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Case Report

A Rare Case of Central Dentinogenic Ghost Cell Tumor: Case Report and Review of Literature

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ABSTRACT

Dentinogenic Ghost Cell Tumor (DGCT) is the solid neoplastic variant of the calcifying odontogenic cyst (COC).DGCT is extremely rare in occurrence, accounting for 2 to 14 % of all COC and only 46 cases intraosseous DGCT are reported in literature till date to the best of our knowledge Hence A series of collaborative studies is being conducted to identify and integrate the clinical and radiological features of DGCT. Here through this case report, we add the 47 th case of central DGCT involving the left maxilla in a young female patient to the literature.

Keywords: Dentinogenic ghost cell tumor, Ghost cell tumor, Calcifying odontogenic cyst, Ghost cell calcifying odontogenic tumor, Dentinoid, Central odontogenic ghost cell tumor.

INTRODUCTION AND REVIEW OF LITERATURE

Dentinogenic Ghost Cell Tumor (DGCT) is the solid neoplastic variant of the calcifying odontogenic cyst (COC). COC was first described and reported by Gorlinet al in 1962 [1] and accounts for 1 to 2 % of all odontogenic tumors; It is a heterogenous lesion and exist both as the cystic and solid variant; hence its nomenclature has been updated multiple times and has been controversial in history. In 2005 WHO renamed cystic variety of the COC as calcifying cystic odontogenic tumor (CCOT) and solid variant as Dentinogenic Ghost Cell Tumor(DGCT. [2] Later in 2009 international collaborative group [3] reviewed the WHO's classification and added malignant variety to the classification and named it as ghost cell odontogenic carcinoma (GCOC). DGCT is extremely rare in occurrence, accounting for 2 to 14 % of all COC. [4] A study conducted in China [5] showed that DGCT comprised only 0.5% of 1,642 odontogenic tumors studied; in a similar study in Sri Lanka count was still less i.e.0.3% of 1,677 odontogenic tumors studied. [6] In 2008, Constantino Ledesma-Montes did an International collaborative study of 114 cases of ghost cell odontogenic tumors by reviewing the data from 14 departments of pathology and oral pathology from Mexico, South Africa, Denmark, The United States, Brazil, Guatemala, and Peru and found that only 5 cases (4.4%) of central DGCT were recorded in comparison of 109 cases (95.6%) of central CCOT. [7] In a study from Iran, only 1 case (3.1%) of DGCT was found among 32 cases of central ghost cell odontogenic tumors. [8] Hence it is the established fact that DGCT is a very rare tumor not only among odontogenictumors, but also compared with CCOT. [7,8] Amos

Buchnerin 2016 did the study to integrate the available data published in single case reports on central DGCT into a comprehensive analysis of its clinical and radiologic features and could find only 45 well documented cases of central DGCT in literature from 1972 to 2014. [9] Following this study a single case of central DGCT was reported in the literature by Rai S in 2015 as we searched the PUBMED MEDLINE and Google. [10] Here we report a 47th case of central dentinogenic ghost cell tumor in English literature. The lesion was identified in a 17 year old female patient involving the left maxilla and was treated with enucleation and curettage and did not show any signs of recurrence during the follow up period of 1 and half years. Furthermore, there is little precedent in the literature to guide management in such a case, and we therefore consider this report to be noteworthy and instructive in this respect.

CASE REPORT

A 17 year old female patient reported to us with the chief complaint of dull continuous pain accompanied with swelling on the left side of face since 1 month with temporary relief of symptoms by taking analgesics (Diclofenac Sodium 50 mg plus Paracetamol 375 mg). Swelling gradually increased in size to the size present on examination. No relevant medical history or dental history present.

On examination diffuse soft, mildly tender swelling was seen with normal overlying skin present at the left ala of the nose, around 2 cm in greatest dimension obliterating the left nasolabial fold and causing slight facial asymmetry.

On intraoral examination diffuse expansile firm tender palatal swelling was present extending from the midpalatine region up to the palatal gingiva in left maxillary canine, first premolar and the second premolar region; buccal cortical plate expansion seen obliterating the buccal vestibule in left maxillary first premolar, second premolar and first molar region; pain on percussion with respect to second premolar and first molar, no mobility with teeth seen. Left maxillary second molar and third molar could not be seen clinically (Figure 1).

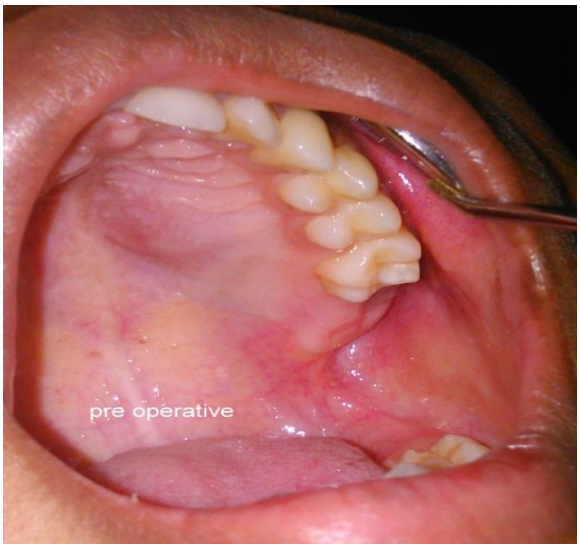


Figure 1: Intraoral photograph showing palatal swelling seen extending in left maxillary canine, first premolar and second premolar region.

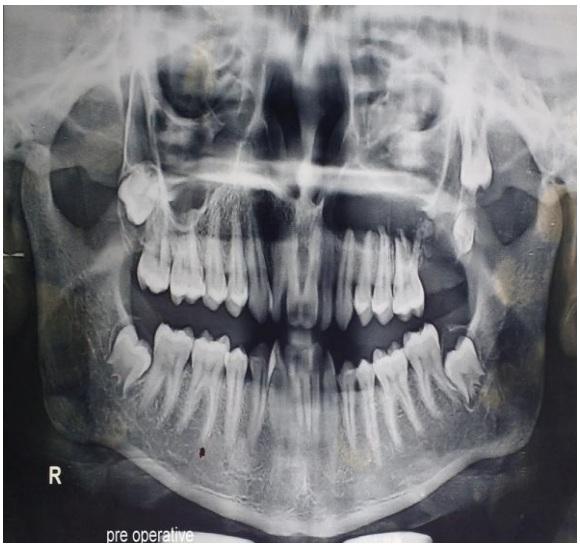


Figure 2: Preoperative Orthopantograph showing well defined unilocular radiolucent lesion with left maxilla, flecks of calcification distal to left maxillary first molar and left maxillary second and third molar pushed upwards close to the lateral wall of orbit.

Fine Needle Aspiration Cytology (FNAC) test showed moderately cellular smear consisting of lymphocytes, macrophages in the background of thin proteineceous fluid giving the impression of the benign cystic lesion.

Clinical provisional diagnosis of cystic lesion involving the left maxilla was

made and dentigerous cyst was considered as the most apt differential diagnosis.

On radiographic examination; Lateralocclusal radiograph showed a radiolucent lesion with flecks of radioopacities encroaching into the left maxillary sinus cavity, apical root resorption with respect to left maxillary canine, first premolar and second premolar region; left maxillary second molar and third molar were seen to be impacted.

Orthopantograph showed well defined radiolucency with partially corticated borders in the periapical region of in left maxillary canine, first premolar, second premolar and the first molar region extending superiorly into the left maxillary sinus with loss of normal architecture lower border and lateral border of the left maxillary sinus not traceable. . Left maxillary second molar and third molar lying laterally close to the left orbit; small flecks of calcification seen lateral to the distobuccal root of left maxillary second molar. Apical root resorption with respect to left maxillary canine, first premolar and the second premolar was seen (Figure 2).

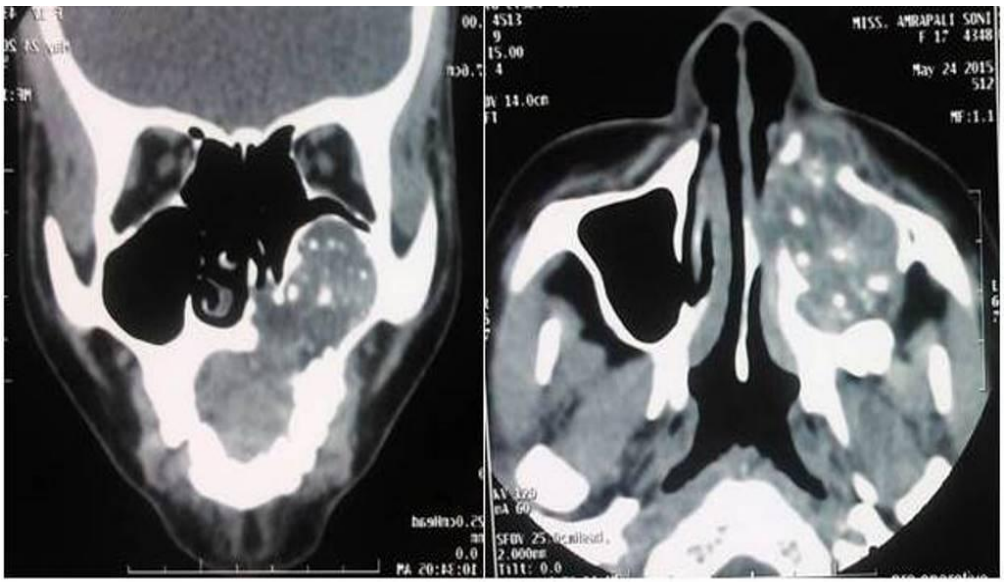
Water's view showed opacification of the left maxillary sinus.

CT scan showed well corticated destructive lesion with flecks of calcification dispersed inside, seen encroaching into the maxillary sinus causing haziness of the left maxillary sinus with the borders of the lesion in close proximity to the borders of the maxillary sinus. Perforation of the palatal cortical plate, posterior wall and anterior wall of the left maxillary sinus was seen (Figure3).

Final provisional diagnosis calcifying odontogenic cyst (COC) was given.

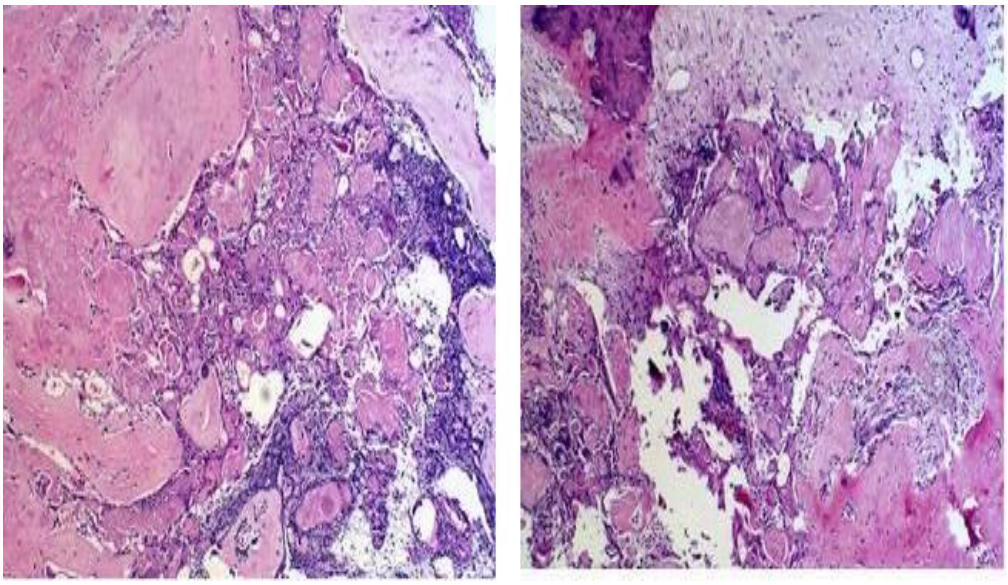
Surgical excision and curettage of the lesion were planned following endodontic treatment with left maxillary canine, first premolar and second premolar, first molar region. Histopathology of the excisional biopsy showed the presence of cystic epithelium composed of ameloblast, with abundant ghost cells and dentinoid material dispersed as shown in the (Figure4); suggesting of dentinogenic ghost cell tumor (DGCT) which is a solid variant of COC.

Patient was followed up for one and half years and patient showed no clinical or radiographic signs of recurrence of the lesion (Figure 5).



A **B**

Figure 3: Cone Beam Computed Tomography scan (A) Coronal section showing palatal cortical plate perforation (B) Axial section showing expansile destructive lesion with left maxillary sinus.



A **B**

Figure 4: (A) Hemotoxylin and Eosin (H&E) stained section showing ghost cells surrounded by odontogenic epithelium & stellate reticulum-like cells with areas of dentinoid formation. (H&E, 25X). (B) Hemotoxylin and Eosin (H&E) stained section showing diffuse area of dentinoid formation formation. (H&E, 100X).

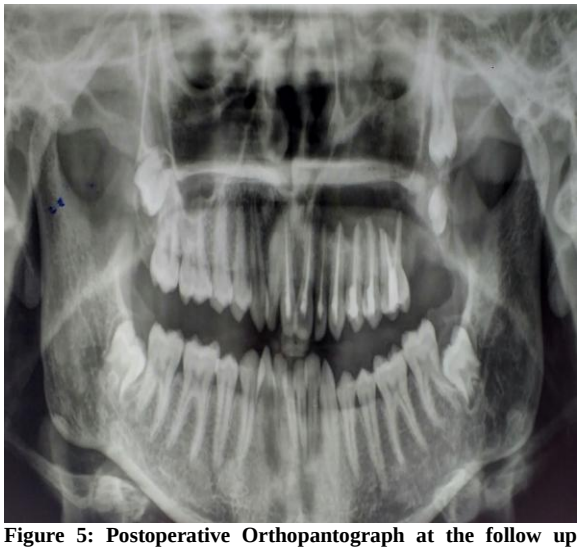


Figure 5: Postoperative Orthopantograph at the follow up period of one and half year; showing no signs of recurrence of lesion.

DISCUSSION

The WHO, in 2005, [2] 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".

Due to the scarcity of the literature on DGCT owing to its rare occurrence; various collaborative studies and comprehensive studies are being conducted worldwide to increase the understanding of the lesion among the medical professionals and make the diagnosis and treatment easier. [7,9]

In these studies [7,9] it is found that the highest prevalence of DGCT occurred from the third to fifth decades of life with the mean age of 40 years. The male-to-female ratio is 1.8:1. The most common location is the posterior mandible followed by the posterior maxilla. Expansion and perforation of the cortical plates, resorption of the roots is the most common clinical and radiographical findings associated with DGCT. Radiographically, the lesions were unilocular with mixed radiolucent and radiopaque appearance manifested by irregular scattered radiopacities. It occurs both as central type and peripheral type with the central type being more common. The transformation of the DGCT into malignancy is found to be extremely rare.

Presented case was identified in 17 year old female patient with a lesion in the posterior maxillashowingbuccal cortical plate expansion with unilocular well corticated mixed radiolucent radiopaque lesion encroaching into the left maxillary sinus causing palatal cortical plate perforation and apical root resorption. These features are consistent with findings of DGCT reported in literature.

Central DGCTs are aggressive neoplasms that show locally invasive behavior and recurrence rates of up to 73%. [9] As for the treatment of choice for DGCT, too few cases have been reported to predict prognosis based on any specific type of surgery. Also, follow-up time is limited in many cases. However a case series from china based on their observation and experience with treatment approaches for DGCT recommended treating DGCT with resection with marginal resection of at least0.5. [11]

The present case was treated with enucleation and curettage under general anesthesia and showed no signs of recurrence at the follow up period of 1 and half year.

CONCLUSION

DGCT is the rare odontogenic ghost cell tumor to come across in literature with many collaborative studies being conducted across the world on this lesion so as to identify and define the clinical and biological behavior of this rare lesion. This case report adds the 47 th case of DGCT to the literature.

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