Dentinogenic ghost cell tumor: Tumor in the garb of a cyst!

Vaishali Korranne, Yash Merchant, Samir Joshi, Rajshekhar Halli

Departments of Oral Medicine and Radiology and Oral and Maxillofacial Surgery, Bharati Vidyapeeth University Dental College and Hospital, Pune, Maharashtra, India

INTRODUCTION

The calcifying odontogenic cyst (COC) was first described by Gorlin et al. in 1962. Formerly, the solid variant of COC was called calcifying ghost cell odontogenic tumor. Primarily, it has the features of a cyst, but it also has several prominent characteristics of a solid neoplasm, it was renamed dentinogenic ghost cell tumor (DGCT) by Praetorius et al. Subsequently, the nomenclature of this tumor was changed to DGCT at the 2005 World Health Organization (WHO) histologic classification of odontogenic tumors.

DGCTs were considered to be the solid, neoplastic tumor counterpart of COCs, an extremely rare odontogenic tumor. COCs account for 1%–2% of all odontogenic tumors and 2%–14% of all COCs are solid tumors.

DGCT may occur intraosseously or less commonly as an extrasosseous variant. Most extrasosseous DGCTs are peripheral lesions arising in the gingival or alveolar mucosa. Their behavior is typically unaggressive and can be controlled by local excision. No recurrences have been reported. Intraosseous DGCTs are more aggressive than extrasosseous DGCTs. More aggressive local resection is recommended for intraosseous DGCT, particularly if the tumor is radiologically ill-defined.

The case report describes a relatively painless, slow-growing asymptomatic lesion in the maxillary anterior gingiva that mimicked a reactive gingival lesion or a globulomaxillary cyst but turned out to be the extremely rare entity of a peripheral DGCT on histopathological examination of the excisional biopsy.

Access this article online

Website: www.jomfp.in
DOI: 10.4103/jomfp.JOMFP_144_16

How to cite this article: Korranne V, Merchant Y, Joshi S, Halli R. Dentinogenic ghost cell tumor: Tumor in the garb of a cyst! J Oral Maxillofac Pathol 2018;22:150.
CASE REPORT

A 26-year-old male reported to the clinic of the oral and maxillofacial consultant with the chief complaint of a growth in the upper left front part of the jaw. The patient did not have any pain, but the swelling was causing discomfort to the patient. The patient revealed that the swelling was slow growing and had attained the current size gradually over the past 12–14 months [Figure 1].

On extraoral examination, the base of the left ala was slightly raised. There was, however, no change in color/texture or erythema over the overlying skin or upper lip.

Intraorally, an ovoid mass was visible on the attached gingival between the maxillary left lateral incisor and canine. It had a reddish-pink color similar to the normal surrounding gingiva.

On palpation, it was firm and had a “ping-pong ball” feel. Written consent was obtained from the patient and the lesion was excised under local anesthesia [Figure 2] and sent for histopathological examination. It was a well-encapsulated lesion [Figure 3].

Histologically, under routine H and E staining, a connective tissue capsule with dentinoid tissue was distinctly visible. The cyst lumen showed characteristic ghost cells with calcification which are the hallmark of the dentinogenic ghost cell lesion [Figures 4-6]. Some fields depicted sheets and rounded islands of odontogenic epithelium in mature connective tissue resembling rosettes. Polygonal and polyhedral cells showing hyperchromatic nuclei were also visible. Mitotic figures were not found in these tumor cells, and there was no evidence of malignant transformation in these patients, but the tumor invaded the surrounding cortical bone and soft tissue.

DISCUSSION

DGCT is an uncommon locally invasive neoplasm, representing approximately 1.9%–2.1% of the overall odontogenic tumors. Peripheral DGCT is a rare odontogenic tumor, representing 13%–21% of all DGCTs. Clinically, it appears as a nodule of the gingival or alveolar mucosa and is usually asymptomatic. Peripheral occurrence of the cystic types of COCs is well documented in the English literature. This may result from cortical bone perforation by a central lesion or more rarely, true peripheral origin from gingival epithelial remnants.

Candido et al., 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. Contrary to other odontogenic tumors, DGCT predominantly occurs in later life. No predominance exists for the maxilla or mandible.

Histologically, DGCT is an infiltrative solid neoplasm composed of odontogenic epithelium associated with
ghost cell formation and production of dentinoid. Dentinoid is hyalinized eosinophilic material suggestive of immature or dysplastic dentin and is located closely near the epithelial sheet. Sheets and rounded islands of odontogenic epithelium are seen in mature connective tissue. The epithelium of the tumor islands resembles that of an ameloblastoma. Minor cysts may form in the epithelial islands. A characteristic feature is the transformation of the epithelial cells into 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, leaving only a faint outline of the original 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.

The aggressive or malignant counterpart of DGCT, odontogenic ghost cell carcinoma (OGCC), combines architectural and cytologic malignant features with prominent mitotic activity, infiltrative growth pattern, locally aggressive, destructive behavior and occasional distant metastasis. The malignant transformation of DGCT into OGCC has been described, so patients with DGCT, especially recurrent DGCT, should remain in long-term follow-up.

Other differential diagnoses include inflammatory and reactive gingival lesions such as epulis, globulomaxillary cysts and the peripheral variant of the ameloblastoma.

DGCT can be distinguished from ameloblastoma by the presence of large numbers of ghost cells and dysplastic dentin.

Some results suggest that conservative curettage and enucleation were not appropriate for treating DGCT, and the biological behavior of DGCT requires further study. The etiology remains an enigma till date and a thorough understanding of the pathophysiology ever since it has been described by Gorlin has been controversial.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.
REFERENCES

1. Gorlin RJ, Pindborg JJ, Odont, Clausen FP, Vickers RA. The calcifying odontogenic cyst: A possible analogue of the cutaneous calcifying epithelioma of Malherbe. An analysis of fifteen cases. Oral Surg Oral Med Oral Pathol 1962;15:1235-43.
2. Praetorius F, Hjorting-Hansen E, Gorlin RJ, Vickers RA. Calcifying odontogenic cyst. Range, variations and neoplastic potential. Acta Odontol Scand 1981;39:227-40.
3. In: Barnes L, Eveson JW, Reichart P, Sidransky D, editors. WHO Classification of Tumors: Pathology and Genetics of Tumors of the Head and Neck. Lyon, France: Oxford University Press; 2005.
4. Buchner A. The central (intraosseous) calcifying odontogenic cyst: An analysis of 215 cases. J Oral Maxillofac Surg 1991;49:330-9.
5. Sun G, Huang X, Hu Q, Yang X, Tang E. The diagnosis and treatment of dentinogenic ghost cell tumor. Int J Oral Maxillofac Surg 2009;38:1179-83.
6. Wong YK, Chiu SC, Pang SW, Cheng JC. Peripheral dentinogenic ghost cell tumor presenting as a gingival mass. Br J Oral Maxillofac Surg 2004;42:173-5.
7. Altini M, Farman AG. The calcifying odontogenic cyst. Eight new cases and a review of the literature. Oral Surg Oral Med Oral Pathol 1975;40:751-9.
8. Bhashar CS. The gingival cyst and the keratinizing ameloblastoma. Oral Surg Oral Med Oral Pathol 1965;19:796-8.
9. Candido GA, Viata KA, Watatube S, Vencio EF. Peripheral dentinogenic ghost cell tumor: A case report and review of the literature. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2009;108:e86-90.
10. Raubenheimer EJ, van Heerden WF, Sitzman F, Heyner B. Peripheral dentinogenic ghost cell tumor. J Oral Pathol Med 1992;21:93-5.
11. Fejerskov O, Krogh J. The calcifying ghost cell odontogenic tumor or the calcifying odontogenic cyst. J Oral Pathol 1972;4:273-87.
12. Goldenberg D, Seiabba J, Tufano RP. Odontogenic ghost cell carcinoma. Head Neck 2004;26:378-81.
13. Sedano HO, Pindborg JJ. Ghost cell epithelium in odontomas. J Oral Pathol 1975;4:27-30.
14. Stone CH, Gaba AR, Benninger MS, Zarbo RJ. Odontogenic ghost cell tumor: A case report with cytologic findings. Diagn Cytopathol 1998;18:199-203.
15. Li TJ, Yu SF. Clinicopathologic spectrum of the so-called calcifying odontogenic cysts: A study of 21 intraosseous cases with reconsideration of the terminology and classification. Am J Surg Pathol 2003;27:372-84.
16. Praetorius F, Ledesma-Montes C. Dentinogenic ghost cell tumour. In: Barnes L, Eveson JW, Reichart P, Sidransky D, editors. WHO Classification of Tumours: Pathology and Genetics of Head and Neck Tumors. Lyon: IARC; 2005. p. 314.
17. Ledesma Montes C, Gorlin RJ, Shear M, Torius FP, Mosqueda-Taylor A, Altini M, et al. International collaborative study on ghost cell odontogenic tumours: Calcifying cystic odontogenic tumour, dentinogenic ghost cell tumour and ghost cell odontogenic carcinoma. J Oral Pathol Med 2008;37:302-8.
Conference Secretariat

Dr. Simarpreet Virk Sandhu
Organising Chairman
883, Circular Road, Amritsar, Punjab-143 001
Mobile : +91-98888 87438
Email : iaompchairman@gmail.com

For Any Enquiry:

- +91-99836 99922
- +91-96808 26508
- iaomp2018@gmail.com
- iaompscientific@gmail.com
XXVII National IAOMP Conference
16-18th November 2018, Amritsar
Theme: Innovate - Integrate - Motivate

OFFICE BEARERS IAOMP

President
Dr. B. K. Das

President Elect
Dr. Veerendra Kumar B

Hon. Secretary
Dr. N. Chaitanya Babu

CONFERENCE ORGANIZING COMMITTEE

Chief Advisor
Dr. Alka Kale

Organizing Chairman
Dr. Simarpreet Virk Sandhu

Conference Secretary
Dr. Vijay Wadhawan

Organizing Secretary
Dr. Raman Deep S. Narang

Treasurer
Dr. Deepthi Sharma

Chairman Scientific Committee
Dr. Thippeswamy SH

IMPORTANT DATES:
Abstract submission deadline: 31st August, 2018
Pre-conference Course: 16th November, 2018
Inaugural ceremony: 16th November, 2018
Banquet: 17th November, 2018
AGM: 17th November, 2018
Valedictory function: 18th November, 2018

Venue: Radisson Blu Hotel, Amritsar