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A unique case of a calcifying cystic odontogenic tumor

Shoko Gamoh[1*], Yukako Nakashima[1], Hironori Akiyama[1], Kimishige Shimizutani[1], Takuro Sanuki[2], Junichiro Kotani[2], Koji Yamada[3], Shosuke Morita[3], Kazuya Tominaga[4], Masahiro Wato[4], Akio Tanaka[4]

[1]Department of Oral Radiology, Osaka Dental University, Osaka, Japan
[2]Department of Anesthesiology, Osaka Dental University, Osaka, Japan
[3]Department of Oral and Maxillofacial Surgery, Osaka Dental University, Osaka, Japan
[4]Department of Oral Pathology, Osaka Dental University, Osaka, Japan
Email: [*]gamo-s@cc.osaka-dent.ac.jp

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ABSTRACT

The calcifying odontogenic cyst was first reported by Gorlin *et al*. in 1962. At that time, it was classified as a cyst related to the odontogenic apparatus, although it was later renamed as a calcifying cystic odontogenic tumor by the WHO calcification in 2005 due to its histological complexity, morphological diversity and aggressive proliferation [2]. Here, we describe a case of a calcifying cystic odontogenic tumor in a 4-year-old boy. The lesion was surgically removed, and the histopathological examination revealed it to be a cystic tumor with ghost cells, a stellate reticulum and small amount of dentinoid tissue in the cystic wall.

Keywords: Calcifying Cystic Odontogenic Tumor; Diagnostic Imaging; Computed Tomography

1. INTRODUCTION

The calcifying cystic odontogenic tumor (CCOT), which is also known as a calcifying odontogenic cyst (COC) or Gorlin cyst, is a rare developmental lesion that arises from the odontogenic epithelium [1]. Although it has commonly been recognized as a benign odontogenic cyst since its original description by Gorlin *et al*. in 1962, this pathologic entity encompasses a spectrum of clinical behaviors and histopathological features including cystic, neoplastic and aggressive variants. As a result of this diversity, different classification schemes and nomenclatures for this type of lesion have been suggested. In 2005, the World Health Organization Classification of Head and Neck Tumors reclassified the lesion as an odontogenic tumor and gave it the name of "calcifying cystic odontogenic tumor" (CCOT) [2].

[*]Corresponding author.

CCOT is a rare lesion that represents about 2% of all odontogenic pathological changes in the jaw [3-5]. It is clinically characterized as a painless, slow-growing tumor, which equally affects the maxilla and mandible, has a predilection for the anterior region of the jaw and usually arises intraosseously, although it may also occur extraosseously. It has a peak incidence during the second and third decades on life, with a mean age of 30.3 years and does not demonstrate any gender predilection [6]. The treatment of choice for cases of CCOT is conservative surgical enucleation. However, recurrence is frequent, especially in neoplastic cases, such as dentinogenic ghost cell tumor [7].

The typical histopathological features of CCOT include a fibrous wall and lining of odontogenic epithelium with columnar or cuboidal basal cells resembling ameloblasts. Stellate reticulum-like cells overlay the basal cell layer, and "ghost cells", which may occasionally become calcified, are also situated in the cyst lining [8].

Radiographically, CCOT may appear as an unilocular or multilocular radiolucent area with either well-circumscribed or poorly-defined margins that also may be observed in association with unerupted teeth [3,7]. Calcification is an important radiographic feature for the interpretation of CCOT, but is detected in only approximately half of the reported cases [9].

This report describes a case of CCOT that occurred in the maxilla of a 4-year-old Japanese boy.

2. CASE REPORT

A 4-year-old boy was referred to the Osaka Dental University Hospital by his home doctor due to 6-month history of asymptomatic swelling in the upper canine area. However, at the time that his mother sought the check-up,

swelling had increased in size and his nose had been bleeding profusely. He had no other concurrent diseases.

The clinical examination revealed the presence of a firm and localized lesion, although the oral mucosa was normal. His mother reported that his pregnancy and birth were normal. In addition, he had no history of traumatic dental injury. The extra-oral examination revealed a discrete facial asymmetry due to swelling in the middle of the face that was close to the right nasal alar rim (**Figure 1**). Conventional radiographs showed a well-defined unilocular, golf ball sized and round radiolucent lesion, extending from tooth 12 to tooth 17 and representing the inter-alveolar septum of the right upper canine and premolar. The lesion included impacted tooth and contained several calcified fragments (**Figures 2(a)** and **(b)**). Computed tomography images revealed a well-defined, expansive lesion with thin cortical margins and containing irregular radiopaque masses. No root divergency or destructive conditions were observed (**Figures 3(a)-(c)**). The differential diagnosis involved analyzing the likelyhood that the mass was a calcifying cystic odontogenic tumor, calcifying epithelial odontogenic tumor, or adenomatoid odontogenic tumor.

Under general anesthesia, an excisional biopsy was carried out (**Figures 4(a)** and **(b)**), and the histopathological examination revealed the presence of cellular and densely fibrous connective tissue. In the overlying layer, cells were loosely and diffusely arranged together with several “ghost cells”, some of which presented dystrophic calcification (**Figures 5(a)** and **(b)**). A microscopic diagnosis of CCOT was made.

There are no signs of recurrence for a year and a half since the complete removal of the tumor in October 2011 (**Figure 6**).

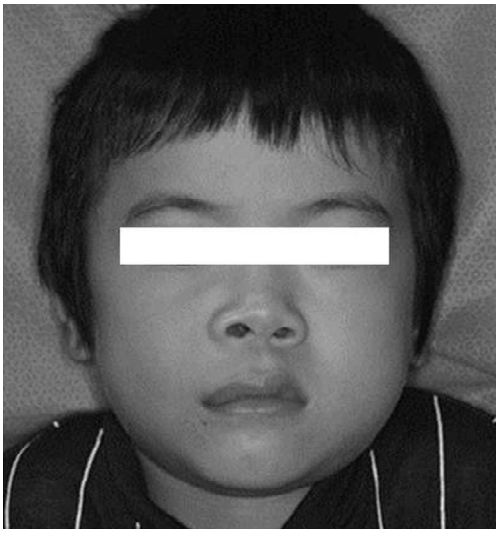
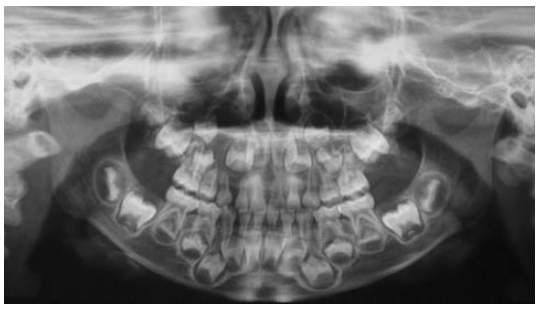
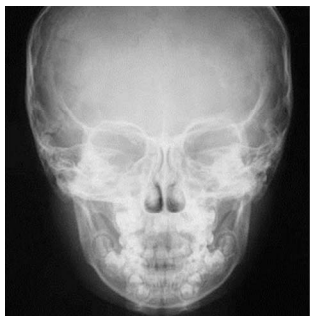


Figure 1. Front face photograph shows discrete facial asymmetry due to swelling in the middle of the face that is close to the right nasal alar rim.

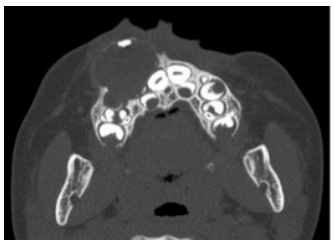


(a)

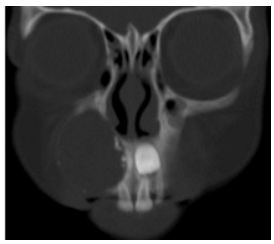


(b)

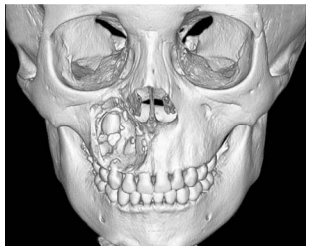
Figure 2. Conventional radiographs. (a) Panoramic and (b) posteroanterior radiographs showed a well-defined unilocular radiolucent lesion associated with radiopaque bodies in the right upper premolar area.



(a)



(b)

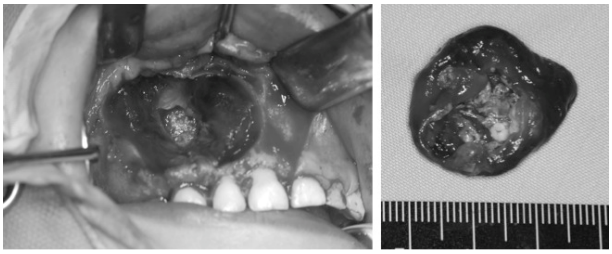


(c)

Figure 3. Computed tomography images. (a) Axial view of CT image, (b) coronal view of CT image, and (c) 3-dimensional reconstructed image of CT image reveal a well-defined and expansive lesion with thin cortical margins and irregular radiopaque masses.

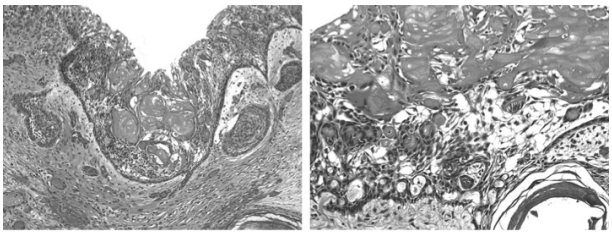
3. DISCUSSION

COC, a rare lesion first described by Gorlin *et al.* [1], has recently been defined as calcifying cystic odontogenic tumor (CCOT) by the World Health Organization (WHO)



(a) (b)

Figure 4. Operation findings. (a) A site of operation. (b) The extracted lesion 25 mm in diameter. The radiopaque masses found on CT image revealed to be a calcified portion in the mass.



(a) (b)

Figure 5. Histopathological findings. (a) The cystic wall is lined by an ameloblast-like epithelium with columnar and cuboidal basal layer. Numerous ghost cells and calcified particles are present within the epithelial lining (hematoxylin-eosin stain, original magnification ×10). (b) Stellate reticulum cells are observed within the epithelial lining (hematoxylin-eosin stain, original magnification ×20).

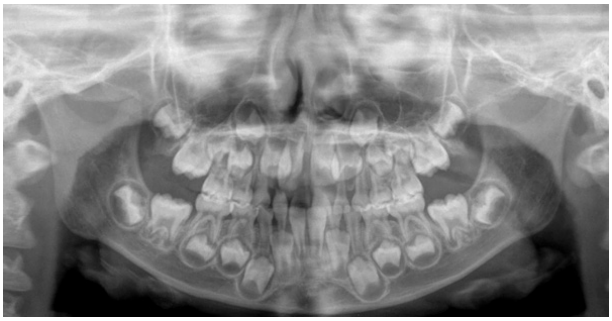


Figure 6. A panoramic radiograph on a year and a half after the operation showed no signs of recurrence.

due to its neoplastic behavior [2]. Alternative names have also been used, such as "keratinizing and calcifying odontogenic tumor" [10,11], "calcifying ghost cell odontogenic tumor" [12], "dentinogenic ghost cell tumor" [13-15] and "epithelial odontogenic ghost cell tumor" [16,17]. The existence of such a variety of terms reflects both the wide clinicopathological spectrum of the formerly named COC and the confusion regarding its cystic or neoplastic nature.

Most CCOTs, as demonstrated by our case, are asymptomatic and often discovered incidentally on radiographic examinations [18]. Because this lesion arises in tooth-bearing areas of the jaws or gingival areas, CCOTs are often located in a periapical or lateral periodontal relationship to the adjacent teeth. Radiographically, they are clearly-delineated and appear as unilocular or multilocular radiolucencies, with calcifications of variable density noted in one-third to one-half of cases. CCOTs can occur alone or in association with other odontogenic tumors such as odontomas (20%), adenomatoid odontogenic tumors and ameloblastomas [18]. However, this association is a challenge for diagnosis using only conventional images, due to the presence of numerous overlapped images of anatomical structures of the jaw region. Root resorption and divergency are common radiographic findings, and an association with an impacted tooth occurs in approximately one-third of cases [19].

Enucleation is the treatment of choice for most intraosseous CCOTs and few recurrences have been reported in the literature [2].

CCOT is an uncommon entity. Although rare, calcifying epithelial odontogenic tumor and adenomatoid odontogenic tumor should be included in the differential diagnosis of jaw lesions. The chief danger is the losing of permanent teeth [20]. Despite their unknown pathogenesis, CCOT should be treated conservatively, and preservation of unerupted teeth is encouraged, especially when intervention occurs at a young age.

In conclusion, this was an observed case of CCOT that was associated with an impacted tooth. A satisfactory esthetic outcome without sequelae was obtained.

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