Case Report

Orthokeratinized odontogenic cyst with calcification: A rare case report of a distinct entity

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Abstract

Orthokeratinized odontogenic cyst (OOC) is a relatively rare odontogenic cyst, distinct from odontogenic keratocyst (OKC). In the 4th edition of WHO Classification of Head and Neck Tumors (2017), OOC has been included as a separate entity in the category of developmental odontogenic cysts. It presents as a unilocular radiolucent lesion in the posterior mandible and is frequently related to impacted teeth, thus mimicking dentigerous cyst. Due to low local aggressiveness and no association with nevoid basal cell carcinoma syndrome, it does not show tendency to recur. When compared to OKC, OOC exhibits substantial number of differences with respect to clinical, pathological and behavioral features and treatment modalities. Hence, recognizing OOC as unique lesion is mandatory to avoid unnecessary overtreatment. This paper aims to report a rare case of OOC associated with impacted tooth, showing calcifications and emphasizes its differences from OKC. Furthermore, the recent concepts about OKC and OOC are discussed.

Keywords: Calcification, female, mandible, odontogenic keratocyst, orthokeratinized odontogenic cyst

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INTRODUCTION

Orthokeratinized odontogenic cyst (OOC) is an uncommon lesion of the jaws and represents 7%–17% of all keratinizing jaw cysts.[1] Although previously thought to be a variant of OKC, OOC is now recognized as a separate entity.[2] OOC usually occurs in 3rd to 4th decade and shows male predominance. It presents as solitary, unilocular radiolucency in the posterior mandible with cortical expansion.[2-4] In contrast, the present case is unique as it occurred in an 18-year-old female patient and showed calcification. Only one case has been reported till date with calcification.

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CASE REPORT

An 18-year-old female patient presented with pain and swelling in the left back region of the lower jaw of one-month duration. Extraorally, the swelling extended from the left angle of the mandible to ramus region and measured 2 cm × 1 cm in dimension. On intraoral examination, 37 was found missing, with partially erupted 38. A diffuse swelling in relation to 37 and 38 was seen obliterating the buccal vestibule. The overlying mucosa was intact. The swelling was hard and nontender on palpation. Expansion of buccal cortical plate was evident. Orthopantomograph showed a well-defined unilocular radiolucent lesion distal to crown of impacted 37 along with expansion and displacement

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of mandibular canal. Tooth 38 was displaced superiorly. However, there was no resorption of 37 and 38 [Figure 1].

Based on clinical and radiographic findings, provisional diagnosis of dentigerous cyst in relation to 37 was rendered. The lesion was treated conservatively by complete enucleation along with removal of 37 and 38. Two-year follow-up was uneventful.

Macroscopic examination of submitted tissue showed thin grayish-white pieces of cystic lining which was partly attached to impacted 37. Hematoxylin and eosin stained sections under microscopic examination showed, 4–6 layers thick, uniform cystic epithelial lining of stratified squamous epithelium with prominent orthokeratinization. Flat to cuboidal basal cells were seen and a prominent granular cell layer was evident [Figures 2 and 3]. The lumen of the cyst was filled with keratin filaments at places. The epithelial and connective tissue interface was flat. The cyst wall showed patchy areas of inflammation along with single concentric mass of calcified material [Figure 4]. Multiple serial sectioning of the tissue did not reveal any features of OKC such as parakeratinization and basal palisading.

Based on these histopathological features, a final diagnosis of OOC was established.

DISCUSSION

Wright in 1981 clearly identified OOC as an orthokeratinized variant of odontogenic keratocyst (OKC). In 2005, WHO classified OKC as keratocystic odontogenic tumor (KCOT) and stated that cystic jaw lesions that are lined by orthokeratinizing epithelium do not from a part of the KCOT.[5] However, the 4th edition of WHO Classification of Head and Neck Tumors (2017) has shifted the so-called KCOT back into the cyst category as odontogenic cyst (OKC).[6,7] This decision has been taken because of lack of evidence to justify the continuation of KCOT as a tumor. In addition, considering the low aggressiveness, lack of recurrence and no association with nevoid basal cell carcinoma syndrome, OOC is now

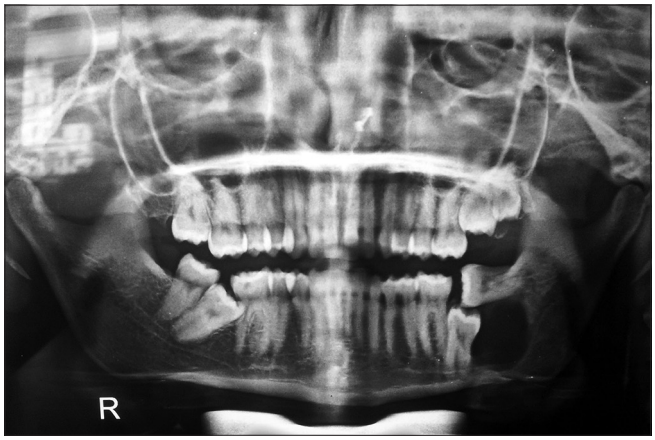

Figure 1: Radiographic image showing well-defined radiolucency in relation to 37

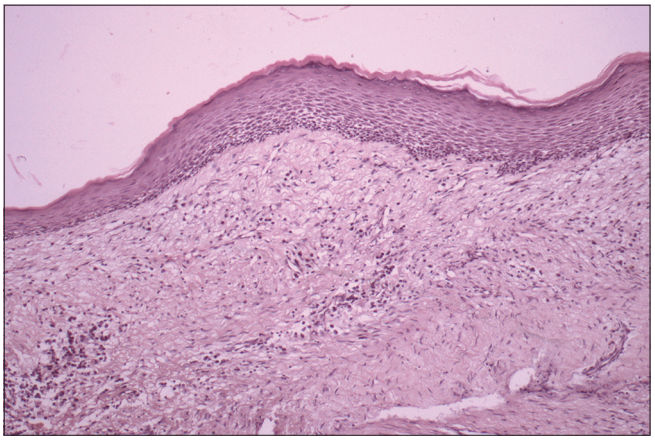

Figure 2: Histopathologic image showing cystic epithelium lined by orthokeratin (H & E, ×4)

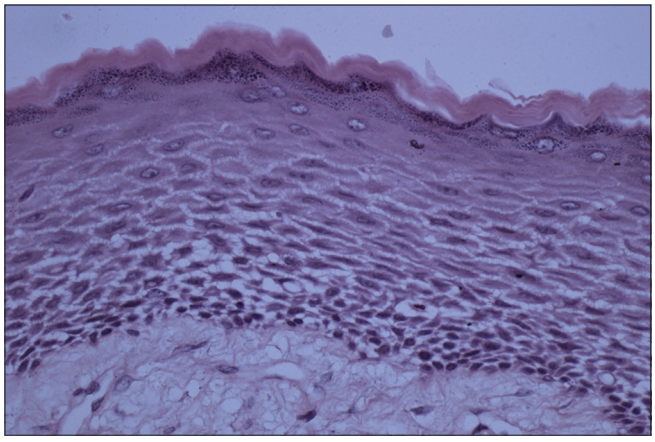

Figure 3: Histopathologic image showing prominent granular cell layer and lack of palisading of basal cells (H&E, x40)

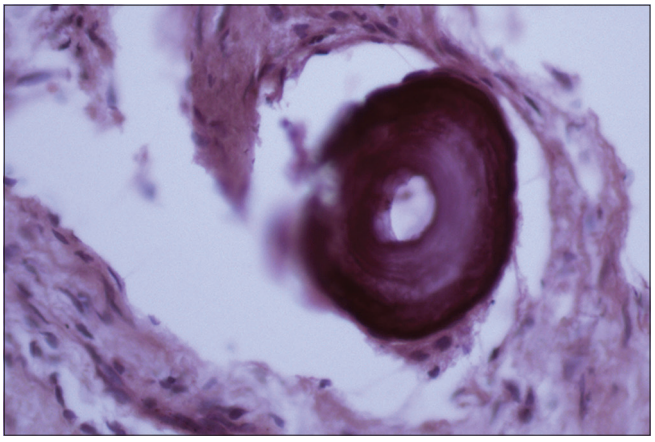

Figure 4: Histopathologic image showing hematoxophilic foci of calcification in the connective tissue wall (H & E, ×100)

classified under developmental odontogenic cysts as an independent entity.[6,7]

Although the histogenesis of OOC is unclear, it is suggested that (a) OOCs could be dentigerous cysts with orthokeratinization as reduced enamel epithelium after tooth formation has the capacity to keratinize under appropriate stimuli, (b) OOCs may represent central dermoid/epidermoid cysts due to similar histological features, (c) OOCs may arise from oral epithelium under the influence of dental papilla or only the oral epithelium and (d) sequestration of the stomodeal ectoderm into the developing jaw during embryogenesis may be a source of origin of OOC.[2,5,8]

Clinicopathological studies have reported that the prevalence of orthokeratinized variant ranges from 3.3% to 12.2%.[5] However, the exact incidence of OOC is not specified, as majority of them were previously considered to be a variant of OKC. OOC is frequently seen in the 3rd–4th decade of life and is predominantly seen in males.[2] On the contrary, our case of OOC was found in female patient of the second decade. A systemic review on OOC among global groups suggests that female predominance in the second decade may be linked to the menarche.[9]

OOC usually presents as a solitary lesion and shows predilection for posterior mandible. OOCs are usually asymptomatic.[5] However, cases with pain and swelling have been reported including the present case. Radiographically, they present as well-defined unilocular radiolucencies and 46.5%–75% of cases are associated with impacted teeth.[2] Multilocular and bilateral cases also have been reported.[2,8] Most of the OOCs show striking feature of buccolingual expansion.[4,10] This helps to differentiate it from OKC which shows anteroposterior growth pattern without causing obvious bone expansion.[1] Although displacement of adjacent teeth may be seen, root resorption is not a feature in OOC.[10] The present case also was seen in posterior mandible, showed bone expansion and was associated with impacted tooth. Literature shows different rare forms of OOC such as peripheral OOC, OOC associated with other cysts and tumors and bilateral OOC.[2] OOC associated with complex odontoma also has been reported.[3]

Keeping in mind the above-mentioned clinical presentation of OOC, the differential diagnosis of OOC should include OKC, dentigerous cyst, radicular cyst and other jaw lesions with similar features.

Histopathologically, OOC is lined by 5–10 cells thick stratified squamous epithelium with onionskin-like luminal surface orthokeratinization. It is characterized by prominent stratum granulosum, an intermediate layer of polyhedral cells with eosinophilic cytoplasm and low cuboidal to flattened basal cell layer with little tendency to nuclear palisading.[10] As opposed to this, typical OKC lining is highly cellular, parakeratinized with surface corrugation and a palisaded, hyperchromatic basal cell layer. OOCs lack satellite cysts and basal cell budding as seen in OKC. The lumen of OOC often shows more abundant leafy keratin flakes whereas the parakeratin squames are very sparse in OKC.[5] Along with these features, our case also showed the presence of calcification within the connective tissue wall which is reported in only one case previously.[8] This calcification can be dystrophic, as part of the lesion showed inflammatory cells. The other possibility can be from odontogenic rests. As the lesion in the present case was of short duration, calcifications could not be appreciated radiologically. The significance of the presence of calcifications lies in the fact that the tissue should be examined carefully with serial sections as cases of OOC associated with odontomes have been reported in the literature.

Studies have suggested that OOC shows low expression of Ki-67 and p53 and negative expression of bcl-2. Immunoprofiling of epithelial lining and capsule using cytokeratins and extracellular matrix proteins have shown that, in contrast to OKC, OOC has less proliferative activity and is a more organized cyst.[5,11,12] Ultrastructurally, OOC shows loose attachment between superficial shreds of orthokeratin and a compact layer of underlying keratin. In contrast, OKC shows the presence of cytoplasmic interdigitations and desmosomal junctions which give rise to the complex surface morphology.[13] The histopathologic differential diagnosis should also include intraosseous epidermoid cyst. However, the absence of skin appendages in OOC is the differentiating feature.[14]

Considering the low recurrence and lack of aggressiveness, OOCs should be treated by conservative approach with complete enucleation along with the removal of impacted tooth.[5,10,15] However, thorough sampling of specimen is required as the presence of parakeratinization and polarization should lead to the lesion being diagnosed as OKC that requires aggressive treatment.[5] The recurrence rate OOC is 2.2% which is far low than that of KCOT (recurrence rate 42.6%).[3,15] Table 1 summarizes the differences between OOC and OKC.

CONCLUSION

As there are considerable differences with respect to pathological and behavioral features among OOC and OKC, OOC should be considered as a unique lesion

Table 1: Comparison of orthokeratinized odontogenic cyst and odontogenic keratocyst

Feature[2,3,7,11,12]	OOC	OKC
Age	3-4th decade	2-3rd decade
Gender	Male > female	Male > female
Association with impacted teeth (%)	46.5-75	25-40
Association with NBCCS	No	Yes
Multiplicity	Rare	Seen
Location	Posterior mandible	Posterior mandible
Radiographic features[2,3,7,11,12]	**Usually unilocular**	**Unilocular or multilocular**
Root resorption	Not seen	Seen
Bone expansion	Seen	Not seen
Histopathologic feature[2,3,7,11,12]	**Orthokeratinized**	**Parakeratinized**
Surface corrugation	Not seen	Seen
Basal cell palisading	Not seen	Seen
Basal cell budding	Not seen	Seen
Recurrence rate (%)	2.2	42.6
Treatment	Simple enucleation	Wide excision

NBCCS: Nevoid basal cell carcinoma syndrome, OOC: Orthokeratinized odontogenic cyst, OKC: Odontogenic keratocyst

of the jaws. This has been reflected in the recent WHO classification that includes OOC as a distinct cyst. Multiple sections of the specimen are necessary to rule out areas of parakeratinization as such finding may point toward the diagnosis of OKC. The frequent association of OOC with impacted tooth necessitates it to be considered in the differential diagnosis of radiolucent lesions that occur in relation to impacted teeth. A thorough knowledge about OOC among the clinicians and pathologists is essential to prevent unnecessary morbidity. The presence of calcifications in OOC needs to be further evaluated as only two cases including our case have reported such a finding.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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