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Case Report

Orthokeratinized Odontogenic Cyst of the Mandible: A Case Report

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Abstract

Orthokeratinized odontogenic cyst (OOC) is a developmental cyst that occurs in the maxilla and the mandible and was initially defined by the World Health Organization as the uncommon orthokeratinized type of odontogenic keratocyst (OKC). However, studies have shown that OOC has peculiar clinicopathologic aspects when compared with other developmental odontogenic cysts, especially OKCs. The orthokeratinized odontogenic cyst is a distinct clinicopathologic entity and is histologically characterized by a thin, uniform, epithelial lining with orthokeratinization and a subjacent granular cell layer. The basal cells are usually cuboidal or flattened. Clinically, the orthokeratinized cyst is a single cyst, shows a predilection for males, and is most often found in the second to the fifth decade. It is not dentigerous cyst but is often mistaken for a dentigerous cyst in the posterior mandible and exhibits little clinical aggressiveness. The purpose of the article is to present a case of OOC arising in the posterior mandible and highlight the importance of distinguishing it from the more commonly occurring Keratocystic Odontogenic Tumor.

Key words: Jaw Cysts;Odontogenic Cyst**;**Basal Cell Nevus Syndrome;Odontogenic Keratocyst; Dentigerous Cyst;Secretory granules.

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Introduction

Orthokeratinized odontogenic cyst (OOC) is a developmental cyst that occurs in the maxilla and the mandible, was initially defined by the World Health Organization (1992) as the uncommon orthokeratinized type of odontogenic keratocyst (OKC).[1] The lesion has been termed variously as an ‘‘orthokeratinized variant of OKC’’ or a ‘‘jaw cyst with orthokeratinization.’’ Li et al suggested a descriptive term ‘‘orthokeratinized odontogenic cyst,’’ which also reflected its most plausible histogenic origin. The World Health Organization new classification (2005) for head and neck tumors has designated OKC as Keratocystic Odontogenic Tumor (KCOT) and reclassified it as a neoplasm in view of its intrinsic growth potential and propensity to recur. According to this new classification, OOC should not be part of the spectrum of KCOT and should be distinguished from the latter.[2,3]

Case report

A 40 year female patient was referred to the Department of Oral and Maxillofacial Surgery with the chief complaint of tenderness and slight swelling in right cheek region since 6 months. Her past medical history was of no relevance and general physical status was good. The patient didn’t present any skin lesion suggestive of basal cell nevus syndrome. Lab findings which include her Hemoglobin %, TLC, DLC, Platelet count and Plasma glucose levels were within normal limits. Head and neck examination showed right facial swelling with an expanded mandibular buccal vestibule. The Orthopantomograph (OPG) showed a radiolucent lesion in a dentigerous relationship with unerupted third molar and resorption of the roots of first and second molars of the right posterior mandibular arch (Fig 1a & b). On the basis of clinical and radiographic findings a differential diagnosis of a dentigerous cyst, odontogenic keratocyst, ameloblastoma and calcifying odontogenic cyst was made. The lesion was enucleated and chemically cauterized with Carnoy’s solution, followed by a packing of iodoform dressing. Postoperatively antibiotics (amoxicillin + clavulanic acid) and analgesics were prescribed for 5 days. Gross examination (Fig 1c) of the excised specimen measuring 3 x 4 x 2 cm showed a cystic sac with a smooth luminal surface which was firm in consistency and brownish black in colour. The lumen showed traces of cheesy material.

Histopathology revealed an orthokeratinized stratified squamous epithelium of varying thickness lining a thin fibrous wall (Fig 2a).

Keratohyaline granules were prominent in the superficial epithelial layer subjacent to the orthokeratin. A hypo cellular spinous cell layer was made up of polyhedral to flattened cells with eosinophilic cytoplasm. The basal layer cells were low cuboidal and exhibited little tendency of nuclear hyperchromatism and palisading (Fig 2b). In places the epithelial lining was relatively thin and was only about 2 cells thick (Fig 2c). The features were suggestive of an Orthokeratinized Odontogenic Cyst. The postoperative course was uneventful and there were no signs of recurrence after a periodic follow-up of 14 months.

Discussion

The orthokeratinized odontogenic cyst clearly identified as an orthokeratinzed variant of the odontogenic keratocyst for the first time by Wright in 1981 owing to it's different histopathology and reduced likelihood to recur.[2] Although both the first two editions of the World Health Organization's histological classification of odontogenic tumors recognized cases with orthokeratosis, the WHO's 2005 edition excluded it from its definition of a KCOT.[3] The 2005 edition reclassified the parakeratotic type as a Keratocystic Odontogenic Tumour and stated ''Cystic jaw lesions that are lined by orthokeratinizing epithelium do not form part of the spectrum of a KCOT. Three histologic variants were recognized initially: a parakeratinized variant, an orthokeratinized variant, and combination of the two. The less aggressive clinical behavior and recurrence pattern of the orthokeratinized variant ultimately warranted the designation of the orthokeratinized variant as a separate entity, "Orthokeratinized Odontogenic Cyst".[3,4]

Vuhahula et al., found that reduced enamel epithelium that had completed its tooth-forming function had the capability to keratinize under appropriate stimuli, thus forming a true dentigerous cyst with keratinization.[5] The association with unerupted teeth suggests that many OOCs may have first developed during adolescence, when the third molars were developing, and were only noticed later either owing to the development of symptoms or as an incidental discovery during investigation of another dental problem.[6] The possibility should be considered that a cyst in a pseudodentigerous relation, in which the crown of an unerupted tooth was not inside the cyst, might be clinically and radiologically misinterpreted as a dentigerous cyst. Although orthokeratinized OKCs also occur as true dentigerous cysts, there is little evidence in the present study as to whether this arose primarily by keratinization of the reduced enamel epithelium or secondarily by the fusion of an extrafollicular orthokeratinized OKC with the lining of a normal dental follicle or dentigerous cyst.[4]

Zhu suggested that while the KCOT may arise from the dental lamina with the presence of the dental papilla required for its development, the OOC may arise from oral epithelium under the influence of dental papilla or only the oral epithelium. The histogenesis of KCOTs and OOCs may thus vary and needs further investigation. Although the histogenesis of OKCs has been debated most authors believe that they originate from dental lamina. This would explain their common occurrence in the posterior mandible, because the dental lamina is more active in this area at the age when many patients develop their cysts. This theory would also support an extra follicular development for dentigerous cysts.

The incidence of KCOT is about eight times more than that of the OOC, which also exhibits a slight male predilection. The clinical or radiographic features of OOC are not distinctive that differentiate them from other inflammatory or developmental odontogenic cysts. OOCs are generally solitary asymptomatic lesions, occurring in the third to fourth decade and with a male predilection.[3] The lesion usually appears as a unilocular radiolucency, but occasionally multilocular lesions are also encountered. About two thirds of OOCs are encountered in a lesion that appears clinically and radiographically as a dentigerous cyst. They occur more commonly in the mandible with an affinity for the posterior region and most often involve an unerupted mandibular third molar tooth. The size can vary from less than 1 cm to large lesions greater than 7 cm in diameter. OOCs are not found in patients with Naevoid Basal Cell Carcinoma Syndrome (NBCCS).[8]

Clinically the two entities (OOC & KCOT) exhibit an overlap in clinical and radiographic presentation. KCOTs also exhibit similar findings regarding age, sex and site of occurrence but they are associated with NBCCS patients and thus tend to exhibit multiple lesions.

Radiographically OOCs tend to be unilocular lesions and are more often associated with impacted teeth as compared to KCOTs.[5,8]

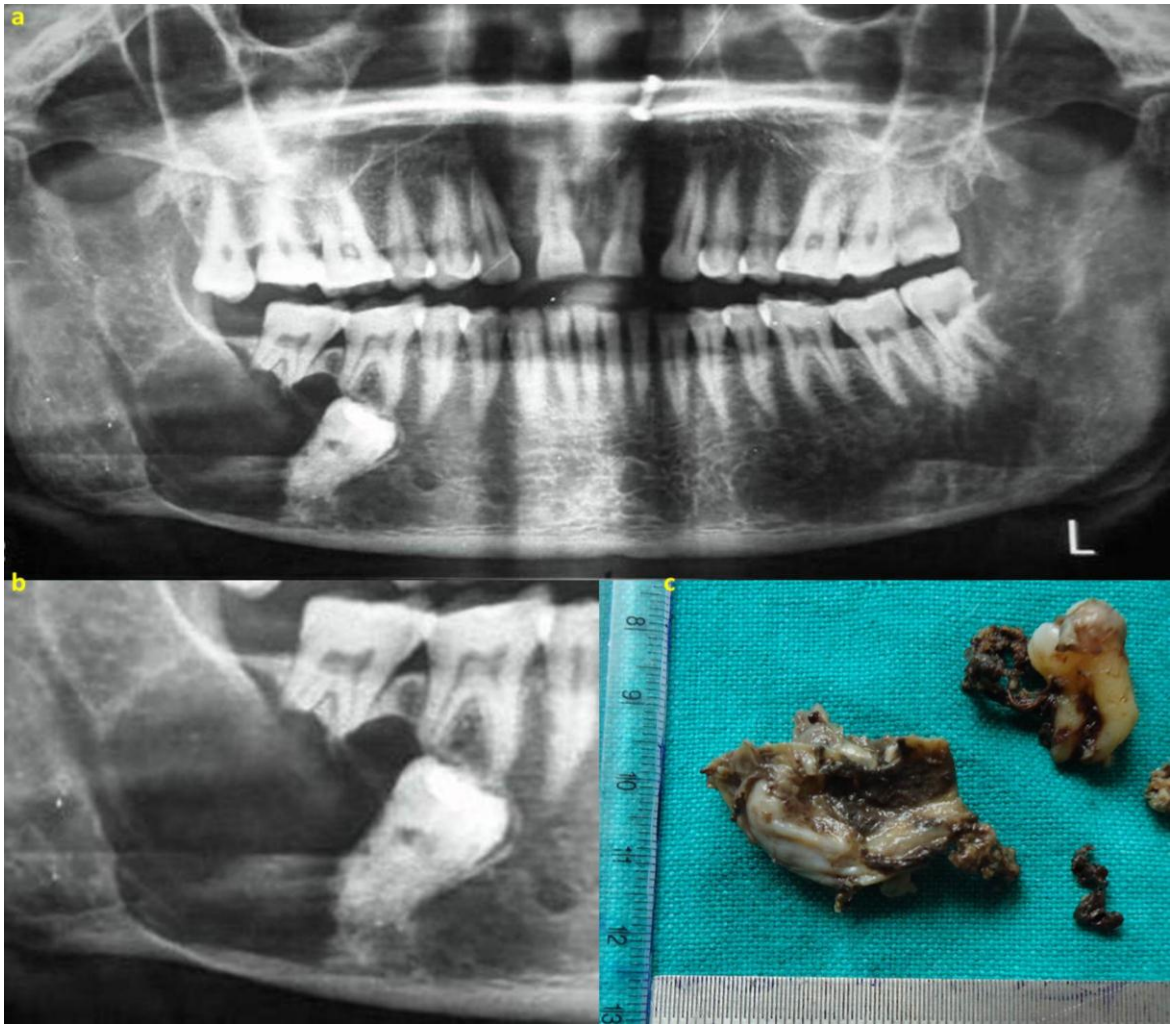

Figure 1: The orthopantomograph showing a radiolucent lesion in the right posterior mandibular arch associated with an impacted third molar with resorption of roots of first and second molar (a), Cropped view of the OPG showing the dentigerous relation of the lesion (b). Gross examination revealed a cystic lesion measuring 3 x 4 x 2 cm (c).

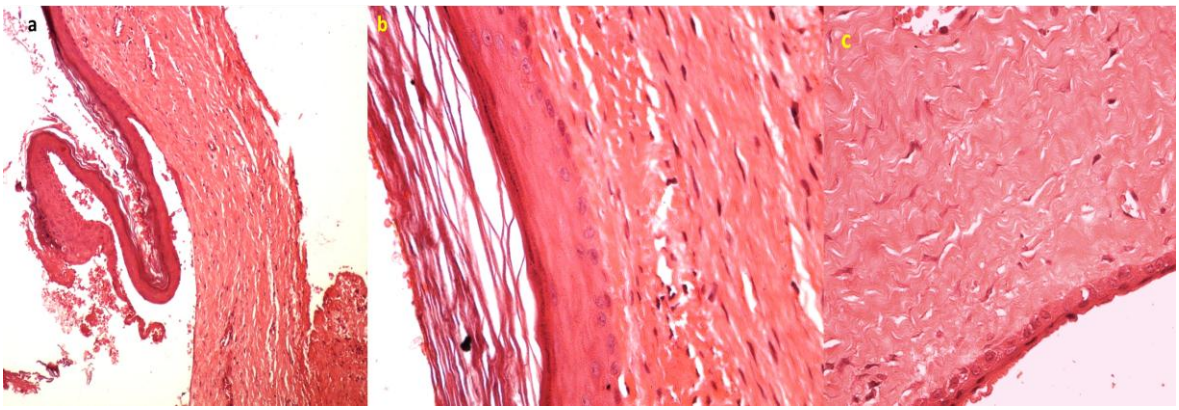

Figure 2: The hematoxylin and eosin stained photomicrograph at low power view showing an orthokeratinized odontogenic cyst exhibiting a uniform orthokeratinized stratified squamous epithelial lining (a). The high power view showing an onion skin–like surface orthokeratinization with prominent granular cell layer, and flattened basal cells (b) and relatively thin epithelial lining several places (c).

Features	Odontogenic Keratocyst	Orthokeratinized Odontogenic Cyst
Incidence	3 to 11% of all odontogenic cysts	7 to 17% of all odontogenic cysts
Clinical Features **Age**	10 to 40 yrs.	Predominantly in young adults
Sex	Slight male predilection	2:1 male to female ratio
Site	Ascending ramus and posterior body of mandible	Posterior region of the mandible
Radiology	Grow in antero-posterior direction within the medullary cavity of the bone without causing obvious bone expansion	No clinical distinct features
	Well defined radiolucent area with smooth and often corticated margins. Large lesions may appear multilocular. Resorption of the roots of the erupted tooth is less common	Unilocular, Occasionally multilocular
Histopathology **Basal cells & Nuclei**	Columnar; Hyperchromatic; Palisading and Polarizied	Cuboidal or flattened, `Less tendency for hyperchromatism, centrally placed nuclei
St. Granulosum	Not Prominent	Prominent
Keratinized layer	Parakeratinized showing surface corrugations	Orthokeratinized layers were relatively thick and onion skin like.
Satellite cysts	Can be seen in the fibrous wall	Not seen
Recurrence rate	High (28%)	Low (4%)
Syndromic Associations	Associated with nevoid basal cell carcinoma syndrome	No association

Table1: Comparative features of Odontogenic Keratocyst & Orthokeratinized Odontogenic Cyst

The margins of all cases of the two small case series of OOCs, reported so far, were well defined, whereas over a third of KCOTs first presented were poorly defined. KCOT first presented with swelling significantly more frequently than OOCs, but OOCs were significantly more associated with unerupted teeth.[9]

Histologically there are several striking differences between the epithelial lining of Orthokeratinized and parakeratinized cysts. The typical KCOT exhibits a highly cellular parakeratinized epithelial lining with surface corrugations and a palisaded layer of basal cells. In contrast the OOC lacks these features and instead the thin, uniform, orthokeratinized lining epithelium is characterized by onion-skin–like luminal surface keratinization, prominent stratum granulosum and low cuboidal or flattened basal cell layer with minimal tendency for nuclear palisading[2] (Table 1).

Enucleation with curettage is the usual treatment for orthokeratinized odontogenic cysts. Recurrence has rarely been noted, and the reported recurrence rate is only 4% in OOC as compared to a high i.e. 28% in KCOTs. The margins of all cases of the two small case series of OOCs, reported so far, were well defined, whereas over a third of the KCOTs were poorly defined.[7]

Conclusion

The significant clinicopathologic differences between orthokeratinized and parakeratinized odontogenic cysts make it imperative that the orthokeratinized cyst be recognized as a distinct entity. Historically, these cysts have been diagnosed as odontogenic keratocyst. Therefore, in order to avoid confusion, it is suggested that they be called Orthokeratinized Odontogenic Cysts.

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