother to the oral and maxillofacial surgery clinic with the chief complaint of fast growing swelling of the jaw. (Figure 1) Medical history was unremarkable except for appendectomy some years previously. Extraoral examination revealed a well nourished other ways healthy patient. Intraoral exam disclosed mild buccal and lingual expansion of the anterior mandible. Teeth 21 to 24 presented severe mobility. Perforation through the lingual cortex was evident. Radiographies show a radiolucent, well circumscribed lesion with a radiopacities. (Figures 2, 3, 4, 5) Aspiration yielded straw colored fluid. With the diagnosis of adenomatoid odontogenic tumor with cystic component vs calcifying odontogenic cystic tumor, an incisional biopsy was performed. The histology revealed a cystic lining showing

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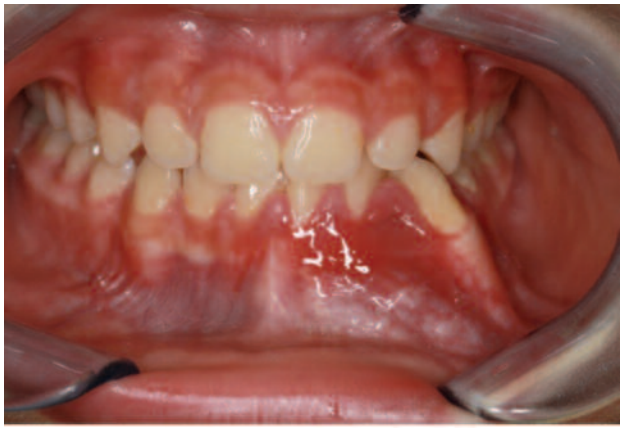


Figure 1. Clinical swelling left mandibular area. Displacement of teeth is evident.

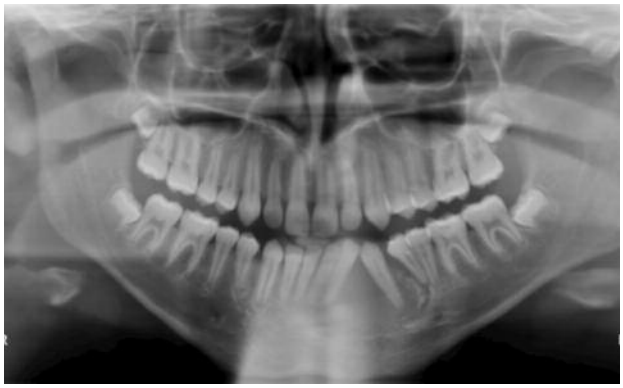


Figure 2. Panoramic radiograph demonstrating a well-defined, radiolucent lesion with multiple small radiopacities that is extending from the mesial area of #21 towards the distal region of #25 of approximately 30 mm in diameter. Notice the displacement of #21, #22, #23 and #24.

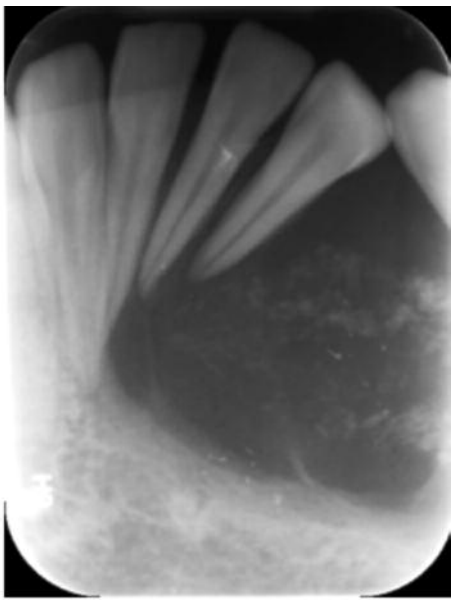


Figure 3. Periapical radiograph of left mandibular anterior region indicating a well defined radiolucency with multiple internal calcifications and displacement of #21, #22, #23 and #24.

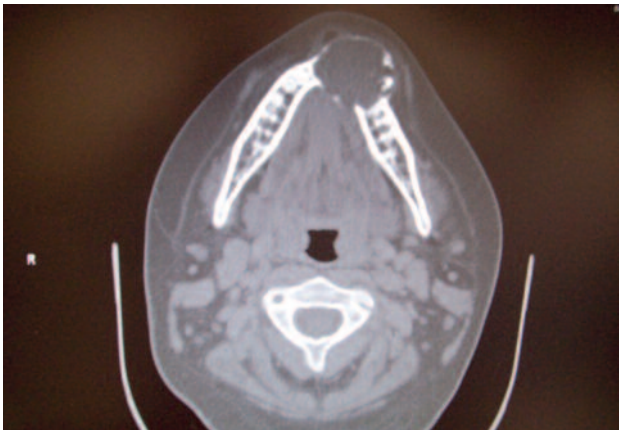


Figure 4. Axial Contrast-Enhanced Computed Tomography (CECT) of the mandibular region reveals an expansile, low density lesion with internal foci that is causing thinning of the buccal and lingual cortices. Notice the teeth displacement and the extension anterior to the platysma muscle and posterior to the floor of the mouth.

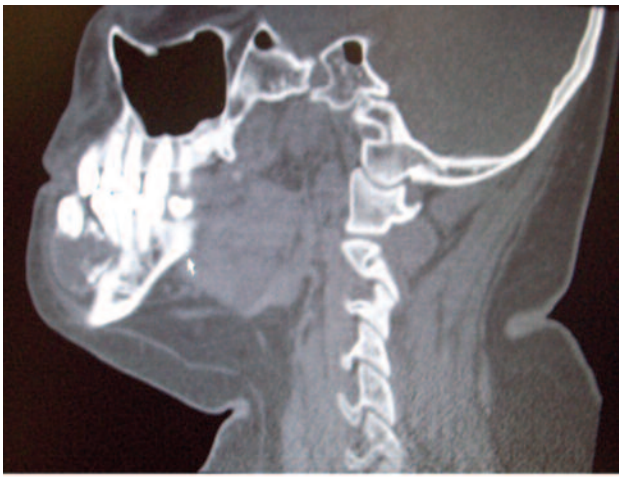


Figure 5. Saggital Contrast-Enhanced Computed Tomography (CECT) shows the expansile, heterogeneous density lesion producing thinning of the buccal, mandibular cortex, tooth displacement.

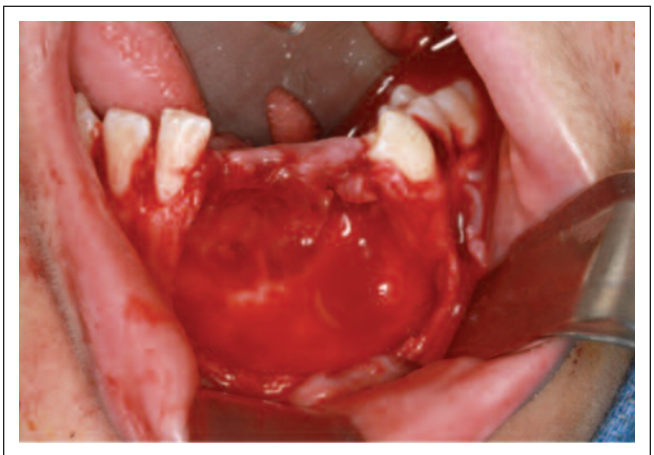


Figure 6. Biopsy procedure. Buccal bone expansion is noted.

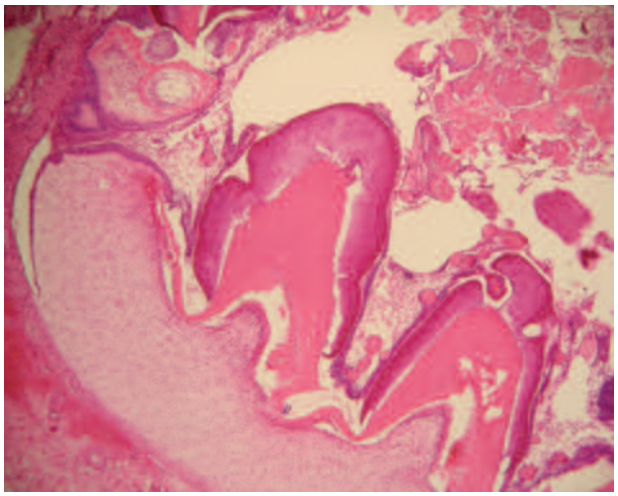


Figure 7. 40X H&E sections of a specimen composed of cystic epithelium with odontoma formation and a large area of pulp-like tissue.

reverse polarization and the typical ghost cells, dentin, enamel matrix, cementum and pulp tissues. (Figures 6, 7)

Case Report 2

Four month old infant exhibited a fast growing swelling of the mandible. Pregnancy, including ecographies, was normal and it was a full term baby. The area didn't appear to be painful and was covered by normal mucosa. MRI and CT showed a large cystic mass with marginal calcifications and teeth during early developmental stage. (Figures 8, 9) The baby was taken to the operating room for enucleation of the lesion. The microscopic examination revealed a cystic lesion showing ghost cells, palisading of the basal cell layer of the cystic epithelium and proliferation of dentin, enamel and cementum.

Both cases were diagnosed as calcifying cystic odontogenic tumor type 2, (associated with odontoma) Enucleation and curettage were curative for these cases.

DISCUSSION

The variations in clinical, radiographic and histologic features of ghost cell odontogenic tumor are a reminder of the wide possibilities regarding behavior and prognosis. This entity may manifest as an extremely indolent cyst, can present different levels of aggressiveness and can be a malignant neoplasm. A simplified classification was proposed by Ledesma *et al*:[3]

1. Calcifying cystic odontogenic tumor. (CCOT)
   a. Types 1: Cystic. The most common type
   b. Type 2: Associated with odontoma
   c. Type 3: With ameloblastomatous proliferation
   d. Type 4: Associated with other odontogenic tumors

2. Dentinogenic ghost cell tumor. (DGCT)
   a. Type 1: Central
   b. Type 2: Peripheral

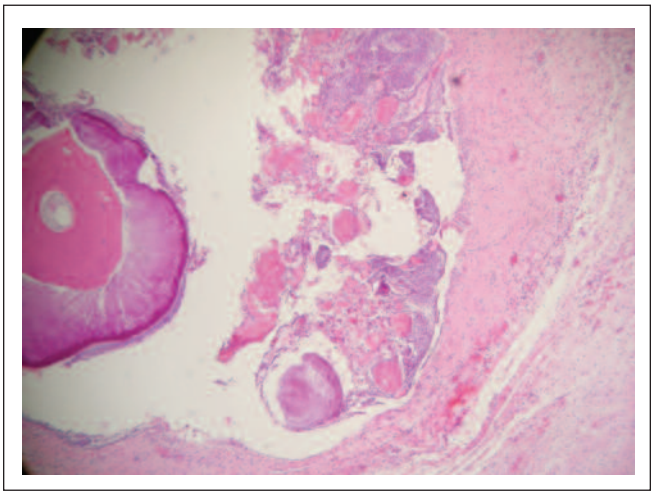


Figure 8. Case 2: 40X H&E sections of a specimen composed of cystic epithelium with ghost cells, enamel, dentin and pulp tissues.

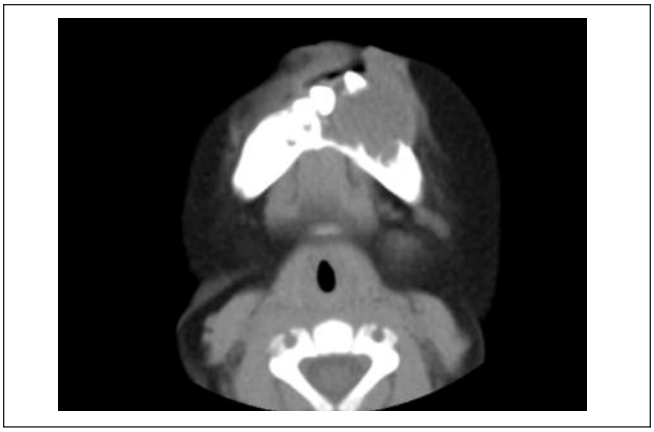
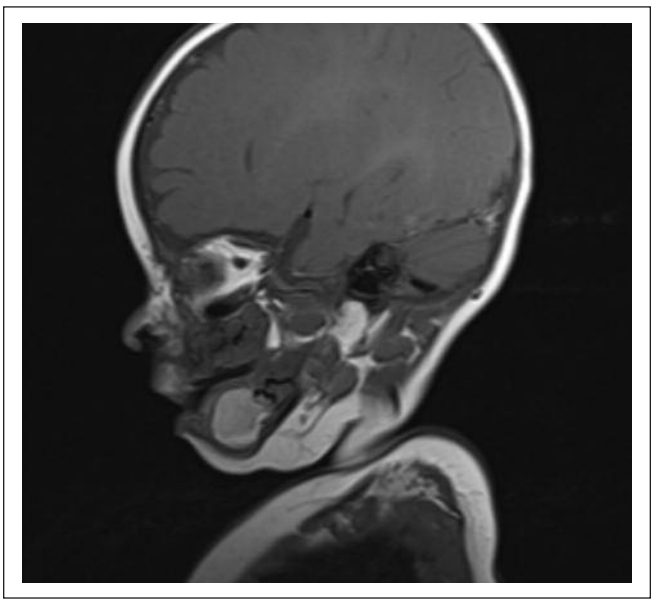


Figure 9. Case 2: Axial soft tissue window Computed Tomography depicts an expansile cystic lesion within the left mandibular anterior region. Thinning of the lingual and buccal mandibular cortex with teeth displacement and soft tissue swelling is demonstrated.



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3. Ghost cell odontogenic carcinoma (GCOC):
   a. Arising de novo
   b. GCOC ex Calcifying cystic odontogenic tumor.
   c. GCOC ex Dentinogenic ghost cell tumor.

The differential diagnosis of this condition is dentigerous cyst, when associated to an impacted tooth and no radiopacities are present. COCT, CEOT, ameloblastoma and ameloblastic odontoma are other possibilities. The final diagnosis is obtained with biopsy and the treatment depends on the type of GCOT.[5,6]

CONCLUSION

The pediatric dentist should be aware of the variability of this lesion and the possibility of development in newborn patients. They should consider it in the differential diagnosis of radiolucent or mixed conditions. To our knowledge, this is the youngest patient reported with a central ghost cell odontogenic tumor.

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