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Rare presentation of two distinct benign odontogenic tumors in a single jaw: Clinical, radiological and histological findings with a brief review of literature

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A B S T R A C T

A wide variety of odontogenic cysts and tumors are seen in human jaw as a result of complex interaction between diverse cellular elements of embryogenesis. Calcifying ghost cell odontogenic tumor (CGCOT) and odontome are benign odontogenic tumors and constitutes 0.37–2.1% and 4–67% of all odontogenic tumors respectively. The most notable features of this pathologic entity are histopathological features, which include a cystic lining demonstrating characteristic "Ghost" epithelial cells with a propensity to calcify. Here we report a unique and probably the first case of concomitant occurrence of CGCOT with odontome and compound composite odontome at two different site of same jaw (mandible) with brief clinical, radiological and histological study along with a brief review of literature.

1. Introduction

Odontogenic tumors are lesions derived from epithelial and/or mesenchymal elements of the tooth-forming apparatus and are, therefore, found within the jawbone or the soft tissue overlying the teeth [1]. These "tooth development" associated tumors are generally benign, but several odontogenic tumors are locally invasive and aggressive in nature, with a high rate of recurrence and a potential for neoplastic transformation [1,2]. The calcifying ghost cell odontogenic tumor (CGCOT) is a rare example of a developmental odontogenic tumor, comprising about 0.37%–2.1% of all odontogenic tumors [3]. Simultaneous occurrence of multiple odontogenic tumors in the same lesion is a well-known fact in the literature [4] but the concomitant occurrence of two different odontogenic tumors at two different sites in the same jaw is a rare phenomenon. Here, we report a case of 12-year-old child with two distinct benign odontogenic tumors in the same jaw (mandible) along with a brief discussion.

1.1. Report of a case

A 12-year-old male patient reported to the outpatient department with the complaint of a persistent swelling in the left side of the lower jaw since 4 months. Extra-oral examination showed facial asymmetry with prominence on the left angle region. Palpation revealed a tender, bony hard expansion of mandible intra-orally in the left alveolo-buccal region distal to 37. Panoramic radiograph revealed a 25 mm × 20 mm unilocular, radiolucent lesion involving most of the left ramus of mandible associated with a small radiopaque area present just distal to roots of 37. A calcified mass with radiolucent halo resembling an odontome was also found adjoining the right mandibular second permanent molar (Fig. 1). The tooth germs of mandibular 3rd molar were missing and the

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lesions occupied their prospective sites bilaterally. No aspirate could be obtained from the lesion. Based on the clinical and radiographic findings, differential diagnoses of adenomatoid odontogenic tumor, fibro-osseous lesion and ameloblastic fibro-odontoma were considered for the lesion on the left side. An incisional biopsy was performed for the lesion on the left side under local anesthesia. Histopathology of this lesion was suggestive of a Calcifying Ghost cell Odontogenic Tumor (CGCOT) in association with an odontome. Enucleation of the hybrid lesions was carried out under general anesthesia. In the same setting, the lesion on the right side was also removed. Histopathological examination of specimen from left side showed a cystic wall composed of plump fibroblasts with few areas lined by thin stratified squamous epithelium of 2–3 cell layers showing basal budding at places. The loose connective tissue stroma showed few tiny islands and cords odontogenic epithelium with few globules of ghost cells. Few foci of calcification were also present. Areas of embryonic connective tissue resembling dental papilla were observed (Fig. 2a and b).

Histopathological examination of specimen from the right side showed a uniform layer of enamel matrix lined on the surface by ameloblasts and cells of the enamel organ. Deeper tissue showed tubular dentin with scalloped dentinoenamel junction and areas of globular mineralization. Few cores of connective tissues were observed within the dentin (Fig. 3a and b). Final histopathologic report of excised tissue was consistent with incisional diagnosis of CGCOT with odontome on left side and compound composite odontome only on the right side of mandible. Patient was kept on a regular follow up and the 1-year follow up OPG revealed a normal bony architecture on both sides of the mandible (Fig. 4). A clinical follow-up of 2 years showed no evidence of recurrence on either side of the mandible.

2. Discussion

Gorlin and colleagues identified CGCOT as distinct pathological entity in 1962 although according to Altini and Farman, the condition had previously been described in German literature in 1932 by Rywkind. The CGCOT has also been reported under a variety of other designations (Table 1) and was earlier thought to be an oral presentation of dermal calcifying epithelioma of Malherbe [5]. Calcifying ghost cell odontogenic tumors constitute about 0.37–2% [3] of all odontogenic tumors and are seen with an almost equal frequency in the maxilla and mandible, with 65% cases occurring in the anterior jaw [2]. In 1992, the World Health Organization (WHO) classified CGCOT as a neoplasm rather than a cyst but confirmed that most cases were non-neoplastic [9,10]. In view of this duality, many different terminologies have been applied to cystic and solid CGOCT variants, but calcifying odontogenic cyst or tumor is the preferred term [7]. Tomich reviewed odontogenic tumors and cyst at the Indiana University School of Dentistry for about 34 years and found that only 51 cases of calcifying ghost cell odontogenic cyst were diagnosed, averaging less than two cases per year. It follows that the average oral and maxillofacial surgeon is likely to see only a case or two during his/her professional career [8].

Odontomes constitute 4–67% of all odontogenic tumors. Compound odontomes most commonly involve the anterior maxilla, whereas complex odontomes most commonly involve the posterior mandible [2]. Both lesions are made up of enamel matrix, dentin, cementum and dental pulp surrounded by a dental follicle or cyst. Compound odontomes appear as a collection of small teeth, leaving only a few entities in its radiographic differential diagnosis, unlike complex odontomes, which appear as radiodense masses of hard tissue that may result in a broader differential diagnosis. Both lesions have radiolucent rims, representing dental follicular tissue or, less commonly, a cyst. Since odontomes are well-encapsulated lesions and have less chances of recurrence, the management consists of a conservative surgical excision [11–13].

CGCOT may occur in association with other odontogenic tumors, the most common of these is the odontome. Buchner [14] showed this association in 35% of his cases, Nagao [15] et al. in 22% and Shamaskin [16] et al. in 47% cases. Hirshberg [17] revealed 52 cases of CGCOT associated with an odontome, analyzed its clinical and histological features, and classified it as an odontocalcifying odontogenic cyst. Abrar and Howell stated that the combined occurrence of CGCOT with another odontogenic tumor could be expected because of multi-potentiality of the odontogenic epithelium [6]. Surgical enucleation for such lesions is the treatment of choice with a recurrence rate of 3–5%. Several rare examples of malignant transformation of these lesions into ghost cell odontogenic carcinoma have also been documented [7]. Shear has stated that it is an intriguing question whether those CGCOTs, which also have features of other odontogenic tumors, develop in them secondarily or whether the CGCOT are themselves a secondary phenomenon in the pre-existing odontogenic tumor [6]. The concept of secondary development of CGCOT from pre-existing odontogenic tumors was

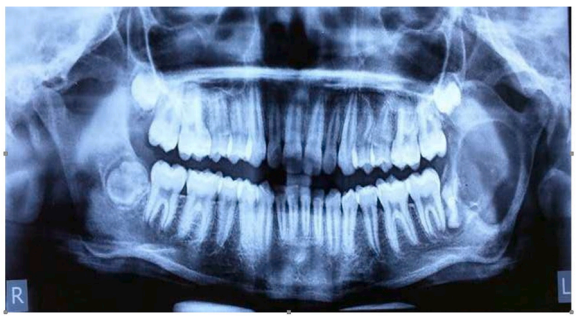

Fig. 1. -Panoramic radiograph showing a 25 × 20 mm unilocular radiolucent area distal to the left 2nd permanent molar extending up to the ramus of mandible and a calcified mass adjoining the right 2nd permanent molar with a radiolucent halo.

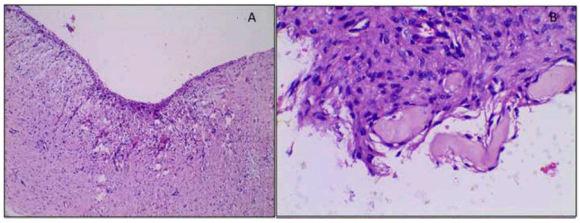

Fig. 2. a - Shows cystic wall lined by 2–3 cell thick straitified squamous epithelium. b - Shows globules of ghost cells within the lining of odontogenic epithelium.

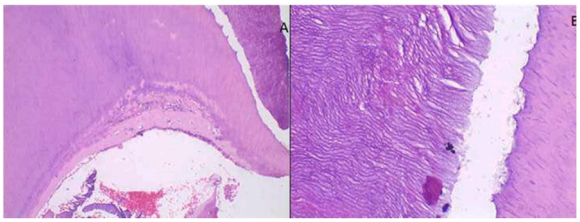

Fig. 3. a,b - Histopathology of both odontomes showing a similar presentation with the enamel matrix lined by ameloblasts, tubular dentin and a scalloped dentinoenamel junction.

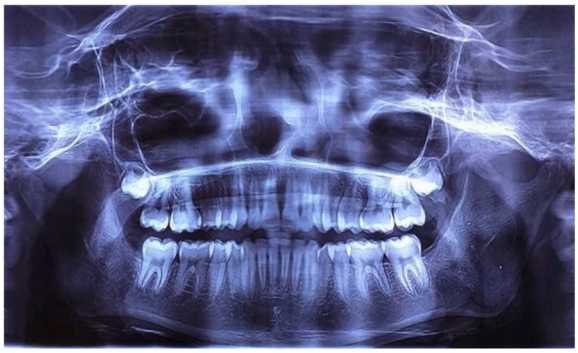

Fig. 4. 1-year post-operative panoramic radiograph showing normal bony architecture on both sides of mandible.

Table 1
Terminology of the calcifying odontogenic cyst.

Gorlin et al., 1962	Calcifying odontogenic cyst
Gold 1963	Keratinizing calcifying odontogenic cyst (KCOC)
Fejerskov and Krogh 1972	Calcifying ghost cell odontogenic tumor (CGCOT)
Freedman et al., 1975	Cystic calcifying odontogenic tumor (COCT)
Praetorius et al., 1981	Dentinogenic ghost cell tumor (DGCT)
Ellis and Shmookler 1986	Epithelial odontogenic ghost cell tumor (EOGCT)
Colmenero et al., 1990	Odontogenic ghost cell tumor (OGCT)

supported by many authors [18]. However, the present case of bilateral odontomes in the posterior mandible is rarely documented in literature.

The central CGCOT (intraosseous) presents as an asymptomatic hard swelling of the jaw that produces expansion, rather than erosion of bone. Pain, if present, suggests a secondary infection [3]. The clinical features in our case were similar to those described by other authors for such lesions. The lesion on the right side of mandible was discovered as an incidental radiographic finding.

Radiographically, early lesions of CGCOT appear completely radiolucent. As they mature, they develop calcifications that produce a well-circumscribed, mixed radiolucent-radiopaque appearance. Patterns of radiopacity range from salt and pepper pattern or flecks to a fluffy cloud-like pattern throughout or even a crescent-shaped pattern on one side of the radiolucency in a “new moon”-like configuration [19].

This case with simultaneous occurrence of a CGCOT with odontome on one left side of the jaw and a compound composite

odontome on the right side in a 12-year-old young child supports the concept of development of CGCOT secondary to an odontoma, as the authors believe that the differentiation of epithelium producing odontome to CGCOT has not yet occurred on the right side. The definitive diagnosis of CGCOT is made histologically, due to lack of the lesion's characteristic clinical and radiological features, as well as its variable biological behavior. The treatment of this lesion involves enucleation with a long-term follow-up, which was done in this case. Prognosis is good for cystic CGCOTs and less certain for neoplastic CGCOTs [20]. The CGCOT may be associated with other odontogenic tumors such as adenomatoid odontogenic tumor, ameloblastic fibroodontoma, ameloblastic fibroma and ameloblastoma, where the treatment and prognosis in such cases is based on the associated tumors [21]. Hence, the authors encourage early management of simple and common lesions developing from the odontogenic epithelium like the odontome to prevent possible secondary development of complex and rare lesions like CGCOT with a malignant potential.

Conflicts of interest

None.

Funding

None.

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