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CASE REPORT

Ameloblastomatous Calcifying Odontogenic Cyst: A Rare Clinicopathologic Entity

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ABSTRACT

The calcifying odontogenic cyst represents a heterogeneous group of lesions that exhibits a variety of clinicopathologic and behavioral features. Therefore, a proper categorization of the cases is needed for better understanding of each variant. Ameloblastoma is one of the well-known odontogenic tumors that could be associated with calcifying odontogenic cyst. Very few cases of ameloblastomatous calcifying odontogenic cyst have been reported in the literature. In this report, we present a case of ameloblastomatous calcifying odontogenic cyst differentiating it from other variants of calcifying odontogenic cyst.

Keywords: Calcifying odontogenic cyst, Ameloblastomatous calcifying odontogenic cyst, Ghost cell.

INTRODUCTION

The calcifying odontogenic cyst (COC) was first described as a separate entity by Gorlin in 1962 and by Gold in 1963, as a benign odontogenic cyst. It is derived from odontogenic epithelial remnants within the mandible or maxilla or from the gingiva.[1] Calcifying epithelial odontogenic cyst (CEOC) represents a heterogeneous group of lesions that exhibit a variety of clinicopathologic and behavioral features and often occurs in association with odontogenic tumors, such as complex odontoma and ameloblastoma.[2,3] In 1992, WHO classification by Kramer and Pindborg used the term calcifying odontogenic cyst and described it as cystic or neoplastic variants in the jaw and the majority of authors also categorized under two basic groups of cyst and tumors. In this report, we present a case of ameloblastomatous COC, emphasizing its features, rarity of its occurrence and distinguishing it from other variants of COC.

CASE REPORT

A 65-year-old male reported with a complaint of swelling and pain in left lower jaw. Patient was asymptomatic 5 years ago when he started a pain in relation to 36 region. On extraoral examination, a large swelling was seen which was firm, tender and nonfluctuant about 4 × 5 cm in diameter extending from the corner of mouth to the angle of mandible in the left side showing facial asymmetry. Intraoral examination revealed very large swelling in the posterior region of mandible obliterating lower vestibule extending from lower left distal surface of first premolar to mesial surface of third molar (Fig. 1). Orthopantomogram findings showed sharply circumscribed bilocular radiolucent lesion extending from the lower left canine to third molar region along the lower border of the body of mandible (Fig. 2).

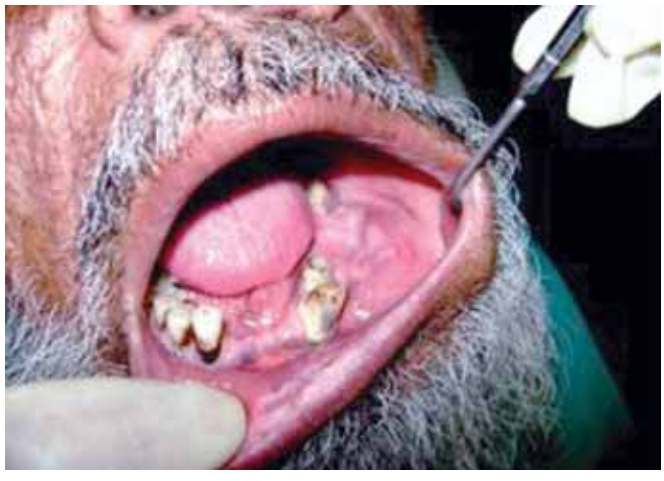

Fig. 1: Swelling in the posterior region of mandible obliterating lower vestibule

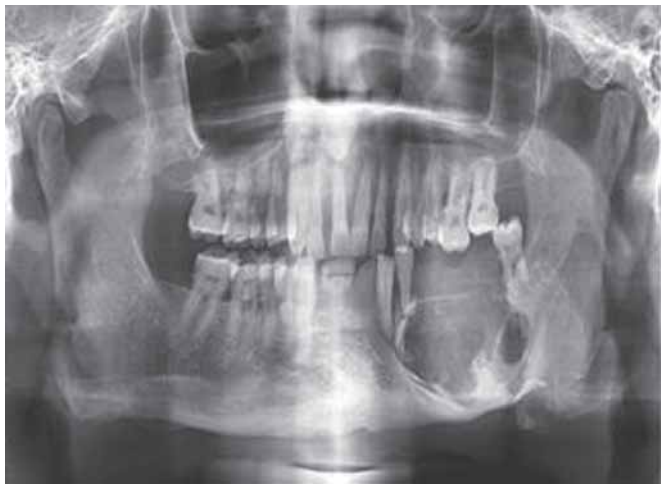

Fig. 2: Bilocular radiolucent lesion extending from the lower left canine to third molar region along the lower border of the body of mandible

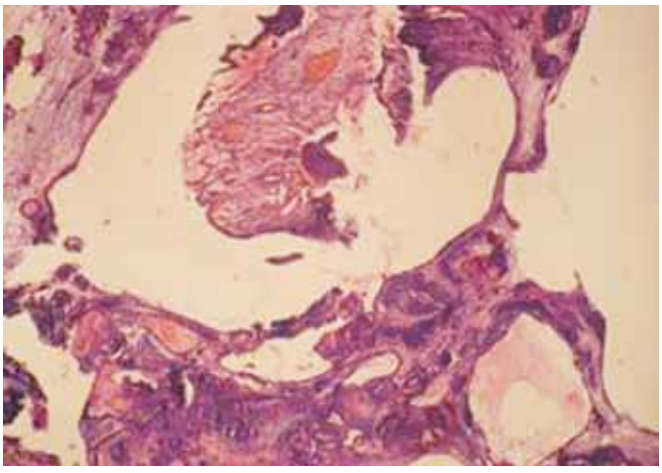

Fig. 3: Cystic areas lined by odontogenic epithelial lining with intraluminal ameloblastomatous proliferation with ghost cells (H and E ×4)

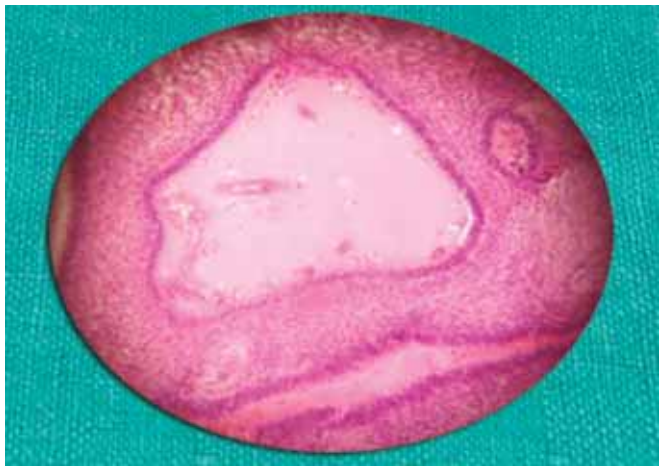

Fig. 4: Cystic spaces lined by tall columnar epithelium with lumina containing amorphous eosinophilic material (H and E × 10)

A clinical diagnosis of ameloblastoma was made. Incisional biopsy was advised and specimen submitted for histopathological examination. During the biopsy, a cystic lesion filled with pultaceous fluid was observed. Microscopically hematoxylin and eosin stained section showed large cystic areas lined by odontogenic epithelial lining with intraluminal and intramural ameloblastomatous proliferation and abundant stellate reticulum-like tissue. Homogeneous eosinophilic areas resembling ghost cells with large keratinizing areas and areas of concentric calcifications were also evident (Fig. 3). Wall of the specimen showed loosely arranged connective tissue with numerous blood vessels and extravasated blood. Based on the histopathological features, diagnosis of calcifying epithelial odontogenic cyst with ameloblastomatous proliferation was given. The patient underwent excisional biopsy along with normal tissue. The biopsy specimen sent for histopathological examination, revealed sheets of uniform appearing odontogenic epithelium with multiple cystic spaces. Cystic spaces were lined by tall columnar epithelium with lumina containing amorphous eosinophilic material (Fig. 4).

Some of the sheets of proliferating cells showed whorled appearance of odontogenic epithelium as seen in adenomatoid odontogenic tumor. Some areas showed loosely arranged stellate reticulum-like cells. Few areas showed the presence of eosinophilic ghost cells and areas of keratinization with specks of hematoxyphilic calcifications (Fig. 5). Large areas of eosinophilic coagulum, presumably dentinoid material or cystic secretory material with inclusions of inflammatory cells were seen. Based on the above features, a diagnosis of multicystic ameloblastomatous calcifying odontogenic cyst was given.

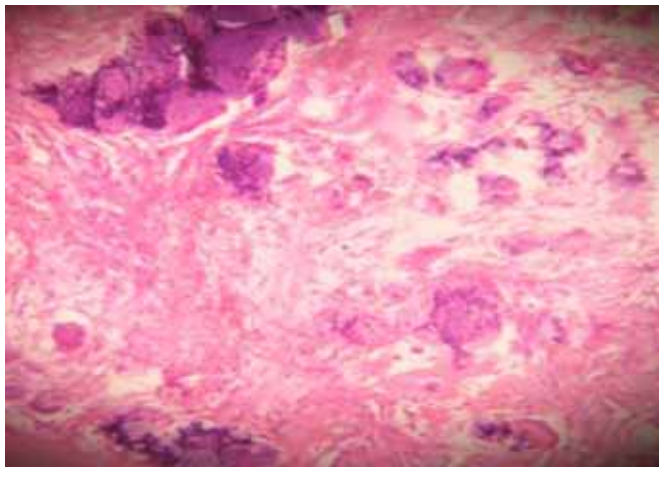

Fig. 5: Eosinophilic ghost cells with hematoxyphilic calcifications (H and E ×10)

DISCUSSION

The calcifying odontogenic cyst (COC) was first described in 1932 by Rywkind who reported a lesion of the jaws which was the same lesion as the cholesteatoma of the ear and, thereafter, called it as cholesteatoma of the jaws. In 1946, Thoma and Goldman described a lesion which they called a strange variant of an ameloblastoma but which was, in fact, a COC with areas which resembled an ameloblastoma. The COC is also referred as a Gorlin cyst and became recognized as a distinct pathologic entity when Gorlin et al described 11 cases and suggested an analog of the cutaneous calcifying epithelioma of Malherbe (1982).[4,11]

COC is an uncommon lesion and accounts for 1% of all odontogenic jaw cysts.[5] The age ranges from 1 to 82 years with peak in the second decade. In an observation of 215 lesions, Buchner and Praetorius et al have drawn attention to bimodal age distribution in support of their contention that two different entities may be involved with second decade and in sixth and seventh decade.[6] The lesion has no sex predilection and is equally distributed between maxilla and mandible, although the cases in the maxilla are more often in older patients. This lesion tends to occur in the canine-incisor portion in both jaws, but those in the maxilla

occur more often at the anterior portion than those in the mandible.[3] The COC are usually intraosseous (70% of the cases) and may account for extraosseous presentation only in 16 to 22% of cases.[5] In 1981, Praetorius et al tried to classify calcifying odontogenic cyst by dividing into two entities: A cyst and a neoplasm.[6] The cystic or non-neoplastic variant of COC is found to occur in 80 to 98% of cases and association with odontoma in 24% of cases. The solid or neoplastic variant of COC accounted for about 11.5% of cases. The cystic lesion can be divided into three basic types: Type 1—a simple unicystic type 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. Type 2—unicystic odontoma producing type with all the characteristics of previous type except that the hard tissue was complex or compound odontoma, and a presence of ameloblastic fibroma-like tissue in the cystic wall extending into the surrounding tissue. Type 3—unicystic ameloblastomatous producing type represents areas similar to unicystic type along with intraluminal and intramural ameloblastomatous proliferation, which are usually plexiform in pattern but can be follicular.[7] Similar features were observed in the present case where follicular type of proliferation was more evident. The neoplastic variant of COC, which shows a solid growth pattern consisting of ameloblastoma-like strands and islands of odontogenic epithelium infiltrating into mature fibrous connective tissue, are further subclassified into ameloblastoma arising from COC and odontogenic ghost cell tumor.

Ameloblastomous COC is very rare. Over 43,500 cases of jaw cysts diagnosed by the Oral Pathology Diagnostic Service at the Indiana University School of Dentistry, only 34 cases of COC were reported (Tomich et al 2004).[8] In the study by Hong et al 92 cases of COCs were reported from the files of AFIP registry of oral pathology out of which only 11 cases (14%) were ameloblastomatous COC.[3,9] Aithal et al[10] and Iida et al[3] also documented single case reports of ameloblastomatous COC. However, it may be difficult to distinguish ameloblastomatous COC from other variants of COC. A simple unicystic COC is characterized by well-defined darkly stained basal cell, and overlying layer of stellate reticulum and few ghost cells which may or may not be calcified. An ameloblastomatous COC also represents features similar to simple unicystic type along with

Table 1: Differentiating features between ameloblastomatous calcifying odontogenic cyst, ameloblastoma ex calcifying odontogenic cyst, and odontogenic ghost cell tumor

	Ameloblastomatous calcifying odontogenic cyst	*Ameloblastoma ex calcifying odontogenic cyst*	*Odontogenic ghost cell tumor*
Clinical features	Age—2nd and 6th decades Sex—no predilection Site—mandible	Age—2nd and 6th decades Sex—no predilection Site—mandible	Age—older than 50 years Sex—male predilection Site—mandible
	Painless swelling causing hard bony expansion Displacement of teeth	Painless swelling of jaws	Jaw expansion Obliteration of maxillary sinus
Radiographic features	Unilocular or multilocular radiolucent lesion but flecks of opacity can be seen	Unilocular or multilocular or mixed radiolucent lesion	Multilocular radiolucent or mixed radiolucent lesion
Histopathological features	Cystic lining lined by columnar cell with an overlying layer of stellate reticulum-like cells with ghost cell that may or may not show calcification	Cystic lining lined by columnar cell with an overlying layer of stellate reticulum-like cells with ghost cell that may or may not show calcification	Ameloblastoma-like areas and odontogenic epithelial islands with ghost cells showing keratinization and calcification
	Cystic lining shows intramural and intraluminal ameloblastomatous proliferation which are usually plexiform in pattern but can be follicular	Ameloblastic proliferation within the cystic wall without ghost cells and calcification	Presence of dentinoid deposition around the proliferation categorizes the tumor as odontogenic ghost cell tumor
	Ghost cells and calcification within the proliferations are seen		
	Ameloblastoma-like cells are not present	Ameloblastoma-like cells can be easily identified	
	(Vickers and Gorlin criteria)[11]	(Vickers and Gorlin criteria)[11]	

intraluminal and intramural ameloblastomatous proliferation that may be plexiform or follicular in pattern also showing ghost cells and calcification within the proliferation. Ameloblastomatous COC may be differentiated from ameloblastoma ex COC which shows ameloblastic proliferation within cystic wall without ghost cells and calcification. Odontogenic ghost cell tumor also shows ameloblastic proliferation as a solid mass with ghost cell and dentinoid deposition around the proliferations. The clinicopathologic differentiating features between these lesions are tabulated (Table 1).

Though the demarcations between these entities are rather slim, the histopathologic presentation in the reported case was more in favor of ameloblastomatous COC and was diagnosed as such. The treatment of cystic lesion involves enucleation with long-term follow-up. Recurrence depends on completeness of cyst removal. Prognosis is good for cystic COC and less certain for neoplastic COC.[10]

Buchner[12] has suggested that if COC is associated with an ameloblastoma, its behavior and prognosis will be that of an ameloblastoma, not of a COC or cystic lesion. Our case did not show any evidence of recurrence after its thorough excision with healthy margins but it is no doubt that careful postoperative observations are necessary for COCs which are associated with an ameloblastoma.

CONCLUSION

COC is a unique lesion possessing both cystic and neoplastic potential and showing considerable number of variants clinically, radiographically and histopathologically. Whether these variants represent unrelated lesions developing simultaneously or single lesion with ghost cell change is an open question and awaits further study. Separation of cases of different variants of COC may lead to a better understanding of each variant and may aid in its classification and treatment modality.

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