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Case Report

Ameloblastomatous Gorlin's cyst

Mala Kamboj[1)] and Manish Juneja[2)]

[1)]Department of Oral Pathology and Microbiology, U.P. King George's University of Dental Sciences, Uttar Pradesh, India
[2)]Department of Oral Pathology and Microbiology, U.P. Dental College and Research Centre, Uttar Pradesh, India

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Abstract: The calcifying odontogenic cyst (COC), first described by Gorlin et al. in 1962, represents a heterogeneous group of lesions that exhibit a variety of clinicopathologic and behavioral features. COC has been categorized under two basic groups namely, cystic and neoplastic. Even after several classifications and sub-classifications, COC remains an enigma. Very few cases of ameloblastomatous COC have been reported in the literature. In this report, we present a case of ameloblastomatous COC / Gorlin's cyst, emphasizing on the rarity of the lesion and distinguishing it from ameloblastomatous ex COC pictographically. (J. Oral Sci. 49, 319-323, 2007)

Keywords: calcifying odontogenic cyst (COC); ameloblastomatous; mandible; ameloblastomatous COC vs ameloblastomatous ex COC.

Introduction

The calcifying odontogenic cyst (COC), first described by Gorlin et al. in 1962 (1), represents a heterogeneous group of lesions that exhibit a variety of clinicopathologic and behavioral features. COC has been categorized under two basic groups namely, cysts and neoplasms. The cystic type of COC comprises the majority of the cases, which are characterized by a unicystic lesion associated with or without an odontoma. They may also show ameloblastomatous proliferative activity intraluminally or intramurally (ameloblastomatous COC). The neoplastic variants of COC, which show a solid growth pattern consisting of ameloblastoma-like strands and islands of odontogenic epithelium infiltrating into mature fibrous connective tissue, are further sub classified into ameloblastoma arising from COC (ameloblastoma ex COC) and odontogenic ghost cell tumors (2). Malignant transformation of COC has also been reported (3).

In this report, we present a case of ameloblastomatous COC, emphasizing the rarity of the lesion and features, and distinguishing it from other types of COC.

Correspondence to Dr. Mala Kamboj, C-1111, Indiranagar, Lucknow, Uttar Pradesh 226016, India
Tel: +91-941-5408764
Fax: +91-522-2623405
E-mail: malskam@gmail.com

Case Report

A 58-year-old female visited the clinic with the chief complaint of pain in the right side of mandible for the past five years, and swelling in the associated region for the last two years. Oral examination revealed a large swelling extending from the canine up to the ramus causing buccolingual expansion of the affected bone. A hard, but fluctuant and cystic lesion was found near the angle and retromolar region of the mandible on palpation. The orthopantomograph (Fig. 1) revealed a multilocular radiolucency

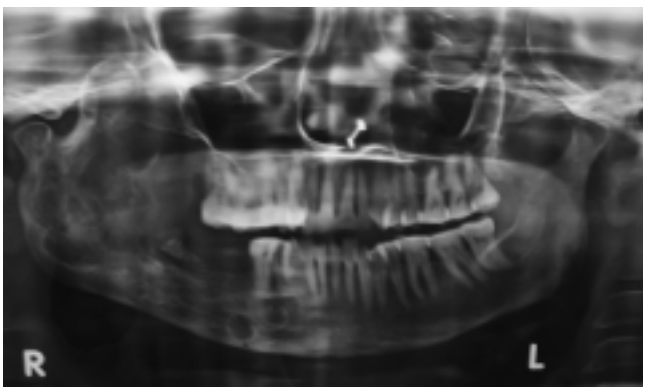

Fig. 1 Multilocular radiolucency on the right side of the mandible.

on the right side of the mandible extending from the canine region up to the condyle and coronoid areas. Based on the past history, clinical features and radiographic appearance, a provisional diagnosis of ameloblastoma or odontogenic keratocyst was made. Right side mandibulectomy was performed and an external plate was fixed in the area. The resected specimen was sent to the Department of Oral Pathology for histopathological examination. The patient has been under regular follow-up since then and has not exhibited any signs or symptoms of recurrence so far.

Histopathological examination revealed a cystic space lined by odontogenic epithelium comprising darkly stained basal cells, stellate reticulum like areas and many masses of ghost cells (Fig. 2). Occasional areas showed juxta-epithelial dentinoid formation (Fig. 3). Calcification of ghost cells was not observed. Ameloblastic proliferative activity was seen both intraluminally and intramurally lacking the histopathologic criteria for early ameloblastoma, as suggested by Vickers and Gorlin (4). The ameloblastomatous proliferations were mainly of follicular pattern with few follicles showing ghost cells (Figs. 4 and 5). Juxtaepithelial dentinoid formation was not seen around the ameloblastomatous proliferations. Based on these findings, a diagnosis of ameloblastomatous COC was made.

Discussion

In 1971, the World Health Organization (WHO) classification of odontogenic tumors defined COC as "a non-neoplastic cystic lesion in which the epithelial lining shows a well-defined basal layer of columnar cells, an overlying layer that is often many cell layers thick that may resemble stellate reticulum and masses of ghost cells that may be in the epithelial cyst lining or in the fibrous capsule.

Fig. 2 Cystic space lined by odontogenic epithelium and ghost cells (H&E, ×10).

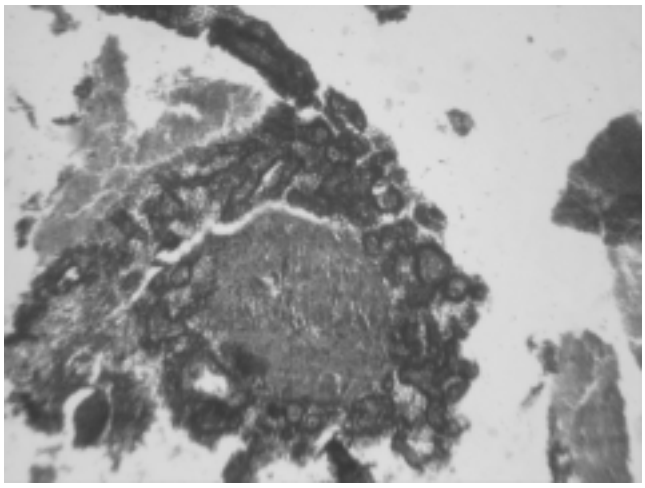

Fig. 4 Intraluminal ameloblastomatous proliferations showing follicular pattern (H&E, ×10).

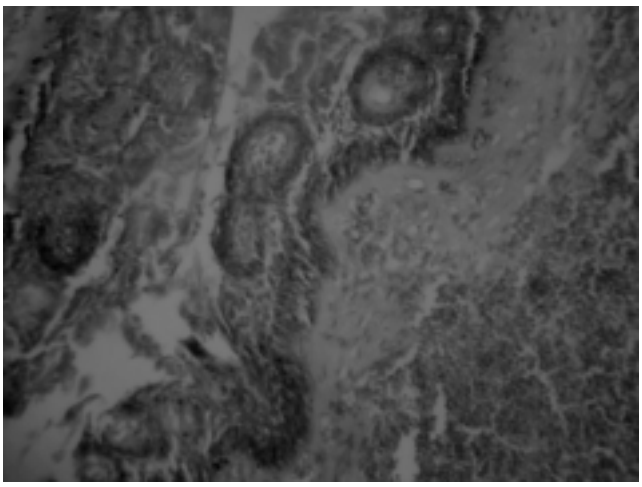

Fig. 3 Juxtaepithelial dentinoid formation (H&E, ×10).

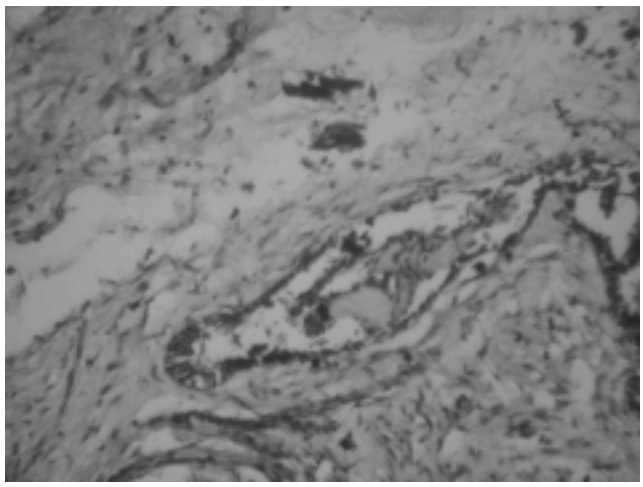

Fig. 5 Follicles showing ghost cells (H&E, ×10).

The ghost cells may become calcified. Dysplastic dentin may be laid down next to the basal layer of the epithelium." (5). Since then, it has been accepted that COC occurs as two different lesions – cystic and solid variants, both having different prognostic implications. Various classification systems have also been introduced to solve the problem of categorizing a particular lesion into its type (2,3,6-8). According to the new WHO classification of histological typing of odontogenic tumors, the COC constitutes a benign cystic neoplasia that presents an epithelium similar to an ameloblastoma, with ghost cells that may display calcifications (9).

COC is an uncommon lesion accounting for 1% of jaw cysts (9,10); the ameloblastomatous COC is even less common. Of 43,500 cases diagnosed by the Oral Pathology diagnostic service at the Indiana University School of Dentistry, only 34 cases of COC were reported. Seventeen more cases were diagnosed in the ensuing 11 years (1974-1984) (11). Hong et al. reported 92 cases of COC from the files of AFIP registry of Oral Pathology, out of which only 11 cases (14%) were ameloblastomatous COC (2). Aithal et al. (10) and Iida et al. (12) also documented single case reports of ameloblastomatous COC.

The cystic lesion can be divided into three basic types according to Praetorius et al.: simple unicystic type, unicystic odontoma producing type, and unicystic ameloblastomatous producing type (6). COC may exist not only with odontoma but also with other odontogenic tumors such as ameloblastic fibroma, ameloblastic fibroodontoma, ameloblastoma and adenomatoid odontogenic tumors, with a high recurrence rate after surgical intervention (2).

A simple unicystic COC is characterized by well-defined darkly stained basal cells, an overlying layer of stellate reticulum like cells and few or masses of ghost cells that may or may not show calcification. Juxtaepithelial dentinoid formation may be seen occasionally (3). However, it may be difficult to distinguish ameloblastomatous COC from ameloblastoma arising from COC (ameloblastoma ex COC). An ameloblastomatous COC represents areas similar to simple unicystic type along with intraluminal and intramural ameloblastomatous proliferation, which are usually plexiform in pattern but can be follicular, as seen in our case. The ameloblastoma-like proliferation typically lacks Vickers and Gorlin criteria of ameloblastoma-like cells (Fig. 6) and they also show occasional ghost cells and calcification within the proliferations. It can be differentiated from odontogenic ghost cell tumor by its obvious cystic structure and absence of juxtaepithelial dentinoid production (2). Ameloblastoma ex COC shows ameloblastic proliferation within the cystic wall without ghost cells and calcification. Vickers and Gorlin criteria for ameloblastoma-like cells can be easily identified (2). Table 1 summarizes the histological and clinical features of ameloblastomatous COC (10,12).

Presence of dentinoid deposition around the proliferations categorizes the tumor as an odontogenic ghost cell tumor, a type of solid variant which has a tendency to recur after

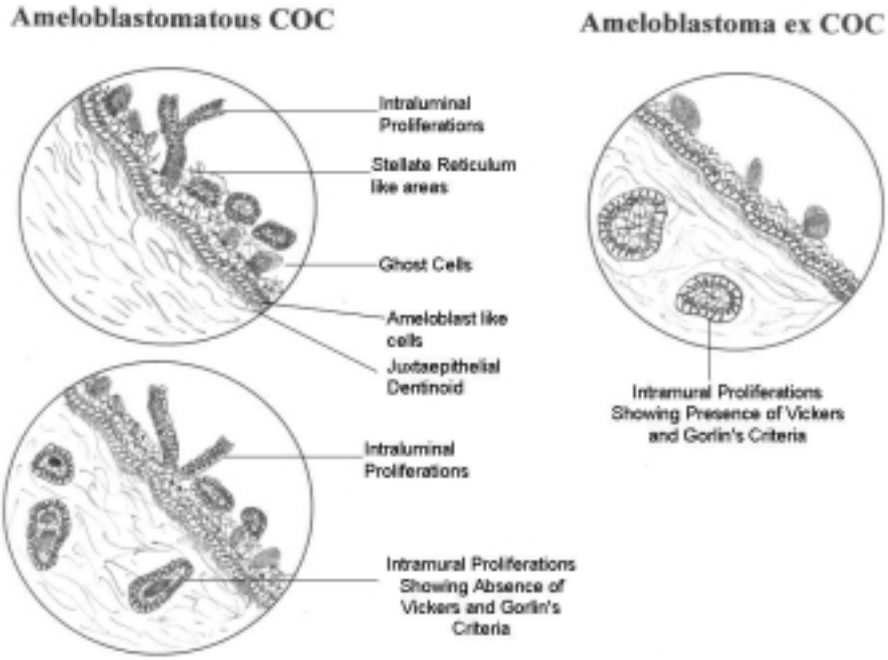

Fig. 6 Pictographic distinction between ameloblastomatous COC and ameloblastoma ex COC.

removal (2). The multicystic calcifying odontogenic tumor and the odontogenic ghost cell tumor can be clearly distinguished by the inductive activity seen around the odontogenic epithelial proliferations and formation of ghost cells in the latter case. This inductive effect of the tumor also distinguishes the odontogenic ghost cell tumor from ameloblastoma ex COC, apart from the presence of Vickers and Gorlin criteria and lack of ghost cells in the odontogenic epithelial proliferations in the latter case (2,3). Some authors (3,6) reported that ameloblastoma ex COC has a clinical course similar to that of ameloblastoma, and thus should be treated accordingly. Keeping in mind

Table 1 Clinical and histological features of ameloblastomatous COC

Authors with year	**Clinical Features**	**Histopathological Features**
Aithal et al. (2003)	*Age:* 28-years *Sex:* Female *Clinical presentation:* Painless swelling in the left posterior region of the mandible. Intraorally, well-defined bony hard, non tender swelling of 2.5 x 2.0 cm with smooth surface in relation to mandibular first and second premolars extending to the floor of the mouth. The mucosa overlying the lesion was intact. *Radiographic findings:* Multilocular radiolucency in the left mandibular posterior region extending from the mesial surface of the canine to that of the second molar.	Ghost cells in the cystic epithelium and juxtaepithelial hyalinization in some areas. Odontogenic epithelium in the form of rosettes and acanthomatous ameloblastic islands in the connective tissue lining of the cyst.
Iida et al. (2004)	*Age:* 17-years *Sex:* Male *Clinical presentation:* Bony swelling of the right mandibular body and facial asymmetry with slight pain on palpation at swollen area. *Radiographic findings:* Well-defined multilocular radiolucency from the right lower second molar to the right ramus with a remarkable bony expansion toward buccal and lingual sides. Involvement of the entire ramus and coronoid process. The lesion contained the unerupted lower second molar dislocated inferiorly to a position below the first molar.	Presence of odontogenic epithelium with many masses of ghost cells with calcification, and solid parts showing ghost cells and ameloblastomatous proliferations seen in the connective tissue of the cyst wall.
Present Case	*Age:* 58-years *Sex:* Female *Clinical presentation:* Pain in the right side of the mandible for the past five years, and swelling in the associated region since the last two year. Intraorally, a large swelling was seen extending from the canine up to the ramus causing bucco-lingual expansion. On palpation, a hard, but fluctuant and cystic lesion was felt near the angle and retromolar region of mandible. *Radiographic findings:* Multilocular radiolucency on the right side of the mandible extending from the canine region up to the condyle and coronoid areas.	Cystic space lined by odontogenic epithelium comprising of darkly stained basal cells, stellate reticulum like areas and many masses of ghost cells. Occasional areas showed juxtaepithelial dentinoid formation. Calcification of ghost cells was absent. Ameloblastic proliferative activity was seen both intraluminally and intramurally lacking the histopathologic criteria for early ameloblastoma as suggested by Vickers and Gorlin. The ameloblastomatous proliferations were mainly of follicular pattern with few follicles showing ghost cells. The juxtaepithelial dentinoid formation around the ameloblastomatous proliferations was not seen.

the above distinguishing features of different types of COC, we classified this lesion as ameloblastomatous COC, a rare entity.

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