CASE REPORT

Dentinogenic Ghost Cell Tumor of Mandible in a Pediatric Patient with Dysplastic Changes

Ankit Natani[1], Santanu Borah[2], Monalisa Borah[3], Swati Agarwal[4], Manas Bajpai[5]

Abstract

Dentinogenic ghost cell tumor (DGCT) is a very rare entity with controversies in its terminology and classification. It is the neoplastic solid counterpart of the calcifying odontogenic cyst (COC), which was first reported by Gorlin et al. in 1962. There are around 31 cases reported in the literature. The mean age of occurrence is 40.27 years, although very rarely is it associated with the pediatric age group. We are reporting a case of DGCT with dysplastic changes in an 11-year-old child which is very rare. The present case deals with the clinical, radiological, and histopathological aspects of the disease and the importance of an appropriate diagnosis.

Keywords: Dentinogenic ghost cell tumor, Dysplastic changes, Ghost cells, Odontogenic tumor, Pediatric age.

International Journal of Clinical Pediatric Dentistry (2020): 10.5005/jp-journals-10005-1884

Introduction

Dentinogenic ghost cell tumor (DGCT) also known as odontogenic ghost cell tumor is a rare neoplastic counterpart of the calcifying odontogenic cyst (COC).[1] It is a locally invasive neoplasm characterized by ameloblastoma-like islands of epithelial cells in a mature connective tissue stroma.[2] Although it can be seen in any age group, its occurrence in pediatric age group is very rare. Dentinogenic ghost cell tumor is generally benign but malignant transformation is also reported.[3]

Dentinogenic ghost cell tumor is characterized microscopically by ameloblastoma-like odontogenic epithelial proliferation, presence of ghost cells, and dentinoid-like material.[4] Due to its diverse histological picture, several terms have been used by different authors to describe this lesion such as dentinogenic ghost cell tumor, calcifying ghost cell odontogenic tumor, keratinizing ameloblastoma, cystic calcifying odontogenic tumor, peripheral odontogenic tumor with ghost cell keratinization, dentinoameloblastoma, ameloblastic dentinoma, epithelial odontogenic ghost cell tumor, and odontogenic ghost cell tumor.[5] Dentinogenic ghost cell tumor 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. It is very rare in children and as per our best knowledge till date only a single case of DGCT has been reported in pediatric age group.[6]

Case Description

An 11-year-old child reported chief complaints of pain and swelling in the right lower posterior tooth region for the last 2 days. Extraoral examination revealed swelling on the right side of the mandible extending from the midline posteriorly up to the angle of the mandible measuring about 2 × 1 cm (Figs 1A and B). Intraorally a firm to hard swelling was seen on a lateral surface of the right side of the mandibular body extending from the mandibular right central incisor to the distal surface of a right mandibular first molar with obliteration of the labial and buccal vestibule (Fig. 1C). The panoramic radiograph revealed a large, well-circumscribed, multilocular, osteodestructive, radiolucent lesion extending from midline to the erupting permanent right second molar and was associated with an impacted canine (Fig. 1D). Based on the history, clinical examination, and radiographic evaluation, a provisional diagnosis of ameloblastoma was arrived at. The patient was referred to the department of oral and maxillofacial surgery and an incisional biopsy was performed. The histopathological examination revealed immature islands of odontogenic epithelium scattered in a connective tissue stroma resembling dental papilla and showed calcification at places resembling dysplastic dentin (Fig. 2A). The epithelium was hyperplastic at places and showed dysplastic features in form of basilar hyperplasia, nuclear hyperchromatism (Fig. 2B). Few aberrant keratinizations of epithelial cells were seen resembling ghost cells suprabasilarly (Fig. 2C). Based on histopathological examination, final diagnosis of DGCT arrived.

Discussion

Dentinogenic ghost cell tumor is a distinct but a rare histological entity among odontogenic ghost cell lesions which have been recently classified into the simple cystic type or COC; cysts associated with odontogenic hamartomas or benign neoplasms;

[1]Department of Pedodontics and Preventive Dentistry, NIMS Dental College, Jaipur, Rajasthan, India

[2]Department of Pedodontics and Preventive Dentistry, Jaipur Dental College and Hospital, Jaipur, Rajasthan, India

[3]Department of Public Health Dentistry, Jaipur Dental College and Hospital, Jaipur, Rajasthan, India

[4]Department of Pedodontics and Preventive Dentistry, Mahatma Gandhi Dental College and Hospital, Jaipur, Rajasthan, India

[5]Department of Oral and Maxillofacial Pathology, NIMS Dental College, Jaipur, Rajasthan, India

Corresponding Author: Ankit Natani, Department of Pedodontics and Preventive Dentistry, NIMS Dental College, Jaipur, Rajasthan, India, Phone: +91 9887783805, e-mail: Ankitnatani@gmail.com

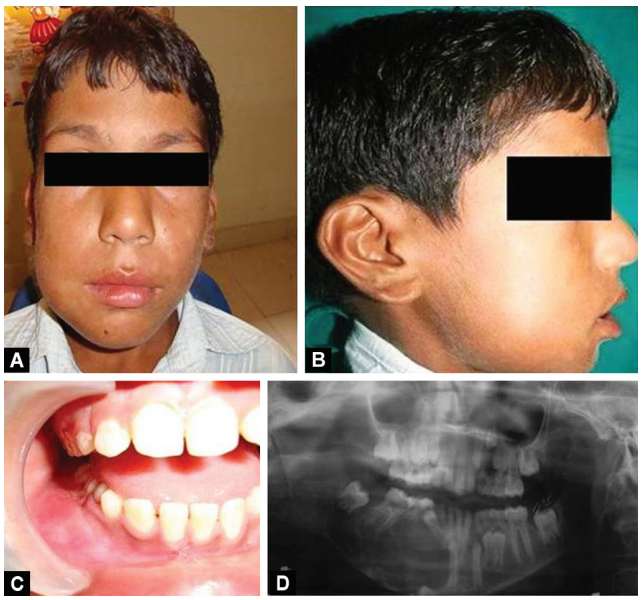
How to cite this article: Natani A, Borah S, Borah M, *et al.* Dentinogenic Ghost Cell Tumor of Mandible in a Pediatric Patient with Dysplastic Changes. Int J Clin Pediatr Dent 2020;13(S-1):S119–S121.

Source of support: Nil

Conflict of interest: None

© Jaypee Brothers Medical Publishers. 2020 Open Access This article is distributed under the terms of the Creative Commons Attribution 4.0 International License (https://creativecommons.org/licenses/by-nc/4.0/), which permits unrestricted use, distribution, and non-commercial reproduction in any medium, provided you give appropriate credit to the original author(s) and the source, provide a link to the Creative Commons license, and indicate if changes were made. The Creative Commons Public Domain Dedication waiver (http://creativecommons.org/publicdomain/zero/1.0/) applies to the data made available in this article, unless otherwise stated.

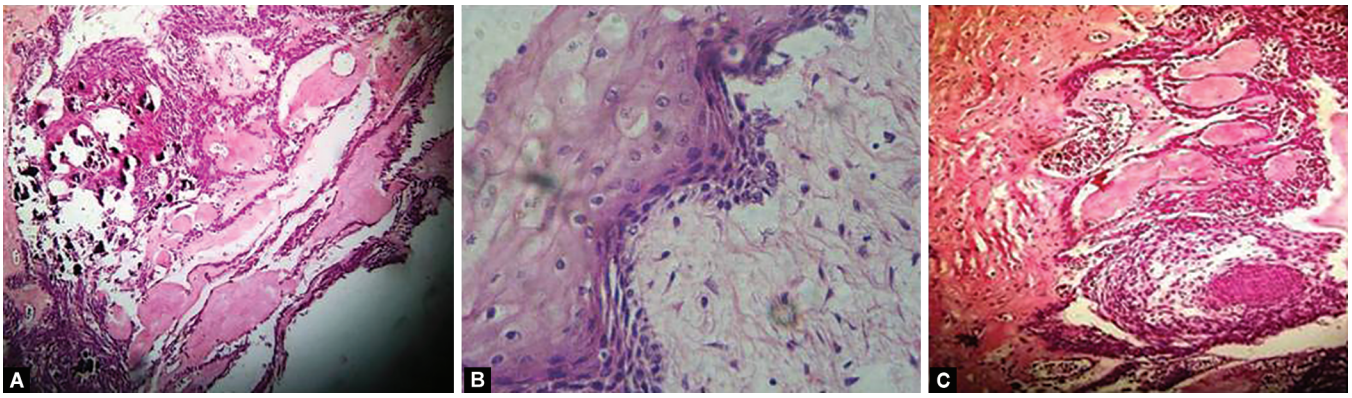
solid benign odontogenic neoplasm, which is same as COC but with dentinoid formation, the DGCT; malignant odontogenic neoplasm-ghost cell odontogenic carcinoma.[3,4] Dentinogenic ghost cell tumor was formerly considered as a solid variant of COC. In 1981, the name of DGCT was proposed first by Praetorius et al.[7] who tried to resolve the question on the cystic or neoplastic nature of the COC. Due to its diverse histological picture, several terms have been used by different authors to describe this lesion. Dentinogenic ghost cell tumor is a locally invasive neoplasm which shows more aggressive behavior and recurrence.[8] Since the first description of DGCT by Fejerskov and Krogh, although they used the term "calcifying ghost cell odontogenic tumor".[9] Approximately 32 cases of DGCT have been reported in English literature in the recent review of DGCT.[7] The mean age of occurrence is 40.27 years.[4] It is very rare in pediatric age group and the present case is so far only the second presentation of DGCT in a child. The first case was reported in a female child in Italy.[6] Present case is the first case of DGCT in a male child. There was no clear predilection for sex in the recent review,[8] whereas a slight predilection for men was documented in the WHO book published in 2005.[4] Dentinogenic ghost cell tumor may occur in any tooth-bearing area of the jaw but it is more commonly seen in mandible.[5,8] Similar pattern of gender and location was noted in the present case. Radiographs show a well-defined, mostly unilocular radiolucent to mixed radiolucent/radiopaque appearance depending on the amount of calcification.[4] However, multilocular appearance has been also reported in at least five cases.[8] The present case was unusual and showed a large multilocular radiolucent lesion resembling ameloblastoma or central giant cell granuloma radiographically. Histologically, the presence of ghost cells within the proliferative ameloblastomatous epithelium is the essential characteristic for the diagnosis. It contains variable quantities of dentinoid in the surrounding stroma, as well as in close contact with the epithelial islands or ghost cells. Also, dysplastic calcification may be found within the aggregates of ghost cells. The dysplastic changes in epithelium is not a very common phenomenon and reported in only seven cases.[6–8] The present case also showed the formation of dentinoid in the surrounding mature stroma with aberrant keratinization resembling ghost cells and dysplastic features in the form of basilar hyperplasia and nuclear hyperchromatism. The prognosis of DGCT seems to be depending on the treatment provided. The recurrence rate of DGCT after resection was reported up to 50% of reviewed cases with follow-up data.[6–9] As DGCT has a high recurrence rate after limited local resection, a wide local resection with an adequate safety margin appears to be the treatment of choice.[8,9] The malignant transformation of the DGCT after several recurrences has been documented,[4] so the patient should be kept under long-term follow-up.



Figs 1A to D: (A) Swelling on the right side of the face; (B) Side profile of the patient; (C) Intraoral picture shows swelling; (D) Panoramic radiograph reveals a radiolucent osteodestructive lesion of the mandible

Conclusion

Dentinogenic ghost cell tumor is a variant of COC. A proper clinical, radiological, and histological evaluation must be established to make a definitive diagnosis and the treatment should be accordingly planned for DGCT cases as it shows more aggressive biological behavior than CCOT and COC. Malignant transformation has also been reported in cases of recurrent DGCT and so regular follow-up is mandatory. Dentinogenic ghost cell tumor in pediatric age group is very rare and it should be diagnosed as early as possible to plan a proper treatment strategy.



Figs 2A to C: (A) Abundant dentinoid formation (hematoxylin and eosin staining ×40); (B) Overlying epithelium shows mild dysplasia (hematoxylin and eosin staining ×40); (C) Abundant aberrant keratinizaton in the form of ghost cells (hematoxylin and eosin staining ×40)

References

1. Quinn M, Brace M. Dentinogenic ghost cell tumor of the maxilla. Otolaryngology-Head Neck Surg 2008;139(4):604–605. DOI: 10.1016/j.otohns.2008.06.004.
2. Praetorius F, Ledesma-Montes C. Dentinogenic ghost cell tumour. World Head Organization classification of tumours. Pathology & genetics head and neck tumours Barnes L, Evenson JW, Reichart P, et al., ed., Lyon: IARC Press; 2005. p. 314.
3. Tomich CE. Calsifying odontogenic cyst and dentinogenic ghost cell tumor. Oral Maxillofac Surg Clin North Am 2004;16(3):391–397. DOI: 10.1016/j.coms.2004.03.006.
4. Barnes L, Everson JW. World Health Organization classification of tumors, pathology and genetics of the head and neck tumors. Lyon: International Agency for Research on Cancer; 2005. pp. 283–327.
5. Bajpai M. Ghost cells – an overview. Int J Clin Prev Dent 2011;7:205–208.
6. Ramaglia L, Esposito D, Bruno MP, et al. Dentinogenic ghost cell tumor: Histopathology and clinical aspects in pediatric age. Minerva Stomatol 2012;61(11-12):509–517.
7. Praetorius F, Hjørting-Hansen E, Gorlin RJ, et al. Calcifying odontogenic cyst. Range, variations and neoplastic potential. Acta Odontol Scand 1981;39(4):227–240. DOI: 10.3109/00016358109162284.
8. 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(5):17–22. DOI: 10.1016/j.tripleo.2009.01.033.
9. Thinakaran M, Sivakumar P, Ramalingam S, et al. Calcifying ghost cell odontogenic cyst: a review on terminologies and classifications. J Oral Maxillofac Pathol 2012;16(3):450–453. DOI: 10.4103/0973-029X.102519.