Calcifying Cystic Odontogenic Tumor in a 5-Year-Old Boy: A Case Report

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Abstract The calcifying cystic odontogenic tumor (CCOT), formerly known as calcifying odontogenic cyst, is a rare developmental neoplasm/cyst of odontogenic epithelial origin with considerable histopathologic diversity and variable clinical behaviour. The occurrence of CCOT in the first decade of life is very uncommon. We report an interestingly rare variant of CCOT with ameloblastomatous proliferation affecting the mandibular left posterior region associated with an impacted permanent tooth in a 5-year-old boy.

Keywords Boy · Calcifying cystic odontogenic tumor · Mandible

Introduction

Calcifying odontogenic cyst (COC) was initially described as a separate pathologic entity by Gorlin et al. [1] in 1962 as a possible oral analogue to the calcifying epithelioma of Malherbe (pilomatrixoma) of the skin, owing to the presence of ghost cell keratinization in both. In 2005 the WHO classified this lesion as an odontogenic, benign tumor rather than a cyst, re-naming this entity as calcifying cystic odontogenic tumor (CCOT) [2]. It can occur at any age, although majority of the cases appear before the fourth

decade of life, affecting equally maxilla and mandible without sex predilection [3, 4]. The majority of CCOTs are cystic in architecture but on rare occasions they appear as solid lesions [4–7]. An associated unerupted tooth is seen in one-third of the cases [4]. Histologically the lesion is characterized by well-delineated cystic proliferation of odontogenic epithelium with ghost cells and fibrous connective tissue wall. The occurrence of CCOT in the first decade of life is very uncommon. This paper reports a case of CCOT involving the mandible in a 5-year-old boy.

Case Report

A 5-year-old boy was referred by the local dentist for evaluation of a painless, slow growing swelling in the left posterior mandible of 1 month duration (Fig. 1). Intraorally the swelling extended from the buccal vestibule of deciduous mandibular left second molar to the edentulous mandible posteriorly. Palpation revealed an asymptomatic vestibular bone enlargement with crepitation. The overlying mucosa was intact and normal in color. No cervical lymphadenopathy or sensory deficits were present. The medical history was non-contributory. A panoramic radiograph revealed a 3×2 cm ill defined unilocular radiolucency around the crown of a developing permanent left second molar (Fig. 2).

Based on the clinic-radiological findings differential diagnosis of benign odontogenic lesions like dentigerous cyst, odontogenic keratocysts, ameloblastic fibroma, unicystic ameloblastoma and adenomatoid odontogenic tumor were considered. In the current case, the presence of pericoronal radiolucency associated with a developing unerupted left permanent second molar and in consideration of the patient's age, the working diagnosis of

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Fig. 1 Pre-operative photograph showing extra-oral swelling on the left side of the face

Fig. 2 The panoramic radiograph showing a well-defined unilocular radiolucency in the left mandibular posterior region associated with a developing permanent mandibular second molar

dentigerous cyst was made. A preoperative incisional biopsy may have been desirable, but considering the age of the patient and the benign radiographic appearance of the lesion, enucleation as the initial approach was preferred. The lesion was enucleated with developing crown of the permanent mandibular left second molar and the permanent mandibular left first molar was also extracted under general anesthesia. The specimen was sent for histopathologic examination. Macroscopic examination showed a cystic

specimen measuring about $3 \times 2 \times 2$ cm in size associated with an impacted crown of developing permanent left 2nd molar. Cut surface of the cystic specimen showed luminal proliferation in some areas (Fig. 3). Microscopic examination of the hematoxylin and eosin stained sections showed a cystic lumen lined by odontogenic epithelium consisting of a prominent basal layer of columnar cells with hyperchromatic nuclei intermingled with islands of ghost cells. Areas of intraluminal proliferations, remaining in continuity with the luminal epithelium, showed elongated epithelial strands with ameloblastic differentiation showing columnar cells at the periphery and stellate reticulum-like cells associated with focal calcifications (Fig. 4). The cystic wall was composed of fibrous connective tissue. The final histopathologic diagnosis of CCOT type 3 was made. The patient's post-operative follow-up of 2 years duration has been satisfactory and free of

Fig. 3 Cut surface of the surgical specimen showing a cystic lesion with intraluminal proliferations

Fig. 4 Photomicrograph showing numerous ghost cells and calcifications in the cyst lining (H&E $\times 50$)

Fig. 5 Post-operative panoramic radiograph showing bone deposition in the left mandibular posterior region after 2 years

recurrence (Fig. 5). Written consent and approval for publication was obtained from the patient's parents.

Discussion

The 2005 WHO classification of odontogenic tumors described CCOT as a benign neoplasm related to odontogenic apparatus and defined it as 'a cystic lesion in which the epithelial lining shows a well-defined basal layer of columnar cells, an overlying layer that is often many cells thick and masses of ghost epithelial cells that may be in the epithelial cyst lining or in the fibrous capsule [8]. It can present as either a cyst or a neoplasm and may be associated with an odontoma, ameloblastic fibro-odontoma, ameloblastic fibroma or supernumerary tooth. The CCOT usually manifests itself as a painless, slow-growing swelling, anterior to the first molar region. Most series of cases dealing with CCOT report no sex predilection, and equal distribution between maxilla and mandible is typically seen [4, 6].

The CCOT may occur at any age, with a peak occurrence in the second and third decades with a mean age of 33 years. The occurrence of CCOT in the first decade of life is very uncommon. To the best of our knowledge, there are 45 documented cases of CCOT occurring in the first decade of life in the English literature (Table 1), of which the youngest recorded case was 2 days old [6]. The low frequency of odontogenic cysts and tumors in the first decade of life, as well as the uncharacteristic clinical and radiographic appearance, makes a preoperative diagnosis of CCOT very difficult, as seen in the present case.

Histologically the epithelial lining shows a well defined basal layer of hyperchromatic columnar cells with an upper cell arrangement of the stellate reticulum-like cells intermingled with mass of ghost cells [5]. After reviewing 122

Table 1 Frequency of COC in first decade of life

Author	Number of cases
Gorlin et al. [1]	1
Fejerskov and Krough [8]	5
Eda et al. [9]	1
Altini and Farman [10]	4
Freedman et al. [3]	3
Praetorius et al. [5]	1
Saito et al. [11]	1
Shear [12]	5
Wright et al. [13]	9
Shamaskin et al. [14]	2
Toida et al. [15]	1
Hong et al. [6]	1
Buchner [4]	8
Oliveira et al. [16]	1
Lin et al. [7]	1
Hogge et al. [17]	1
Total	45

cases Ledesma-Montes et al. [18] classified CCOT into 4 types, namely, type 1 (cystic), type 2 (associated with odontoma), type 3 (with ameloblastomatous proliferation) and type 4 (associated with other odontogenic tumors). Based on this, present case was diagnosed as CCOT type 3. This lesion is uncommon and shows considerable diversity in its clinical presentation and in its biological behaviour, because of its histological complexity and morphologic diversity, it is still debated whether CCOT is a cyst or neoplasm. The question raised by Shear, whether CCOT that have features of other odontogenic tumors developed this tumor secondarily, or whether the CCOT is itself a secondary phenomenon in an odontogenic tumor, remains unanswered [12]. Taking the potentials of odontogenic epithelium into consideration, both possibilities are plausible. Enucleation seems to be the appropriate treatment for most lesions, and the prognosis is considered to be good. Although rare, recurrences of both cystic and solid lesions of CCOT have been reported after one to 8 years [3], therefore adequate follow-up over a period of 10 years would be desirable [3, 19]. Although malignant transformation of CCOT have been reported [20, 21], not a single case in pediatric patients have been documented so far.

Conflict of interest None.

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