Calcifying Odontogenic Cyst With Compound Odontoma Of Maxilla Report Of A Rare Case With Radiological And Histopathological Correlation

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Abstract

Calcifying Epithelial Odontogenic Cyst (CEOC), also known as the Gorlin cyst, is a rare, benign odontogenic lesion of the jaws that displays both cystic and neoplastic features. It typically presents in the second to fourth decades of life and is more commonly found in the mandible. This article reports an unusual case of CEOC in a 14-year-old male, presenting as a painless swelling in the anterior maxillary region. The lesion was associated with an unerupted tooth and showed characteristic radiographic features, including a well-circumscribed radiolucency with internal calcifications. Despite the lesion's size and potential to cause facial asymmetry, the patient reported no symptoms. This case emphasizes the importance of including CEOC in the differential diagnosis of mixed radiolucent-radiopaque lesions in paediatric patients and highlights the need for timely radiographic and histopathological evaluation for effective management.

Introduction:

The Gorlin cyst, sometimes called calcifying epithelial odontogenic cyst (CEOC), is a rare benign odontogenic disease that exhibits both cystic and neoplastic features. Although maxillary involvement is also observed, it slightly prefers the front parts of the jaws, especially the mandible, and makes up 1% to 2% of all odontogenic tumors and cysts.[1] CEOC has no discernible gender preference and can develop at any age, but it is more frequently observed in the second to fourth decades of life. [2]

Clinically, CEOC frequently manifests as a painless, slowly expanding swelling that could be connected to an unerupted tooth. Until it results in facial asymmetry or cortical enlargement, the lesion is typically asymptomatic. [3] Due to dystrophic calcifications or "ghost cells," CEOC usually manifests radiographically as a unilocular or multilocular radiolucency with varying degrees of internal

radiopacities. However, these characteristics require histological confirmation because they are not pathognomonic. [4]

Ameloblastoma like epithelial lining, eosinophilic "ghost cells," and varying calcification within the cystic epithelium or lumen are histological characteristics of CEOC. Ghost cells are modified epithelial cells that have lost their nuclei but still maintain their essential cellular outlines. [5] To learn more about CEOC's biological behaviour, immunohistochemical markers like CK19, β-catenin, and Ki-67 should be investigated. [6]

Despite being seen as innocuous, certain variations of CEOC behave aggressively locally. [7] The cornerstone of treatment is surgical enucleation. Recurrence is rare but has been seen in situations of inadequate removal. [8] To avoid possible problems including bone loss or the displacement of nearby structures, early diagnosis and suitable surgical care are crucial.

Case Report:

A 14 year old male patient reported to the department of oral medicine and radiology with the chief complaint of swelling in the upper front and left back tooth region for 3 years, which was initially smaller in size and gradually attained to present size during last three years. The patient did not experience any discomfort or bleeding from the same. In the past, there was no history of trauma, fever, loss of appetite, or weight reduction of the patient. Overall, the patient remained healthy. On general physical examination patient was moderately built, nourished and the vital signs were within the normal range. There were no clinical signs of anaemia, Jaundice, cyanosis etc were detected in the patient. On clinical examination, permanent left maxillary canine and second premolars were missing. Retained deciduous left maxillary canine and second molars were present. The examination of specific lesion revealed a solitary swelling occupying left maxilla, measuring approximately 5x1.5 cm extending anteroposteriorly from frenal attachment to mesial aspect of 26 with well-defined borders with no secondary surface changes. The lesion was non tender and hard in consistency (Fig 1). Based on the history and clinical examination, provisional diagnosis of Adenomatoid odontogenic tumor was considered.

Patient was then subjected for radiological examination. Panoramic radiograph (OPG) revealed a solitary mixed radiolucent and radiopaque areas with well defined corticated borders and impacted teeth, extending from the distal aspect of 22 to mesial aspect of 26. The lesional area was consisting multiple radioopacities along with two impacted teeth and resulted in the displacement of adjacent teeth away from the epicentre (Fig 2). Cone beam computed tomography (CBCT) revealed expansion of buccal and palatal cortical plates with impacted 23, 25 and root resorption of 63, 24, 65 without Maxillary sinus involvement (Fig 3,4,5,6).

Figure 1: The swelling involving the left maxillary arch

Figure 2: Panoramic image showing calcifications, impacted teeth within the lesion along with mesial displacement of permanent left lateral incisor.

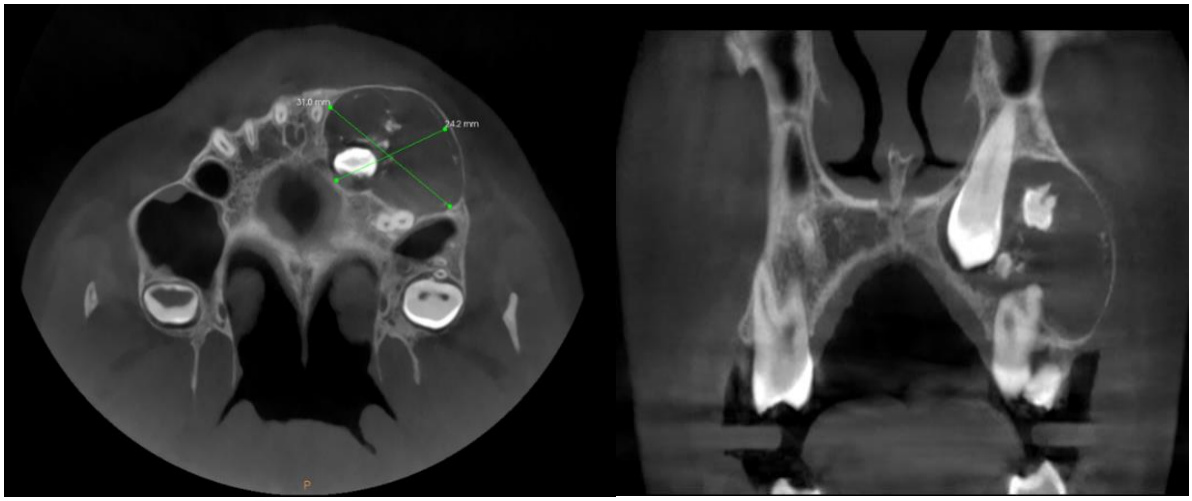

Figure 3: Axial CBCT reveals involvement of Buccal and Palatal cortical plates

Figure 4: Coronal CBCT reveals impacted canine displacement towards nasal fossa

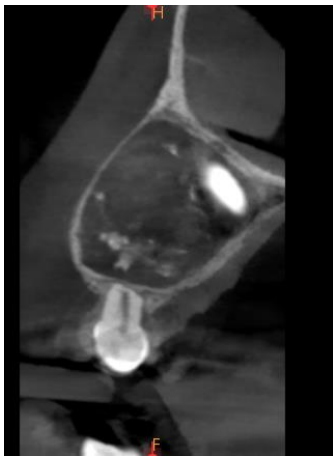

Figure 5(a)

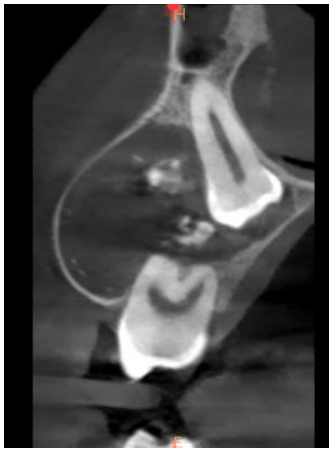

Figure 5(b)

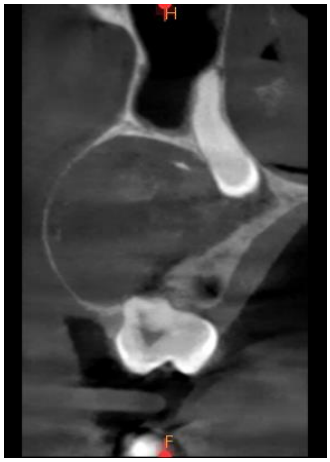

Figure 5(c)

Evidence of root resorption 5(a) deciduous canine (b)permanent left premolar (c) deciduous molar

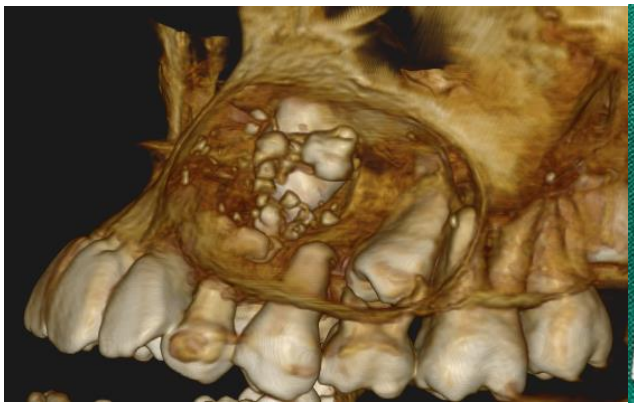

Figure 6: 3D CBCT reveals calcifications and impacted teeth

Figure 7: Excised specimen

Following the Haematological evaluation and Radiological investigation; surgical excision of the swelling was done under local anaesthesia. The excised sample was sent for the histopathologic examination (Fig 7)

Prominent aggregates of Ghost cells are seen with foci of dentinoid with lamellar calcifications arranged in concentric patterns (Fig 8)

Discussion:

Calcifying odontogenic cyst is a rare benign odontogenic lesion, which is often referred to as the Gorlin cyst as Gorlin et al. were the first to report it in 1962[9]. Over time, the World Health Organization (WHO) reclassified COC, and when it exhibits solid, neoplastic features, it is now known as the Calcifying Cystic Odontogenic Tumor (CCOT).[10]

Usually COC can arise as a single lesion or in conjunction with other odontogenic tumors. [11]In these situations, the cyst may enclose or be close to the odontoma, and a sizable percentage of lesions are discovered by chance on radiographs done for swelling or delayed tooth eruption. [12] COC is commonly detected in the anterior jaw region and is typically asymptomatic and slow-growing.[13] It can present across a broad age range, but most frequently affects

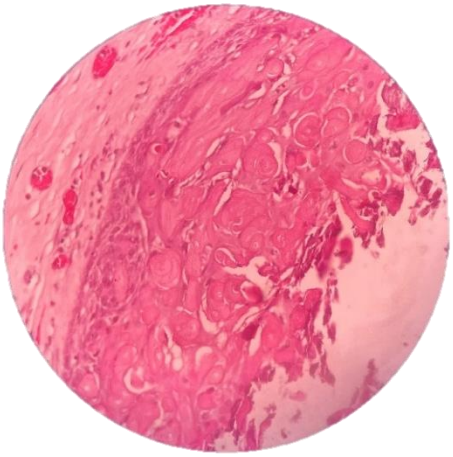

Figure 8(a)

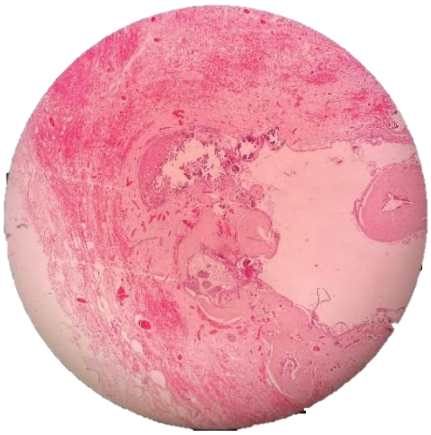

Figure 8(b)

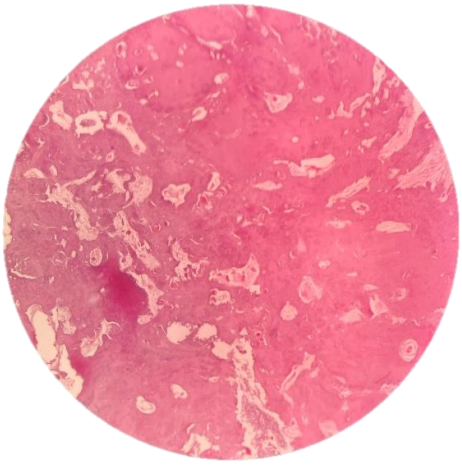

Figure 8(c)

Figure 8(a), 8(b), 8(c) histological pictures of the lesion

individuals in the second to third decades of life, with no significant sex predilection.[14]

On the radiograph, the lesion usually appears as a well-defined unilocular or multilocular radiolucency, frequently with dispersed radiopaque foci from calcified ghost cells or an accompanying odontoma. Internal radioopacities aid in differentiating COC from other radiolucent lesions of the jaw.[15] Odontomas might show up as distinct radiopaque masses inside the radiolucent cystic structure in mixed lesions, making the radiographic diagnosis more difficult and requiring histological confirmation.[16]

Dentigerous cyst, adenomatoid odontogenic tumor (AOT), ameloblastic fibro-odontoma, and calcifying epithelial odontogenic tumor (CEOT) are among the differential diagnoses for COC. They all occur in similar age groups and sites and may exhibit radiopaque components. Ghost cells, which are anucleate epithelial cells that have the potential to calcify, are a characteristic that is pathognomonic for COC.[17,18]

According to histology, COC is distinguished by an odontogenic epithelial lining that resembles that of ameloblastoma and contains ghost cells that are susceptible to dystrophic calcification. Mature dental hard tissues such as cementum, dentin, and enamel matrix are frequently seen inside the lesion in odontoma instances.[19] In certain situations, ghost cells may cause a foreign body giant cell reaction.[5]

The lesion is normally treated with conservative surgical enucleation, and the biologic nature of COC is generally non-aggressive. Prognosis is generally favorable, with low recurrence rates if completely excised.[20]However, long-term follow-up is advised, especially in cases

associated with odontomas or where the lesion exhibits aggressive features.

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