

A rare variant of calcifying odontogenic cyst with ameloblastoma presentation

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SUMMARY

Calcifying odontogenic cyst is a rare entity which was first described by Gorlin, and also accounts for 1% of the jaw cysts according to Shear. Due to its diverse histopathology and variable clinical features, there has been a doubt regarding its nature as a cyst or a neoplasm. In this report we present a case of calcifying odontogenic cyst with mural ameloblastomatous presentation in the left body of the mandible in a 19-year-old male patient.

Keywords: calcifying odontogenic cyst, ameloblastoma.

INTRODUCTION

Calcifying odontogenic cyst (COC) is a rare developmental odontogenic cyst, first described by Gorlin in 1962 (1). According to Shear, it accounts for 1% of jaw cysts (2). COC has been classified under two basic groups, namely cysts and neoplasms. The cystic type of COC comprises the majority of the cases, which are characterized by an uncystic lesion associated with or without an odontoma. They may also show ameloblastomous proliferative activity intraluminally or intramurally (3). Malignant transformation of COC has also been reported (4). The aim of this report is to present the clinical radiological and histopathological features of a calcifying odontogenic cyst with mural ameloblastomatous proliferation which has been rarely reported.

CASE REPORT

A 19 yr old male patient visited our department with pain on the left side of the jaw since 4 months. It was also associated with an extra oral swelling which was diffuse, 3 cm below the alatrugal line,

inferiorly to the inferior border of the body of the mandible, anteriorly 1cm away from the chin and posteriorly to the angle of the mandible. Overlying skin appeared normal with local rise in temperature (Figures 1, 2). On intra oral examination, a swelling extending from the premolar to the ramus of the mandible with bucco lingual expansion of the involved cortical bone (Fig. 3). A hard but fluctuant and cystic lesion was seen at the ramus and the retromolar area. Hard tissue examination did not reveal decay or mobility of the teeth present. Pulp vitality testing was done. 33, 34, 35, 36 showed no response. 31 and 32 showed delayed response at 7 and 6 units respectively. The Orthopantomograph revealed a multilocular radiolucency extending from the first premolar to the third molar, involving the crown of the second molar. Root resorption was seen with respect to second premolar and first molar (Fig. 4A). Submentovertex view revealed cortical thinning and expansion in the left body of the mandible (Figure 4B). Axial CT of the mandible showed an expansile lytic area with few septations with soft tissue density in the body of the mandible on the left side extending into symphysis menti measuring 4.5×3.6×3.8 cm respectively (Fig. 5). There is thinning of cortex noted and small cortical breaks are noted on the medial and lateral aspects. The 2nd molar crown is seen within the cyst and there is resorption of the first molar root. No soft tissue was seen infiltrating outside the lesion. Based on the clinical appearance and the radiological features, differential diagnosis of ameloblastoma and odontogenic keratocyst were listed. Marginal mandibulectomy was done and the resected specimen was sent for histopathological examination. It

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Fig. 1. Clinical photograph of the patient in profile view



Fig. 2. Clinical photograph of the patient showing lateral expansion of the swelling



Fig. 3. Clinical intraoral photograph showing obliteration of the buccal sulcus



Fig. 4. Orthopantomograph of the lesion showing extent of the lesion

revealed odontogenic epithelium with tall columnar basal cells resembling ameloblasts having reverse polarity & overlying epithelium resembling stellate reticulum. Epithelium shows aberrant keratinization & induction activity at epithelial connective tissue interface. Odontogenic epithelium shows proliferation into underlying connective tissue (Mural Proliferation). Connective tissue is collagenous & shows few small odontogenic epithelial islands, bone trabeculae and blood vessels, suggestive of calcifying odontogenic cyst with mural ameloblastomatous proliferation (Fig. 6).

DISCUSSION

In 1971, the World Health Organisation described COC as “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

maybe in the epithelial cyst lining or in the fibrous capsule (5). Calcifying odontogenic cyst occurs intraosseously or extraosseously, with intraosseous being more predominant. Prior to separation of this entity by Gorlin et al, it was often regarded as some form of ameloblastoma (1). The COC is an uncommon lesion demonstrating considerable histologic diversity and presenting with variable clinical behaviors. Although, it is broadly considered to represent a cyst, some investigators prefer to classify it as a neoplasm (6). The question concerning the nature of the cyst appeared to be clarified by Toida, who recently categorized COC into a cyst and neoplasm (7). In the new classification of World Health Organisation(2005), the term calcifying cystic odontogenic tumor was replaced by calcifying odontogenic cyst (COC) which constitutes a benign cystic neoplasia presenting an epithelium with ghost cells which may display calcification in it (8).

According to Praetorius *et al.*, the cystic lesion can be divided into three basic types simple unicys-



Fig. 5. Submentovertex view of the patient showing lateral expansion of mandibular cortex



Fig. 6. Axial CT section showing an expansile lytic lesion in the body of the mandible

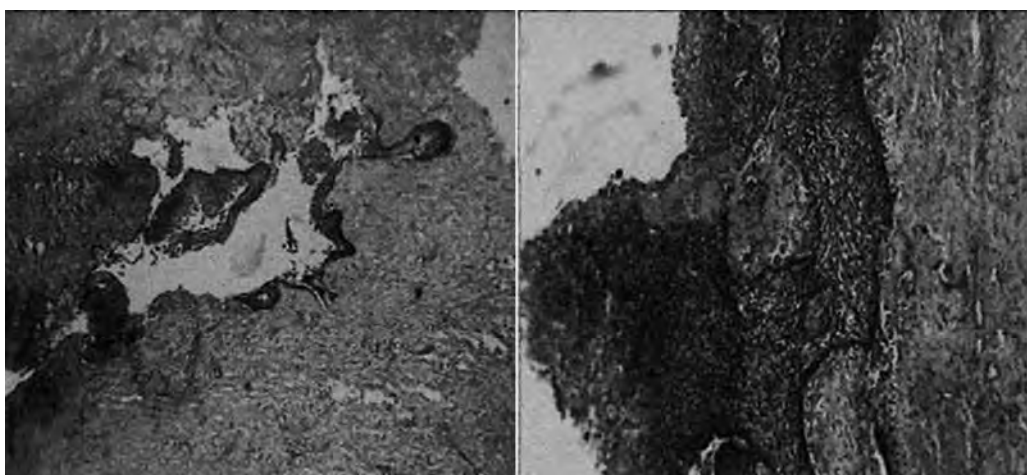


Fig. 7. H&E, 40X, Histopathological sections showing odontogenic epithelium and mural proliferation



Fig. 8. Post treatment orthopantomograph

tic type, unicystic odontoma producing type, and unicystic ameloblastomous producing type (9). Microscopically ameloblastomous COC resembles unicystic ameloblastoma except for the ghost cells and calcifications within the proliferative epithelium. Ameloblastomatous COC occurs only intraosseously (10) Ameloblastoma ex COC designates an ameloblastoma which arises from the cyst lining of COC(11). It can also occur intraosseously, appearing as cyst-like radiolucent lesion. Whether these tumors have the same disruptive potential and tendency for occurrence as a typical ameloblastoma is unknown (12)

Calcifying odontogenic cyst with mural ameloblastomatous presentation is a rare histologic variant. Our case did not show any signs of recurrence after surgery, but it is clear that periodic post operative observations are essential for COC's associated with ameloblastoma.

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