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CASE REPORT

An Intricate Clinicopathologic Presentation of Calcifying Odontogenic Cyst

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ABSTRACT

Calcifying odontogenic cyst (COC) is considered to be an uncommon benign entity which was first recognized by Gorlin. Calcifying odontogenic cyst accounts for only 2% of all the odontogenic tumors and is a relatively rare lesion which is characterized by histological diversity as its clinical and radiological features are not pathognomonic. Here, we report a case of COC in a 45 years old male patient with a long standing swelling.

Keywords: Gorlin cyst, Tumor, Ghost cells, Dentinoid.

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INTRODUCTION

True odontogenic cysts encompass an extensive part of jaw lesions wherein calcifying odontogenic cyst (COC) is an uncommon benign entity showing considerable amount of histopathological miscellany.[1] Calcifying odontogenic cyst was first recognized by Gorlin (thus named after him as 'Gorlin cyst'), Pindborg, Praetorius-Clausen and Vickers in 1962 and later by Gold. Ever since its recognition, controversy and confusion have existed regarding the relationship between non-neoplastic, cystic lesions and solid tumor masses and is considered to occupy a position between a cyst and an odontogenic tumor, having some characteristics of both. Because of this diversity, a 'dualistic' concept has been proposed by World Health Organization (WHO).[1,2] Calcifying odontogenic cyst, accounting for only 2% of all the odontogenic tumors, is a relatively rare lesion which has not been fully analyzed.[3]

The histologic variation of COC has led to different terminologies by various authors, such as calcifying ghost cell odontogenic tumor, dentinogenic ghost cell tumor, epithelial odontogenic ghost cell tumor and odontogenic ghost cell tumor. The array in the biologic behavior of these lesions have led to several theories and based on these, many nomenclatures and classifications have been published.[4] Here, we report a case of COC in a 45-year-old male patient with a long standing swelling.

CASE REPORT

A 45-year-old male patient reported with a swelling in the upper left back jaw region since 1 year. The swelling was gradual in onset and increased in size over a period of time to attain the present size. It was associated with pain which was intermittent, dull and localized in nature and loosening of teeth. The patient also gave history of exfoliation of tooth in the same affected area. There was no history of any purulent or blood discharge, dysphagia and paresthesia. Medical, dental, family and personal histories were inconspicuous. There was no abnormality detected on general physical examination, and the vital signs were found within satisfactory limits. Extraorally, there was a gross facial asymmetry on the left middle one-third of the face.

On intraoral examination, solitary swelling was noted in the left maxillary posterior alveolar ridge region measuring approximately 5 × 3 cm in size. The swelling extended from the maxillary left canine region to the left retromolar region anteroposteriorly. Mediolaterally, it extended 1.5 cm away from the midpalatine raphae to the buccal vestibular region. On palpation, the swelling was mild tender, firm in consistency and fluctuant in nature. The teeth associated with the swelling were 23, 24, 25, 27, 28 which were mobile and 26 was clinically missing (Fig. 1). The patient was subjected to radiographical investigations. Panoramic radiograph revealed well-defined homogenous unilocular radiolucency with a thin sclerotic radiopaque border extending from 22 to 28. Root resorption of 22, 23 and 24 was also seen (Fig. 2). Fine needle aspiration cytology did not yield any positive result.

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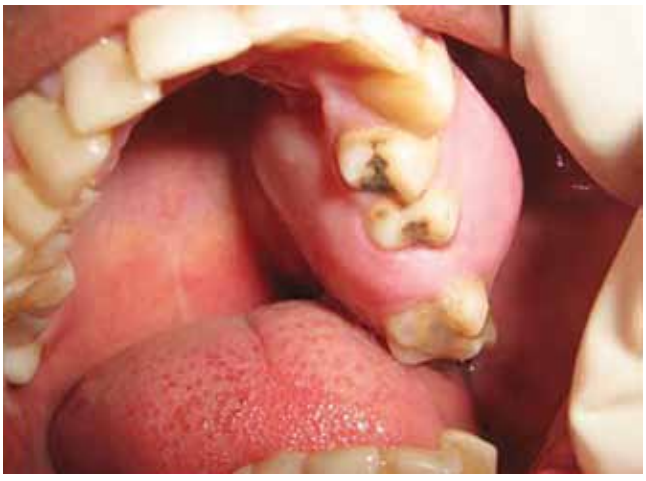

Fig. 1: Solitary swelling was seen in the left maxillary posterior alveolar ridge region

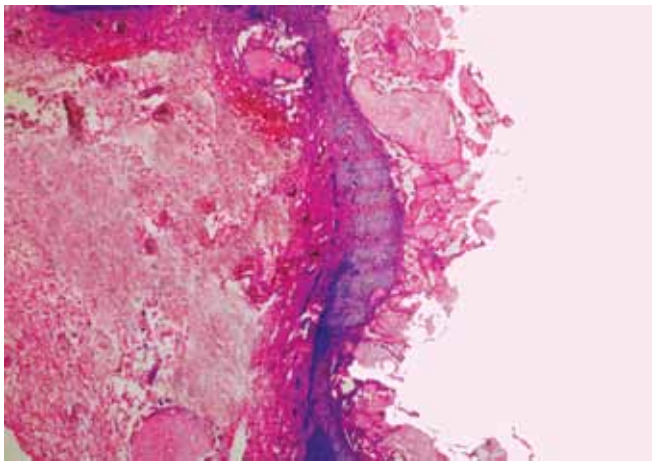

Fig. 3: Hematoxylin and eosin showing odontogenic epithelial lining with presence of ghost cells and eosinophilic dentinoid material

Furthermore, surgical intervention was carried out, and the specimen was sent for histopathological analysis. On microscopic examination, hematoxylin and eosin stained section revealed a well-defined cystic lesion with a fibrous capsule. There was presence of proliferative odontogenic epithelial layer with columnar and cuboidal like cells. Variable number of eosinophilic masses was seen within the epithelial component which represented ghost cells. These ghost cells masses are characterized by the loss of nuclei with preservation of the basic cell outline. A single area adjacent to the epithelial component is seen to be consisting of an eosinophilic matrix that represented dentinoid like material. Some ghost cells are seen to be consisting of fine basophilic granular material which may be due to the inductive effect of the odontogenic epithelium on the adjacent mesenchymal tissue. Underlying connective tissue stroma shows dense collagenous matrix with areas of hemorrhage present with a few inflammatory cells present (Fig. 3).

Based on all the aforementioned findings, final diagnosis of calcifying odontogenic cyst was obtained. Following surgery, prosthetic rehabilitation was done. The

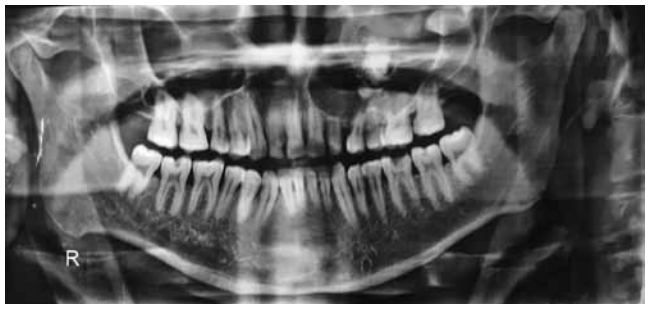

Fig. 2: Panoramic radiograph showing well-defined homogenous unilocular radiolucency with a thin sclerotic radiopaque border

patient was being followed-up after 3 months, 6 months and 1 year. No recurrence was noted henceforth.

DISCUSSION

Gorlin et al identified COC as a distinct pathological entity although, a similar occurring condition was previously reported in German literature in 1932 by Rywkind that was earlier thought to be an oral presentation of dermal calcifying epithelioma of Malherbe.[5] Calcifying odontogenic cyst was previously described as a non-neoplastic cystic lesion, but in 1992 according to the WHO classification of odontogenic tumor groups, the COC with all its variants was classified as an odontogenic tumor rather than a cyst. Calcifying odontogenic cyst is relatively a rare lesion that is characterized by histological diversity as its clinical and radiological features are not pathognomonic.[6,7]

The odontogenic origin of the COC is widely accepted. Praetorius et al suggested that it is a unicystic process that develops in the dental follicle, gingival tissue or bone from remnants of either odontogenic epithelium or reduced enamel epithelium. Dentinoid alone or an odontome associated with COC might be found in the cystic wall, induced by the epithelial lining. It can be thus said that the development of COC can be *de novo*.[2,8]

In 1981, Praetorius et al tried to classify COC by dividing it into two entities—cystic and neoplastic. Cystic entity was further classified into three types: Type 1A—a simple monocystic type with or without dentinoid calcified tissue, type 1B—cysts associated with odontogenic hamartomas or benign neoplasms and type 1C—monocystic ameloblastomatous proliferating type with dentinoid formation. Neoplastic entity is an odontogenic tumor characterized by presence of 'ghost cells' also known as dentinogenic ghost cell tumor.[9,10]

Toida M et al (1998) proposed basic classification based on 'dualistic' concept to overcome the shortcomings of the previous classifications. He proposed a classification in which he called CGCOC (calcifying ghost cell odontogenic cyst) for the cystic variant and calcifying ghost cell odontogenic tumor (CGCOT) for the neoplastic variant. He further subdivided the neoplastic group into—

cystic CGCOT and solid CGCOT, to include neoplasm showing cystic architecture and neoplasm with solid pattern respectively.[11]

The classification of the odontogenic ghost cell lesions was revised by Praetorius in 2006 as: Group 1—simple cysts; COC, group 2—cysts associated with odontogenic hamartomas or benign neoplasms: calcifying cystic odontogenic tumors (CCOT), group 3—solid benign odontogenic neoplasms with similar cell morphology to that in COC, and with dentinoid formation—dentinogenic ghost cell tumor and group 4—malignant odontogenic neoplasms with features similar to those of the dentinogenic ghost cell tumor—ghost cell odontogenic carcinoma.[9,10]

Calcifying odontogenic cyst predominantly is a central (intraosseous) lesion but the peripheral (extraosseous) lesion has also been reported. The intraosseous variant occurs with an equal frequency in the mandible and maxilla (1:1) and same gender predilection. It is found rarely in patients in the first decade of life, where it can occur in association with odontoma. The intraosseous variant is commonly seen in individuals of age ranging from 5 to 92 years, with peak incidence in the 2nd and 6th decades of life, whereas the extraosseous variety is more likely to occur in the 6th decade.[12] Our present case was also within the similar age range and site predilection.

The central COC normally appears as a painless, hard, slow-growing tumor, affecting the jaws that produce expansion rather than erosion of bone, with predilection to the incisor or canine region. However, our case presented with swelling in the cuspid region which extended up to the molars. The extraosseous COC is a localized, sessile or pedunculated circumscribed elevated gingival mass. In some cases, it may be associated with unerupted tooth mostly seen with the cuspids. Sometimes, the root resorption or displacement of adjacent teeth is reported and can be occassionally associated with the perforation of the cortical plate.[13,14]

The intraosseous COC usually presents as a unilocular or multilocular radiolucency with well-defined margins. With maturation, irregular radiopacities signifying calcifications or tooth-like densities are seen occasionally giving a salt and pepper appearance. The extraosseous lesions show saucerization of the bone and sometimes displacement of adjacent teeth.[12] Although, the reported case showed an unilocular radiolucency with well-defined margins and no calcifications.

Histologically, COC essentially is a well-defined cystic lesion with fibrous capsule and lined by odontogenic epithelial lining of approximately 4 to 10 cell layers thick and shows a palisaded basal layer of columnar or cuboidal cells with hyperchromatic nuclei. A loosely arranged overlying layer of cells that may resemble the stellate reticulum and usually containing masses of ghost cells which may be located within the epithelial lining or in the fibrous capsule may occasionally be seen.[14,15] Budding from basal layer into the connective tissue is often seen. Ghost cells are the most characteristic feature of COC. These are aberrant eosinophilic, ballooned, ovoid, swollen or elongated elliptoid epithelial cells which are devoid of nuclei and retain their basic cell outline. However, the nature of the ghost cells is not clear and many hypotheses have been proposed and are still debatable. Some of the hypotheses are as follows:

- Ghost cells have been considered as abnormal keratinized bodies.
- Might be a result of coagulative necrosis.
- They may represent simple cell degeneration or a form of enamel matrix, or,
- Might derive from the apoptotic process of odontogenic cells or represent different stages of normal and abnormal keratin formation, therefore, deriving from metaplastic transformation of odontogenic tumors. Presence of an atubular dentinoid is often found in the wall with close relation to the epithelial lining.[2,5,8]

The present reported case also showed similar histological features with presence of lining epithelium and eosinophilic ghost cell masses and dentinoid like material.

Imuunohistochemical analysis of lining epithelium in COC with Ki-67, proliferating cell nuclear antigen (PCNA), Bcl-2 have also been shown to give positive results in most of the cases reported, whereas ghost cells expressed only a few cytokeratins like AE1/AE3 and 34βE12.[8,15] However, the present case was clearly indicative of COC in histopathological analysis, thus, immunohistochemistry was not performed.

The treatment of choice for COC is enucleation with long-term follow-up. Recurrence depends on completeness of cyst removal. Malignant transformation COC is an exceptionally unusual.[6]

CONCLUSION

Calcifying odontogenic cyst may impersonate multiple odontogenic or nonodontogenic lesions, therefore, the clinical and histological diagnosis is complex. Henceforth, it is essential for the clinician to persuade thorough knowledge based on the clinicopathologic correlation of such multifaceted entities to arrive at a confirmative diagnosis.

REFERENCES

1. Eshghyar N, Jalayer-Nadery N, Ashery R. Calcifying odontogenic cyst: an analysis of thirty-six cases. Acta Medica Iranica 2006;44(1):59-62.
2. Sonawane K, Singaraju M, Gupta I, Singaraju S. Histopathologic diversity of gorlin's cyst: a study of four cases and review of literature. J Cont Dent Prac 2011;12(5):392-397.

3. Resende GR, Brito JA, Souza LN, Gomez RS, Mesquite RA. Peripheral calcifying odontogenic cyst: a case report and review of the literature. Head and Neck Pathol 2011;5(1):76-80.
4. Rastogi V, Pandilwar PK. Calcifying epithelial odontogenic cyst associated with complex odontome of maxilla. J Maxillofac Oral Surg 2013;12(1):1-5.
5. Vatsyayan A, Adhyapok AK, Debnath SC, Malik K. Calcifying odontogenic cyst: case report and review of literature. International J Dent Med Res 2014;1(4):69-75.
6. Cazal C, Sobral APV, Da silva VCR, De araújo VC. Extraosseous calcifying odontogenic cyst: a case report and a literature review. J Bras Patol Med Lab 2006;41(6):443-446.
7. Balaji SM, Rooban T. Calcifying odontogenic cyst with atypical features. Annals of Maxillofacial Surgery 2012;2(1):82-85.
8. Shear M, Speight P. Cysts of the oral and maxillofacial regions. 4th ed. Oxford, UK: Blackwell Publishing Ltd; 2007. p. 103-107.
9. Praetorius F, Hansen EH, Gorlin RJ, Vickers RA. Calcifying odontogenic cyst: range, variations and neoplastic potential. Acta Odont Scand 1981;39(4):227-240.
10. Knezevic G, Sokler K, Kobler P, Manojlovic S. Calcifying odontogenic cyst-Gorlin's cyst: report of two cases. Coll Antropol 2004;28(1):357-362.
11. Toida M. So-called calcifying odontogenic cyst: review and discussion on the terminology and classification. J Oral Pathol Med 1998;27(2):49-52.
12. Jinbu Y, Tsukinoki K, Tanobe K, Yamaguchi K, Itoh H, Watanabe Y, et al. Calcifying odontogenic cyst associated with complex and compound odontoma: report of a case and immunohistochemical study. Oral Med Pathol 2001;6(1):51-55.
13. Swan RH, Houston GD, Moore SP. Peripheral calcifying odontogenic cyst (Gorlin cyst). J Periodontal 1985;56(6):340-343.
14. Buchner A. The central (intraosseous) calcifying odontogenic cyst: an analysis of 215 cases. J Oral Maxillofac Surg 1991;49(4): 330-339.
15. Johnson A, Fletcher M, Gold L, Chen SY. Calcifying odontogenic cyst: a clinicopathologic study of 57 cases with immunohistochemical evaluation for cytokeratin. J Oral Surg 1997; 55(7):108-111.