

Ameloblastomatous Calcifying Odontogenic Cyst: Case Report

ATOUSA AMINZADEH¹ and ALIREZA SADIGHI^{2*}

¹Department of oral maxillofacial pathology, school of dentistry, Islamic Azad university Isfahan (khorasgan) branch, Isfahan, Iran.

²Department of oral maxillofacial surgery, school of dentistry, Islamic Azad university Isfahan (khorasgan) branch, Isfahan, Iran.

Abstract

Calcifying odontogenic cyst (COC) or calcifying cystic odontogenic tumor was first introduced in 1962 by Gorlin *et al.*, as a possible oral counterpart of calcifying epitheliomas of Malherbe in skin. This lesion is a rare odontogenic lesion with variable clinico-histological characteristics. Three different histologic subtypes has been reported for COC. In this study we presented a female patient diagnosed with ameloblastomatous COC a very rare variant of this lesion and challenges regarding microscopic diagnosis and treatment of it is discussed.



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Introduction


Calcifying odontogenic cyst (COC) or calcifying cystic odontogenic tumor was first introduced in 1962 by Gorlin et al as a possible oral counterpart of calcifying epitheliomas of Malherbe in skin. COC is a rare odontogenic lesion with variable clinico-histological characteristics. According to literature only 2% of all odontogenic lesions are COC. Mostly COCs grow as cysts; however in less than 5% COCs can occur as a solid tumor-like mass.^{1,2}

The basic histopathologic criteria introduced for microscopic diagnosis of cystic COC is an odontogenic ameloblastoma-like epithelium containing numerous ghost cells and calcification.¹ But in regard to some microscopic details observed in a few cystic COCs three different histologic subtypes has been considered for cystic COC : simple unicystic type, odontoma producing type and ameloblastomatous proliferating type.³ 65% of cases are simple cystic which is composed of

CONTACT Alireza Sadighi ✉ Ar.sadighi@khuisf.ac.ir 📍 Department of oral maxillofacial surgery, school of dentistry, Islamic Azad university Isfahan(khorasgan) branch, Isfahan, Iran.



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squamous or stellate reticulum like epithelium with or without palisading of basal cells, large ghost cells with eosinophilic cytoplasm which may go through dystrophic calcification, dentinoid material and melanin deposits. Odontoma associated type actually shows the microscopic features of simple cystic type plus an odontoma associated with it and composes nearly 22% of COCs. Ameloblastomatous type shows stellate reticulum like areas with palisaded basal cells and reverse nuclear polarization and ameloblastoma proliferations in cyst wall.^{3, 4}

According to a literature review in pubmed only a few Ameloblastomatous COCs have been reported which are summarized in table 1.

In regard of proper diagnosis and treatment of this lesion the question is how to differentiate it from unicystic ameloblastoma and Ameloblastoma ex coc. And what would be the best treatment planning for this lesion in favor of patient and possibility of recurrence.

Table1:Ameloblstomatous COCs reported in Pubmed

Author (s)	Year	Country	Age	Gender	Place	Clinic	Treatment	Recurrence
Aithal <i>et al.</i> , (5)	2003	India	28	Female	Mandible	painless swelling well-defined bony, hard, non-tender swelling of 2.5 cm × 2 cm with smooth surface	-	No after 24 month
Iida <i>et al.</i> , (6)	2004	Japan	17	Male	Mandible	Painful swelling from second molar to ramus coronoid process	Enucleati-on	No after 13 years
Kamboj <i>et al.</i> , (7)	2007	India	58	Female	Mandible	Pain for 5 years and history of swelling for 2 years from canine to ramus, condyle and coronoid	Mandibul-ectomy	No
Singh <i>et al.</i> , (8)	2013	India	24	Female	Mandible	Swelling for 6 months Bone destruction and cortical thinning	Enucleati-on	No after 24 months
Samuel <i>et al.</i> , (9)	2013	India	13	Female	Mandible	Painless swelling with mild facial asymetry	Enucleati-on	No After 6 months
Menat <i>et al.</i> , (10)	2013	India	20	Male	Mandible	Swelling for 2 years from mandibular left first molar up to ramus condyle and coronoid Surface mucosa was intact cortical thinning	Hemiman-dibulecto-my with disarticul-ation	No after 12 months
Devaraju <i>et al.</i> , (2)	2015	India	65	Male	Mandible	Painful swelling watery discharge on chewing food Right mandible crossing midline	Not mentioned	Not evaluated
Bhat <i>et al.</i> , (11)	2015	India	19	Male	mandible	Swelling extended from premolar to ramus Small cortical perforations	Marginal mandibul-ectomy	NO

In this paper, a case of ameloblastomatous COC is reported which occurred in upper jaw pushing toward maxillary sinus and obliterating its space without bone perforation. In two year follow up after enucleation of cyst no recurrence was observed.

Case Presentation

A 63-year-old female with a two-year history of swelling in her right cheek and an aspiration biopsy report suggestive of hemangioma was referred to oral maxillofacial surgeon. In extra oral examination,



Fig. 1: Extra oral swelling in right cheek is observed

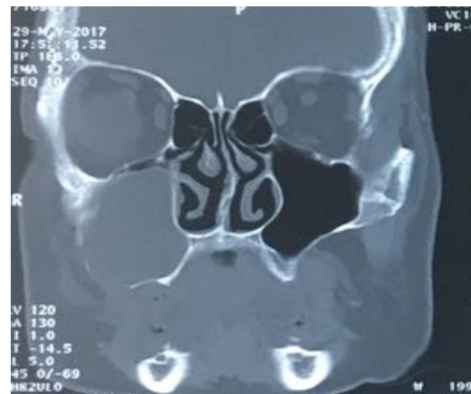


Fig. 2: CT scan showed obliteration of right maxillary sinus

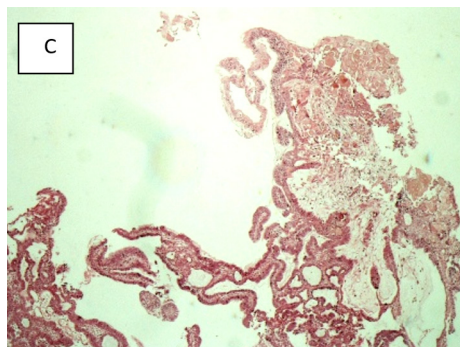
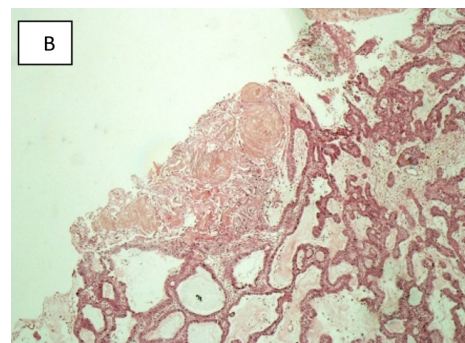
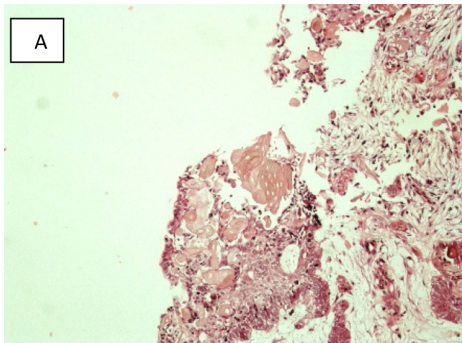


Fig. 3: A: cystic epithelium with numerous ghost cells was seen. B: Plexiform ameloblastomatous growth in cyst wall was observed. C: cystic epithelium, ghost cells and plexiform ameloblastoma growth is seen (HandE stain 10X)

a considerable asymmetry was noticed on the right side of zygomatic area (figure1).

Intraoral examination showed an expansion of right maxillary vestibule making her denture hard to fit. The patient had no history of pain or paresthesia. No cervical lymphadenopathy was noticed.

A doppler sonography was performed which denied the vascular nature of lesion. Facial and paranasal CT scan showed fullness and expansion of the lateral and medial wall of right maxillary sinus, but the orbital floor was not elevated. Other sinuses were clear, with no sign of sinus perforation. (Figures 2) Her past medical history was irrelevant. No history of allergy or other systemic diseases was reported by the patient.

With a provisional diagnosis of sinus mucocele and differential diagnosis of odontogenic cysts, a complete excisional surgery under general anesthesia was performed. A brown-gray soft tissue lesion in several pieces some with cystic structure was submitted to pathology laboratory in 10% buffered formalin. In microscopic examinations a cystic lumen lined with columnar basal cells and surface stellate reticulum like cells was observed (Figure3 a, b, c). Numerous ghost cells and spherical calcifications within the ghost cells was seen. A plexiform ameloblastomatous proliferation was observed in the cyst wall with multiple ghost cells. Accordingly a definitive diagnosis of ameloblastomatous COC was made. Patient was under post-operative follow up with no reported complication and recurrence 24 months after surgery.

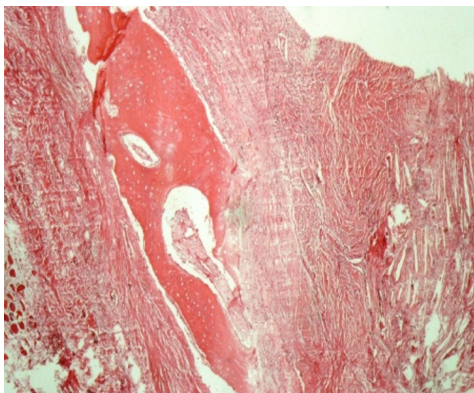


Fig. 4:lesion free sinus bony walls

Discussion

COC represents only one percent of all odontogenic cysts.³ The World Health Organization (WHO) has classified COC as a neoplasm and applied the term calcifying cystic odontogenic tumor for benign cystic types of COC and dentinogenic ghost cell tumor for benign solid-type COCs. Various clinical and histological features have been reported for this lesion. However, no united definition exists regarding the classification of COCs. For many years, the nature of COC resulted in conflicting theories on whether it is a cyst, neoplasm or hamartoma.¹²

The basic histopathologic criterion introduced for cystic COC is an odontogenic ameloblastoma-like epithelium containing ghost cells which may go through calcification.¹

Three different histologic subtypes have been reported for cystic COCs, including simple unicystic type, odontoma producing type and ameloblastomatous proliferating type.³ It is important to differentiate ameloblastomatous COC from unicystic ameloblastoma and ameloblastoma ex COC to prevent any unnecessary invasive treatment procedure. According to a literature review in PubMed, only a few ameloblastomatous COCs have been reported previously, which are summarized in table 1. With regard to the proper diagnosis and treatment of this lesion, the questions that arise are how is it differentiated from unicystic ameloblastoma and ameloblastoma ex COC? and what would be the best treatment plan for this lesion in favor of both patient and possibility of recurrence? In brief would this type of COC have different

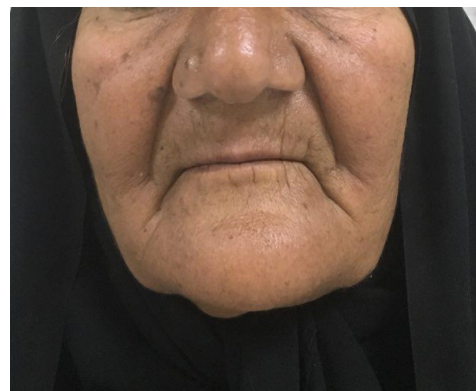


Fig. 5: 24 months after surgery

biological behavior, treatment and prognosis than other histologic subtypes?^{3,13,14,15}

COCs show nuclear and cytoplasmic positivity for beta catenin.⁴ But the immune-histochemistry staining is not available all time as it was not available for our patient so a histologic criteria is necessary for differentiating between these similar lesions with different treatment and prognosis. Ameloblastomatous COC shows intra luminal and intramural plexiform (sometimes follicular) ameloblastomatous proliferation admixed with ghost cells and calcification. Unicystic ameloblastoma has both intra-luminal and mural ameloblastomatous proliferation but lacks the ghost cells and calcifications seen in ameloblastomatous COC. Ameloblastoma ex COC shows ameloblastic proliferation within the cyst wall without any ghost cell or calcification.¹³

Adenoid ameloblastoma and odontogenic carcinoma can as well be considered in differential diagnosis of ameloblastomatous COC. Adenoid ameloblastoma also contains ghost cells and dentinoid similar to ameloblastomatous COC but has a pseudocribiform morphology. Odontogenic carcinoma has the dentinoid material but very few ghost cells and shows infiltration to adjacent stroma.⁴

In the present case selecting the proper surgical treatment was challenging. In the previously reported cases shown in table 1, ameloblastomatous

COCs have been mostly treated by enucleation, hemi-mandibulectomy was reserved for large cases accompanying bone perforation. In our case, although lesion pushed itself toward maxillary sinus, the sinus wall was clear and intact. (figure 4) Thus, it was decided to omit the lesion by enucleation and curettage, with preservation of the infraorbital nerve.

In a 24 month follow up, no sign of recurrence was noticed clinically and in CT scan (image not available) (Figure 5). In all the cases reported before, no recurrence was seen, which shows this lesion does not have a neoplastic nature despite the tumoral growth in its wall. The clinical significance of mural ameloblastomatous proliferation in COC cyst wall is still unclear.¹⁴ So until today enucleation and close follow remains the best treatment plan for small ameloblastomatous COCs without bone infiltration.

Conclusion

Ameloblastomatous COC should be differentiated from ameloblastoma ex COC, unicystic ameloblastoma and calcifying ghost cell odontogenic tumor to prevent unnecessary invasive approaches. Complete microscopic examination of the entire sample and a careful post operative follow-up is highly recommended.

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