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Dentinogenic ghost cell tumour of anterior mandible crossing the midline: Report of a rare case with a review of literature

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Abstract

Ghost cell revealing odontogenic tumours are very elusive and controversial and pathophysiology of these tumours are still debatable. Dentinogenic ghost cell tumor (DGCT) is a rare tumorous form of calcifying odontogenic cyst and only a small number of cases have been described. Dentinogenic ghost cell tumor (DGCT) is an exceptional odontogenic tumor accounting for only 2-14% of the COCs. Till today only less than 40 cases of DGCT have been reported in English literature. The main purpose of presenting this case is to present a rare case of DGCT in the anterior mandible of 30 yrs old female exhibiting curious histological behaviour, along with review of literature.

Keywords: Calcifying odontogenic cyst, dentinogenic ghost cell tumor, odontogenic ghost cell tumor, DGCT

Introduction

Calcifying odontogenic cyst was first reported by Gorlin *et al.* in 1962. DGCT is a very rare entity with controversies in its terminology and classification. It is the neoplastic solid counterpart of calcifying odontogenic cyst. Less than 40 cases have been reported so far in the literature ^[1].

Because of the confusing and controversial terminologies associated with classification of Calcifying odontogenic cyst it is very necessary to properly understand its clinical, radiographical and histological behavior. DGCT generally represents as a benign tumour, however, malignant transformation is also reported ^[1,2].

Case Report

A 30 year old female patient presented with a swelling in the anterior mandibular region since four and half months and pain since last one month. There was no history of trauma, swelling started insidiously and steadily increased in size. Medical, surgical, dental, family and personal histories were not relevant.

On extra oral examination confirmed a solitary diffuse swelling on the right mandibular canine area, extending half a centimeter below the vermilion border and a centimeter above the inferior mandibular border of the mandible. Submandibular, sublingual and cervical lymph nodes were palpable. No secondary changes such as, sinus or fistula and local rise of temperature.

Intraoral examination revealed a solitary diffuse swelling in the mandibular labial sulcus, extending from the 32 region and crossing the midline extending up to 45 region. Mucosa over the lesion was normal and hard in consistency. Both labial and lingual cortical plate expansion was observed, however, but lingual expansion was more in relation to 42, 43, and 44 (Fig-1). These clinical findings favored the differential diagnosis of included central giant cell granuloma, ameloblastoma, central ossifying fibroma, fibrous dysplasia, traumatic bone cyst and aneurysmal bone cyst.

Orthopantomography (OPG) confirmed a pear shaped well defined radiolucency, extending from the root of 32 up to the mesial margin of 45. Divergence of root of mandibular canine was also observed within the radiolucent areas and no evidence of any root resorption (Fig-2). Considering this radiographic differential diagnosis of ameloblastoma, traumatic bone cyst, aneurysmal bone cyst, central giant cell granuloma, calcifying epithelial odontogenic tumor and calcifying odontogenic cyst was made.

The lesion was surgically enucleated and subjected for histopathological examination. Macroscopic examination of the enucleated specimen was brownish black in color, soft to firm in consistency, with irregular superficial surface. Further, tissue was fixed in buffered formalin and processed electronically and stained with H& E Stain.

The H&E stained sections expressed (Fig 4.) cystic lining of odontogenic epithelium composed of ameloblast like cells and few stellate reticulum like cells along with mild proliferative features. Within the connective tissue capsule, abundance of dentinoid material admixed with odontogenic epithelium arranged in or ameloblastoma like follicles. Numerous ghost cells are also observed along with abundant dentinoid like material. These histological findings favored the diagnosis of dentinogenic ghost cell tumor. Further, special stains like; Van Gieson showed positive for ghost cell and negative for congo red in the dentinoid like areas ruled out the possibility of amyloid like materials in the tissue.

Review of Literature

First description of DGCT was given by Fejerskow & Krough in 1972 and terminology was first used by Pretorius *et al.* (1981). It accounts 2-14% of all COCs [2-6] Calcifying odontogenic cyst (COC) is a cystic lesion in which the epithelial lining shows a well defined basal layer of columnar cells, an overlying layer that may resemble stellate reticulum and masses of ghost epithelial cells either in the epithelial cyst lining or in the fibrous capsule.^{7,8} nomenclature and terminologies associated with calcifying odontogenic cyst is interesting and debatable because COC’s generally exhibits histological complexity and morphological diversity and accounts 2% of all odontogenic lesions [1, 9-11] thus, in 2005 WHO renamed the cystic version of COC as calcifying cystic odontogenic tumor (CCOT) and solid version of COC as Dentinogenic ghost cell tumor (DGCT) [3, 12-14]

Pretorius (2006) proposed a classification system for the odontogenic ghost cell lesions and included DGCT as group 3- Solid benign odontogenic neoplasm (DGCT) [2, 9] and suggested various criteria’s for this group of lesions

- A. May have an infiltrative growth pattern
- B. Predominantly solid lesion
- C. Combined with features of ameloblastoma, ghost cells and dentinoid [1, 10, 15]

Further, F. Praetorius and C. Ledesma- Montes (2006) defined this lesion 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 along

with varying amounts of dysplastic dentin.” The proliferative epithelium and ghost cells are interspersed with the abundant dentinoid [15, 16]

DGCT has been reported mainly occurring in the mandibular anterior region, with a male predilection (5:3), with in the age range of 40-50 years [17-19]. Radiography generally shows mixed radiolucency which may be uni/multi locular with expansion of cortical plates. Histopathology reveals proliferative epithelium and ghost cells interspersed with dentinoid and ameloblast like follicles in mature stroma. Ghost cell may infiltrate the stroma to form foreign body granulomas [18, 19] Surgery is the preferable choice of treatment and is performed by surgical resection which can be segmental or enblock. [15, 20]. Many cases have been treated with conservative enucleation procedures but high rate of recurrences have been reported after 5 -8 yrs [17, 19]

The term “odontogenic ghost cell carcinoma” [1, 10, 16] should be used to designate a malignant tumor with potential to metastasize. In odontogenic ghost cell ameloblastoma [1, 9], which are exceedingly rare, enamel deposition as well as dentin formation are also been reported in the literature.

Gunhan *et al.* prefer the term epithelial odontogenic ghost cell tumor [1, 4], because epithelial odontogenic ghost cells are the main component of this tumor. Scott and Wood reported a case with the presence of ameloblastomatous proliferation with masses of poorly differentiated or basaloid cells and suggested that these features might point to the existence of a neoplasm which is a subgroup of ameloblastoma rather than a variant of COC and they suggested the term “dentinogenic ghost cell ameloblastoma” [1, 16].

Discussion

In the present case features like location anterior mandible, unilocular radiolucency, cortical expansion, extensive dentinoid, ghost cells, sheets of proliferating odontogenic epithelial cells and ameloblastoma like follicles in mature stroma, favored the diagnosis of DGCT.

Presence of dentinoid and ghost cells can be observed in various other lesions such as ameloblastic fibrodentinoma, odontoma, dentinoma and dentinogenic ghost cell tumor. In the present case, Ameloblastic fibrodentinoma was ruled out because of lack of primitive ectomesenchyme like areas. Similarly, odontoma was eliminated as there were neither enamel spaces nor cementum like calcifications. We also ruled out dentinoma⁶ because even though dentin matrixes with ghost cells were present, absence of ectomesenchyme eliminated the possibility of dentinoma.

Following are the few features of DGCT reported in the literature with the current case.

Table 1: Clinical and radiological features

Dgct	Present Case	Matching Features
Central lesion more frequently	Central lesion	+
4 th decade	4 th decade	+
Male predilection (5:3)	female	-
Mandibular anterior region	Mandibular anterior region	+
Bicortical expansion present	Bicortical expansion, more lingually	+
Mixed radiolucency	Full radiolucency	?
Root resorption apically	Root divergence	?

All the histological features present case was in accordance with the previously reported literatures of DGCT except for the exuberance of dentinoid which could be due to

1. Interrupted epithelial – mesenchymal interaction as dentinoid was seen juxta epithelially ^[21].
2. Foreign body reaction to ghost cells leading to dystrophic calcification ^[21, 22].
3. Metaplastic calcification due to metaplastic changes in the connective tissue ^[23].

Conclusion

COC has been seen to be of extensive diversity in its clinical and histopathological features as well as in its biological behavior. The present case of 30-year-old female was diagnosed as DGCT, a tumorous form of COC, due to its characteristic histological features; numerous ghost cells and dentinoid material.

The excessive dentinoid and minimal cellularity, coupled with the unusual radiographic features in the present case, rendered a difficult diagnosis. More documentations of DGCT are definitely necessary to make better correlations.



Fig 1: Intraoral view depicting a diffuse swelling



Fig 2: Panoramic radiograph showing pear shaped well defined radiolucency

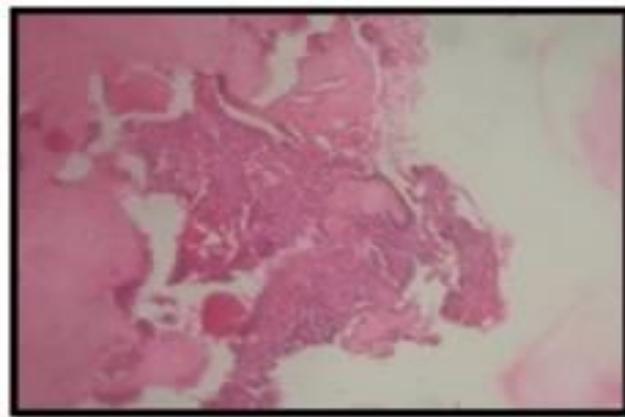


Fig 3: Sheets of proliferating odontogenic epithelium

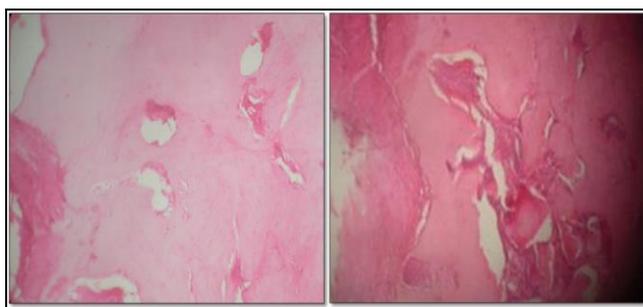


Fig 4: Ameloblastoma like follicles in mature connective tissue stroma (H&E).

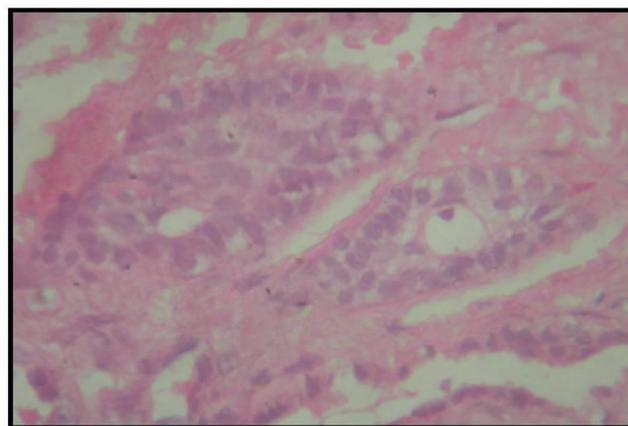


Fig 5: Ghost cell areas in the odontogenic epithelium just adjacent to dentinoid material.

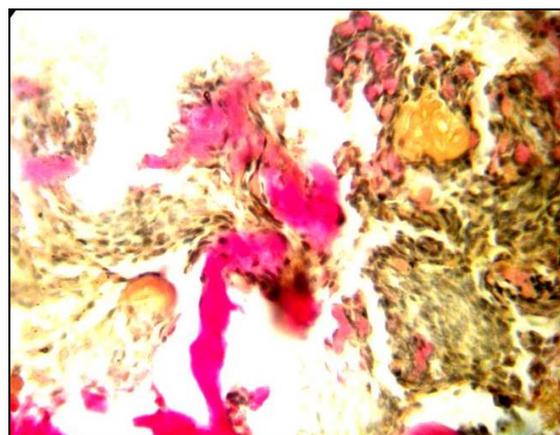


Fig 6: Red stained dentinoid (Van-gieson) & Yellow color ghost cells (Van-gieson).

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