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Dentinogenic Ghost Cell Tumor - A Rare Neoplastic Form of Calcifying Odontogenic Cyst

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Abstract

Introduction: Dentinogenic ghost cell tumors (DGCT) are very rare tumors considered as solid variants of calcifying epithelial odontogenic cysts (CEOC). They are locally invasive neoplasms and occur as two forms; intra osseous (central) and extra osseous (peripheral).

Methodology: A 14 year old patient presented with a chief complaint of well-circumscribed swelling in the mandibular right lateral incisor region with a diameter of 11x11 mm since 1 year. There was no history of pain associated with the swelling. Medical and family history was not relevant. After SRP, an excisional biopsy was performed. The lesion grew back in the size of 9x12 mm within one month. A deeper excision was performed and the tissue was send for histopathological examination.

Results: The lesion showed odontogenic epithelium, ghost cells, dentinoid material, and giant cells. The final microscopic diagnosis was a Dentinogenic Ghost Cell Tumor.

Conclusion: DGCT is an extremely rare tumor. The peripheral, extra osseous lesion can be easily confused with other gingival lesions such as reactive or inflammatory lesions or other peripheral odontogenic tumors. The clinical appearance of all of these lesions is similar; therefore a definitive diagnosis and a regular follow-up are imperative.

Keywords: CEOT, DGCT, Intraosseous, Extraosseous

Introduction

Calcifying odontogenic cyst (COC) was recognized as a distinct clinicopathological entity by Gorlin et al in 1962.[1] The solid variant of COC was called calcifying ghost cell odontogenic tumor. Even though it has the features of a cyst, it also has several prominent characteristics of a solid neoplasm. Hence it was renamed as Dentinogenic ghost cell tumor (DGCT) by Praetorius et al. [2] The WHO defined DGCT as a locally invasive neoplasm characterized by ameloblastoma-like islands of epithelial cells in a mature connective tissue stroma. Aberrant keratinization may be found in the form of ghost cells in association with varying amounts of dysplastic dentin. [3] DGCT is usually considered to be a rare condition. DGCT can exhibit either a benign or a malignant form or can undergo malignant transformation.[4]

The aim of this paper is to report a case of DGCT and briefly review the scientific literature about it.

Case Report

A 14 year old male patient visited our department with a chief complaint of swelling with respect to lower right

front tooth region. Patient gives no history of pain or extra oral swelling. The patient reported the onset of lesion one year ago. (Figure 1&2).

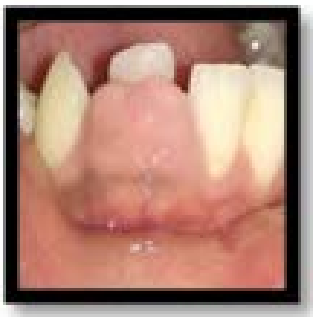
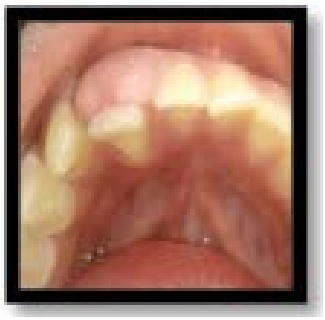

Fig 1 & 2 Localized swelling of 11X11mm size

Clinically intra oral examination revealed a pale pink, hard, non tender sessile swelling of size 11 X 11mm with respect to marginal gingiva of 42. No sign of pus discharge or mobility was associated with the tooth. Radiographic examination revealed no bony involvement. (Figure: 3) Soft tissue shadow was seen.

Fig 3: Radiographic View

All the hematological parameters were within normal range. Scaling and Root Planing was done. After 2 weeks surgical excision of the lesion (fig:4, 5) was done and the specimen (fig 6,7) was sent for a histopathological examination.

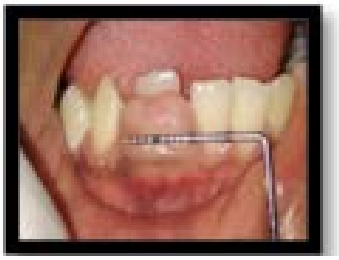
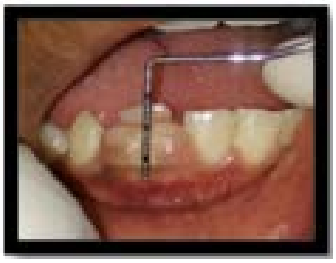

Fig 4, 5:Pre-operative view

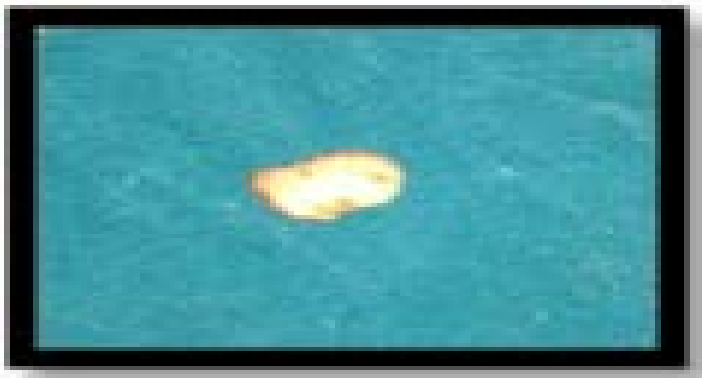

Fig: 6 Excised Tissue

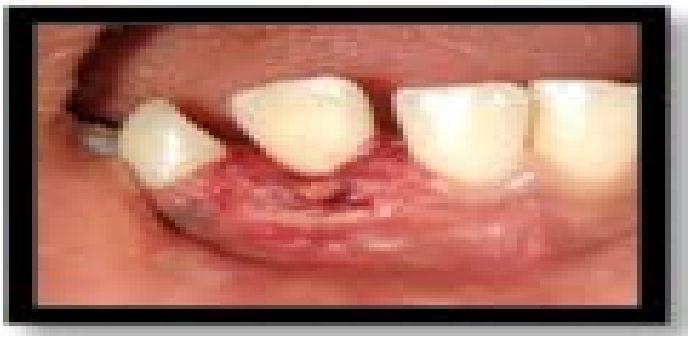

Fig: 7 Post-operative view

Histopathological report (fig: 8) reveals parakeratinized stratified squamous epithelium. Deeper connective tissue reveals presence of two teeth like structures composed of dentine/dentinoid material. Numerous ghost cells undergoing calcification can be seen.

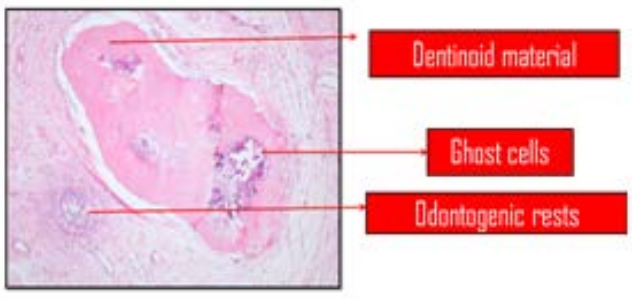

Fig: 8 Histopathological view

Features were suggestive of peripheral developing Odontome but a deeper excisional biopsy was advised as the tumor component was located close to edge of the biopsy and it looked like complete excision was not done.

During the follow up (fig: 9,10,11,12), it was seen that the lesion was recurring back and by 4th week it had grown back to its almost original size (9X12mm).

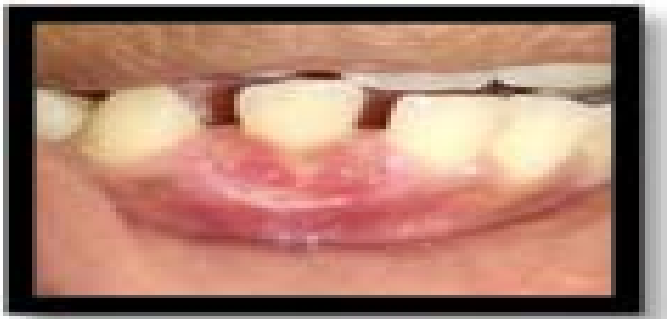

Fig: 9- Post operative view after 1st week

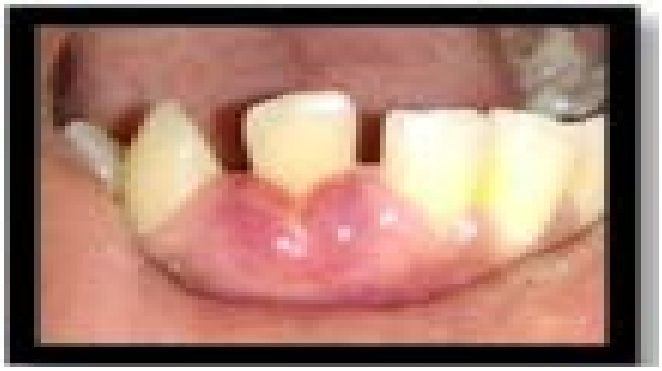

Fig: 10- Post operative view after 2nd week

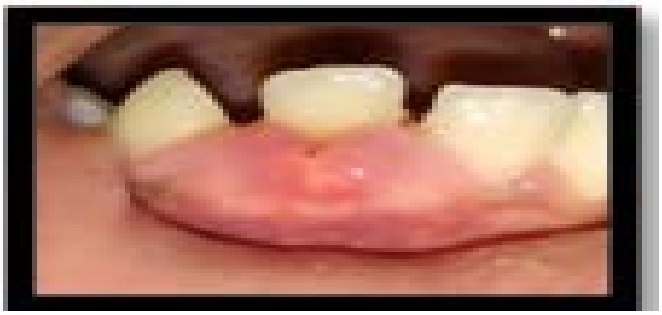

Fig: 11- Post operative view after 3rd week

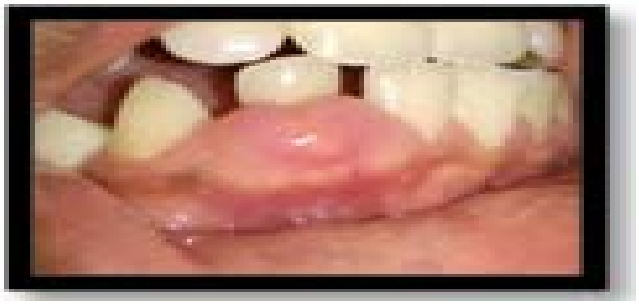

Fig: 12- Post operative view after 4th week

So, a second biopsy was planned. (Fig: 13, 14) A deeper excision was done followed by curettage of the involved area. (Fig: 15, 16, 17) The obtained specimen was sent for histopathological examination.

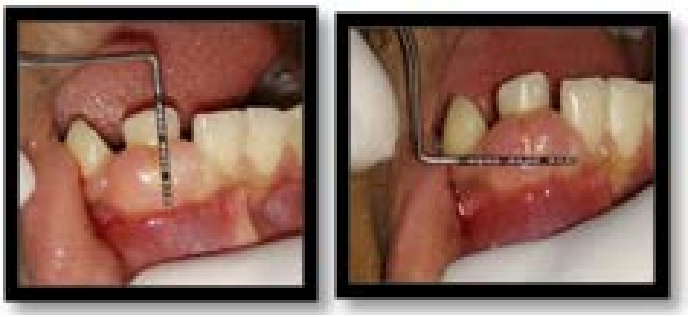

Fig: 13, 14- Swelling of 9X 12mm

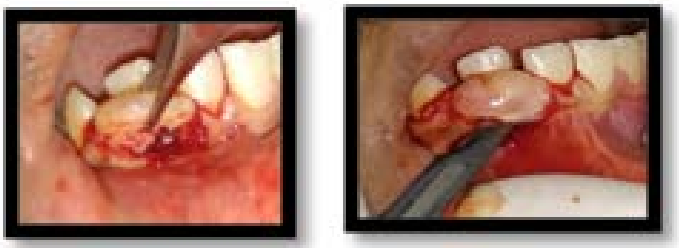

Fig: 15-Excision of lesion

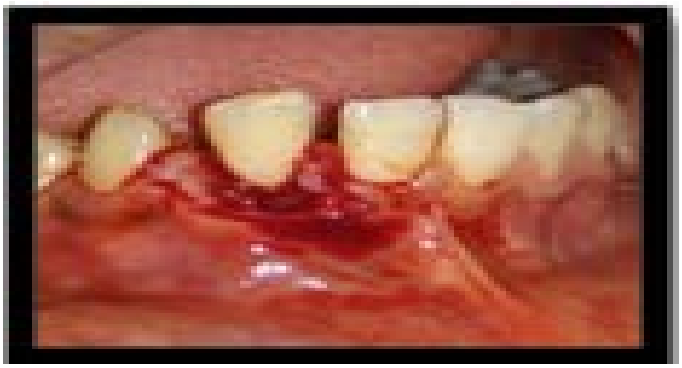

Fig:16 After excision

Fig: 17-Sutures

The histopathological report (fig: 18) revealed sheets and islands of proliferating odontogenic epithelial cells in connective tissue. Eosinophilic anucleate, swollen, spherical to ellipsoidal structures were suggestive of ghost cells. Masses of dentinoid with basophilic globules of calcification admixed with odontogenic epithelial cells. Features were suggestive of Dentinogenic ghost cell tumor.

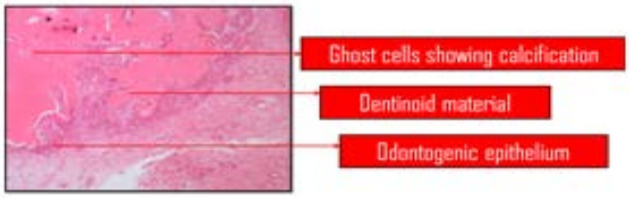

Fig: 18 Histopathological view

Post operative pictures: (Fig 19-23)

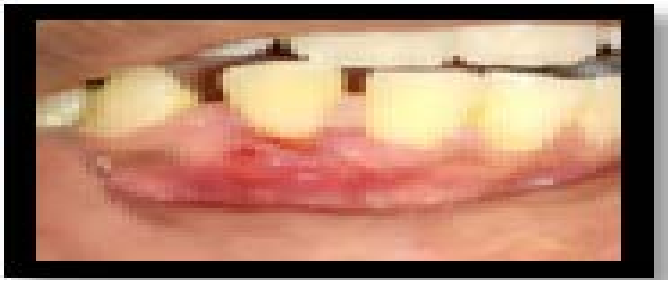

Fig: 19-Post operative 2nd week

Fig: 20- Post operative 1st week

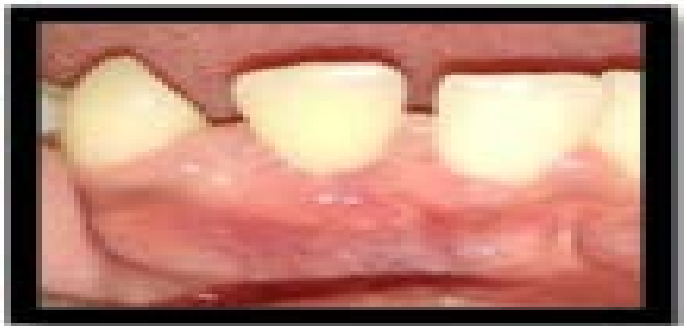

Fig 21- Post operative 3rd week

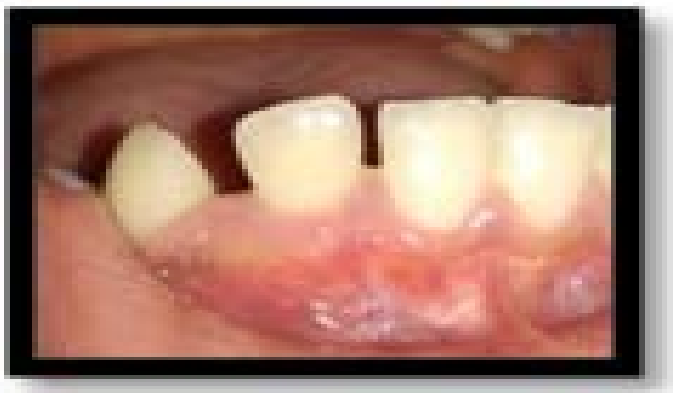

Fig 22- Post operative 3 months

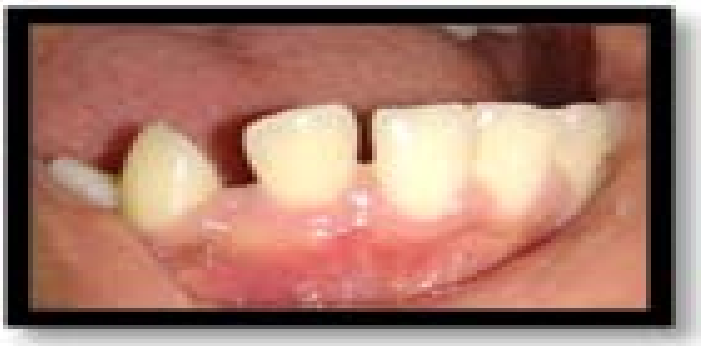

Fig 23- Post operative 6 months

Discussion

The first ever description of dentinogenic ghost cell tumor (DGCT) was given by Fejerskov and Krogh[5] in 1972. They used the term “calcifying ghost cell odontogenic tumor.” In 1981, Praetorius et al.[2] suggested the term “dentinogenic ghost cell tumor.” The term DGCT was used by the authors because dentinoid formations were seen in relation to the epithelial islands and Ghost cells of varying degrees were also found. In 1983, Shear[6] used term “dentinoameloblastoma” because of its similarities with ameloblastoma and dentinoid production. In 1986, Ellis and Shmookler[4] used the term “epithelial odontogenic ghost cell tumor” since they thought that the ghost nucleated keratinizing cell was the most distinctive histopathological feature. In 1991, Hong et al.[7] supported the use of term “epithelial odontogenic ghost cell tumor” as characteristic of these neoplasm are the odontogenic epithelial proliferations with some inductive activity and

the formation of ghost cells. The authors did not use the term 'dentinogenic' as they believed that dentinogenic seemed to connote a mesenchymal tissue origin and production of true dentin. Later in 2003, Li and Yu[8] suggested the term "odontogenic ghost cell tumor". This term emphasized its origin, neoplastic nature, and most striking histopathological features.

Over the years several terminologies have been used to designate this rare variant of calcifying odontogenic cyst (now termed as calcifying cystic odontogenic tumor [CCOT]), until recently when in 2005, the World Health Organization (WHO)8 decided to retain the term dentinogenic ghost cell tumor as initially described by Praetorius et al.[2]

DGCT represents approximately 1.9% to 2.1% of the overall odontogenic tumors. [9] Two variants of DGCT have been described: a locally invasive intraosseous tumor (central) and a noninvasive extraosseous tumor (peripheral). Peripheral DGCT is a rare odontogenic tumor; representing 13% to 21% of all DGCTs.[10, 11]

Candido et al., [12] in their review of the literature, concluded that peripheral DGCTs mostly affect the canine region or the anterior part of the jaw. The patient age ranged from 41 to 83 years with an average age of 62.

Histologically, DGCT is an infiltrative solid neoplasm which is composed of odontogenic epithelium associated with ghost cell formation and production of dentinoid. Dentinoid is hyalinized eosinophilic material suggestive of immature or dysplastic dentin. It is located closely near the epithelial sheet. A characteristic feature of DGCT is ghost cells. Individual as well as large islands of ghost cells may be seen. Some ghost cells undergo calcification and lose their cellular outline. Ghost cells are swollen ellipsoidal keratinized epithelial cells that have lost their nuclei. Ghost cells are essentially requisite for the diagnoses of DGCT and COC. Large islands or individual eosinophilic ghost cells are found in the epithelium as well as in the connective tissue.[13]

Ledesma-Montes et al[14] who described the two variants of DGCT reported that peripheral occurrence of DGCT is rare and only few reports with clinical radiographic documentations are available. This is in consensus with Bello et al[15] who reported that only 24 cases were stated in the literature prior to their case in 2011.

Differential diagnosis includes ameloblastoma, odonto-ameloblastoma, ameloblastic fibro-odontoma, odontoma, adenomatoid odontogenic tumor, and cementoma. The presence of ghost cells and dentinoid material may distinguish DGCT from other odontogenic tumors, such as peripheral ameloblastoma and peripheral calcifying epithelial odontogenic tumor (CEOT).[12]

De Arruda[16] et al did a systematic review study on COC, DGCT and GCOC. From their study it was concluded that reports on DGCT was scarce and only 55 cases were reported so far. The age distribution of DGCT patients ranged from four months to 89 years. Of 55 cases, 36 were reported in males and 19 in females (male-to female ratio: 1.9:1). The lesions most often occurred in the mandible. 11 cases were in the posterior mandible and 9 in the anterior mandible. In maxilla, 8 cases were in the anterior maxilla and 4 in the posterior maxilla. The maxillary sinus was affected in three reports. The most common sign was swelling. 15 DGCTs were peripheral. In 39 cases, conservative surgery was performed. In 32, no recurrence was reported. Of the 11 cases in which recurrence was reported, the period of lesion recurrence ranged from two weeks to six years after treatment.

The peripheral extra osseous lesion can be easily confused with other gingival lesions such as reactive or inflammatory lesions or other peripheral odontogenic tumors. The clinical appearance of all of these lesions is similar; therefore, the definitive diagnosis depends on

histology and biopsy with a mandatory microscopic examination. Early diagnosis of DGCT is essential for better prognosis of the patient. Dentinogenic ghost cell tumor can be either benign or malignant, depending on the histopathological features. Malignant DGCTs can show aggressive clinical behavior and can metastasize.[13]

In our case; the patient was treated by surgical excision followed by curettage and is under follow up. No recurrence of the tumor has been observed till date and the patient was asymptomatic.

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