Peripheral Dentinogenic Ghost Cell Tumor- Diagnostic Challenge in a Gingival Epulis: Report of a Case with Update of all Dentinogenic Ghost Cell Cases Reported in English Literature

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Abstract

Dentinogenic ghost cell tumor (DGCT), a variant of the calcifying odontogenic cyst, is considered to be a benign epithelial and mesenchymal neoplasm containing aberrant epithelial keratinization, ghost cells, and spherical calcifications. While there can be peripheral and central variants of this entity, the extraosseous type is rarer and usually innocuous in its clinical presentation. The aim of this article is to report a case of peripheral DGCT on the maxillary anterior region in a 14-year-old female evolving for 5 years and to emphasize the importance of histopathologic examination of gingival growths to avoid diagnostic pitfalls. A compilation of all reported cases in the English literature till date with details on the site, size, age sex, symptoms radiographic features, treatment follow-up, and recurrences has been attempted for better understanding of the biologic nature of this rare neoplasm.

Keywords: *Calcifying odontogenic cyst, extraosseous, gingival growth, peripheral dentinogenic ghost cell tumor*

Introduction

Calcifying odontogenic cyst (COC) is a rare, well-circumscribed, solid, or cystic lesion of odontogenic epithelial origin containing "ghost cells and spherical calcifications."[1] It represents a diverse group of lesions having both cystic and neoplastic variants. The neoplastic variant of COC with dentinoid formation was termed as dentinogenic ghost cell tumor (DGCT).[2] This rare neoplastic entity exists as both central and peripheral variants. The extraosseous type of DGCT is much rarer than the central variant presenting as painless swelling or nodule on the gingiva. The present case is being reported due to its innocuous clinical presentation with emphasis on the importance of histopathologic examination of gingival growths to avoid diagnostic pitfalls.

Case Report

Soft-tissue excisional biopsy specimen was received in the Department of Oral and Maxillofacial Pathology in ESIC Dental College and Hospital, Rohini, Delhi. The lesional tissue was fragmented and brownish in color. The clinical description was that of a swelling in the upper anterior gingiva in a 14-year-old female patient, which had been present for 4–5 years with a history of previous excision of the lesion from the same site 1 and a ½ years ago. The size of the present lesion as described was 1 cm × 0.5 cm in the region of the right maxillary central incisor. While no clinical photograph was available to substantiate the clinical appearance of the lesion, the orthopantomography revealed no evidence of bone involvement [Figure 1]. Differential clinical diagnosis was given as fibroma or pyogenic granuloma.

Histopathologic Description: hematoxylin- and eosin-stained sections showed superficial parakeratinized stratified squamous epithelium with elongated rete ridges. Underlying fibrocellular connective tissue showed areas of calcification which were trabecular as well as globular in appearance. Strands of proliferating ameloblastic odontogenic epithelium were seen with adjacent areas of eosinophilic dentinoid material. Eosinophilic amorphous globular ghost cells were noted in the received sections. Van Gieson special stain was undertaken to identify the ghost cells and dentinoid material in the

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sections. Dentinoid areas were stained red and located in close approximation to the ameloblastomatous strands of odontogenic epithelium. Ghost cells took up yellow stains and were seen scattered throughout the sections [Figure 2]. Based on the histopathologic appearance, a diagnosis of peripheral DGCT was given.

Discussion

COC was first described by Gorlin, Pindborg, Clausen, and Vickers in 1962. When reporting this new entity, the cystic nature of the lesion was described along with its peculiar histological features and pathogenesis.[3] The definition given in 1971 by the WHO was, "A nonneoplastic cystic lesion in which the epithelial lining shows a well-defined basal layer of columnar cells, an overlying layer that is often many cells thick and that may resemble stellate reticulum, and masses of (ghost) epithelial cells that may be in the epithelial cyst lining or in the fibrous capsule. The (ghost) epithelial cells may become calcified. Dysplastic dentin may be laid down next to the basal layer of the epithelium."[3] The term "dentinogenic ghost cell tumor" was suggested because of the prominent formation of dentinoid in relation to the epithelial islands, along with ghost cells in varying numbers,[4] and was considered to arise from entrapped odontogenic epithelial remnants in the maxilla, mandible, or within the gingiva. According to the dualistic concept, COC has two variants cystic and neoplastic[5] termed "calcifying cystic odontogenic tumor" and "dentinogenic ghost cell tumor" (DGCT), respectively.[1] With only 2%–14% of COC being solid tumors,[2] DGCTs are less common than the cystic variant and may exist both intra- and extraosseously and are notable by the presence of ameloblastomatous odontogenic epithelium, ghost cells, and dentinoid material. By 2019, there were only 57 cases of DGCT published (18 peripheral and 39 central).[3] A total of 99 cases of DGCT (40 peripheral and 59 central) have been reported till date [Table 1]. The peripheral variant is rare and usually occurs significantly later in life with a mean age of occurrence around 50 years (oldest age reported-92 years/female) with a slight predilection for males, anterior mandible,[6-8] and gingival soft tissues of the canine premolar region.[9] Our case, however, was of a young female (14 years) with an innocuous gingival lesion in relation to the maxillary right central incisor. Although PDGCT generally does not recur after excision,[6] the patient in our case gave a long-standing history of 5 years with a history of recurrence after surgical removal 1 ½ years ago. Since the previous surgery was not done in our hospital, the question of incomplete excision with subsequent growth till the present size of 1 cm × 1 cm × 0.5 cm is a possibility. It is of clinical significance to note that this lesion may be confused with reactive or inflammatory lesions of the gingiva, such as peripheral giant cell granuloma, pyogenic granuloma, irritation fibroma, epulis, or parulis,[10] and histopathologic examination is of utmost importance in diagnosing the case as PDGCT and not sign it off as an inflammatory lesion of the gingiva. Radiographically mild

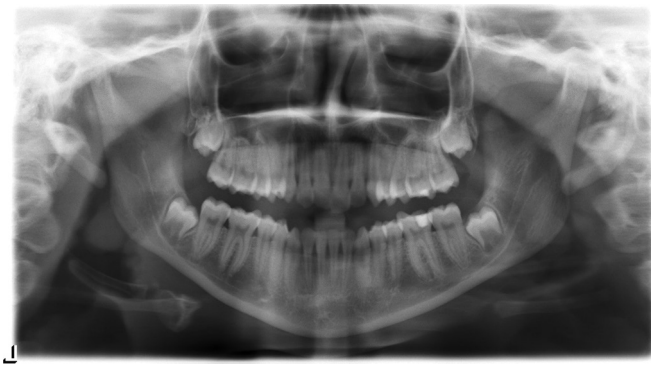

Figure 1: Orthopantomogram-showing no bone involvement in the Maxillary anterior tooth region

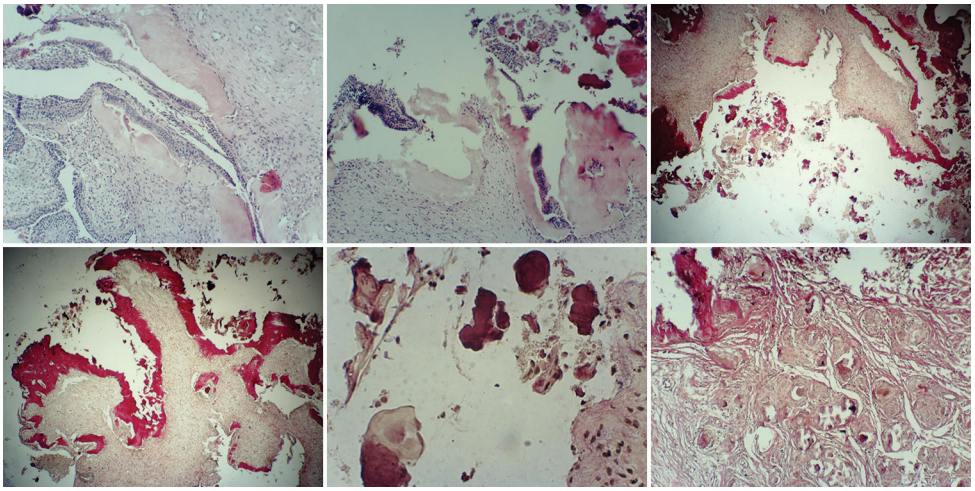

Figure 2: Right-to-Left. Row 1: 1–3; Row 2: 4–6. (1) ×10 H and E-stained section showing strands of ameloblastic cells with adjoining dentinoid material. (2) ×10 Van Gieson-stained section shows red color stained dentinoid material and loosely arranged connective tissue. (3) ×10 H and E-stained section showing odontogenic epithelium, dentinoid structure, and eosinophilic ghost cells. (4) ×40 Van Gieson-stained section showing ghost cells in variable degrees of calcification. (5) ×4 Van Gieson-stained section showing dentinoid material and ghost cells. (6) ×10 H and E-stained section showing ghost cells

Table 1: A summary of cases of dentinogenic ghost cell tumors reported to date

Dentinogenic Ghost cell Tumour [99 cases reported till date (Central :59 Cases; Peripheral :40 cases)]		
Peripheral Dentinogenic Ghost cell Tumour (40 cases reported*)		
Gender (No. of reported cases*)	Male (24*)	Female (16*)
Age-Oldest	82 years	92 years
Age-Youngest	8 years	14 years (our case)
Duration of Lesion before treatment	3 months to 8 years	
Site predilection	Mand.Antr > Max.Antr > Mand.postr. > Max.postr	
Treatment	Surgical excision	
Recurrence	No recurrence seen in any of the reported cases except for our case	

erosion or saucerization of the underlying cortical bone may be seen in 20% of cases,[11] although no such evidence of bone involvement was noted in our case [Figure 1]. The present case was also clinically diagnosed as pyogenic granuloma and the excised specimen was sent for routine histopathologic examination. The finding of ameloblastic strands along with masses of dentinoid material and the presence of ghost cells in various stages of calcification was confirmatory of PDGCT as the diagnosis. Van Gieson staining helped us to see the ghost cells as yellowish stained structures which are thought to be aberrant keratinization, whereas dentinoid material took up the red stain, thus confirming it as a connective tissue derivative.[9] The lesional ghost cells if in contact with the connective tissue could evoke a foreign-body reaction with the formation of multinucleated foreign-body type giant cells and may show calcifications ranging from fine basophilic granules to small spherical or globular bodies.[12] While peripheral DGCTs can be misdiagnosed as peripheral ameloblastomas,[13] they are positive for cytokeratin 19 and have a low Ki-67 index on immunohistochemical analysis.[14] The ghost cells themselves are cytokeratin AE1/AE3 positive, and the dentinoid material is positive for p53.[15]

Conclusion

The low number of cases published as peripheral DGCT makes case reports important in providing information that helps in their diagnoses and management. The present case was unique as it is among the 14 cases reported to fall under the age of 16 years and is among the 5 cases (3 male and 2 female) to have been diagnosed as PDGCT with 3 involving anterior maxillary region; 1 in the ethmoid sinus and 1 in the anterior mandible.[13,16-18] Another notable feature of most PDGCTs is their innocuously long evolution ranging from 3 months to several years. Fewer reported cases of peripheral DGCT may be attributed to the misdiagnosis of such cases as peripheral ameloblastomas.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal patient identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Sivapathasundharam B. Shafer's Textbook of Oral Pathology. 8th ed. India: Elsevier Publication; 2016. p. 73.
2. Bafna SS, Joy T, Tupkari JV, Landge JS. Dentinogenic ghost cell tumor. J Oral Maxillofac Pathol 2016;20:163.
3. Praetorius F, Hjørting Hansen E, Gorlin RJ, Vickers RA. Calcifying odontogenic cyst. Range, variations and neoplastic potential. Acta Odontol Scand 1981;39:227-40.
4. Juneja M, George J. Dentinogenic ghost cell tumor: A case report and review of the literature. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2009;107:e17-22.
5. Wright JM, Soluk Tekkesin M. Odontogenic tumors: Where are we in 2017? J Istanb Univ Fac Dent 2017;51:S10-30.
6. Wong YK, Chiu SC, Pang SW, Cheng JC. Peripheral dentinogenic ghost cell tumour presenting as a gingival mass. Br J Oral Maxillofac Surg 2004;42:173-5.
7. Cabo I, Saura A, Cardoso AR, Cardoso JA. Peripheral dentinogenic ghost cell tumour. BMJ Case Rep 2022;15:e245513.
8. Pommer B, Dvorak G, Rappersberger K, Jordan RC, Watzek G, Pogrel MA. Pyogenic granuloma mimicking peripheral dentinogenic ghost-cell tumour recurrence: Case report and meta-analytic literature update. Oral Surg 2013;6:98-103.
9. Kumar U, Vij H, Vij R, Kharbanda J, Aparna I, Radhakrishnan R. Dentinogenic ghost cell tumor of the peripheral variant mimicking epulis. Int J Dent 2010;2010:519494.
10. Bello IO, Qannam A, Al Zahrani A, AlDosari A. Peripheral dentinogenic ghost cell tumor: Report of a case and literature review. Int J Surg Pathol 2012;20:494-9.
11. Praetorius F, Ledesma Montes C. Dentinogenic ghost cell tumour. In: Barnes L, Eveson JW, Reichart P, Sidansky D, editors. World Health Organization Classification of Tumor. Pathology and Genetics of Head and Neck Tumours. Lyon: IARC Press; 2005. p. 314.
12. Hirshberg A, Dayan D, Horowitz I. Dentinogenic ghost cell tumor. Int J Oral Maxillofac Surg 1987;16:620-5.

13. Buchner A, Merrell PW, Hansen LS, Leider AS. Peripheral (extraosseous) calcifying odontogenic cyst. A review of forty-five cases. Oral Surg Oral Med Oral Pathol 1991;72:65-70.
14. da Silva Barros CC, de Souto Medeiros MR, de Azevedo RA, da Costa Miguel MC, Dos Santos JN, da Silveira ÉJ. Peripheral dentinogenic ghost cell tumor-report of two cases and review of the literature. Oral Maxillofac Surg 2021;25:575-9.
15. Pinheiro TN, de Souza AP, Bacchi CE, Consolaro A. Dentinogenic ghost cell tumor: A bibliometric review of literature. JODM 2019;3:9-17.
16. Abrams AM, Howell FV. The calcifying odontogenic cyst. Report of four cases. Oral Surg Oral Med Oral Pathol 1968;25:594-606.
17. Soluk Tekkesin M, Erdem MA, Ozer N, Olgac V. Intraosseous and extraosseous variants of dentinogenic ghost cell tumor: Two case reports. J Istanb Univ Fac Dent 2015;49:56-60.
18. Liu G, Li JN, Liu F. Peripheral dentinogenic ghost cell tumor of the ethmoid sinus: A case report. Medicine (Baltimore) 2020;99:e18896.