

Orthokeratinized Odontogenic Cyst of the Mandible: Case Report with Unusual Histopathologic Features with Review

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DOI: <http://dx.doi.org/10.13005/bpj/662>

(Received: August 15, 2015; accepted: September 20, 2015)

ABSTRACT

Orthokeratinizedodontogeniccysts(OOC's) are relatively uncommon developmental cysts lined with orthokeratinized epithelium consisting of prominent granular layer and a basal layer of low cuboidal flattened cells that show no tendency for nuclear palisading. OOC's has been considered a distinct entity from odontogenickeratocyst since they exhibit a less aggressive behavior and a very low recurrence rate. This report describes a 27 year old female patient who presented with a cyst with unique histopathological features. The purpose of this article is to present a case of OOC in the posterior mandible and also emphasize on the need for differentiating it from keratocysticodontogenictumor(KCOT) with a comprehensive literature review.

Key words: Odontogenic cyst, Orthokeratinized odontogenic cyst, Keratocysticodontogenic tumor.

INTRODUCTION

Orthokeratinizedodontogenic cyst (OOC) refers to an odontogenic cyst that presents microscopically with an orthokeratinized epithelial lining. It was earlier referred to as orthokeratinized variant of odontogenickeratocyst (OKC) and does not denote to any specific clinical type of odontogeniccyst. Ortho keratinized odontogenic cyst represents about 7-17% of all keratinizing jaw cysts¹ Orthokeratinizedodontogenic cyst is considered to be clinicopathologically different from parakeratinizedodontogenickeratocyst. (1). It shows considerable differences in cytokeratin, epithelial membrane antigen (EMA) and carcinoembryonic antigen (CEA) immune reactivity from the parakeratinized OKC. These differences also prove that the orthokeratinized variety or OOC shows less aggressive behavior and should be considered as

a different entity from the parakeratinized odontogenickerato cyst.

Case report

A 27 year old female patient visited the dentist in a private dental office complaining of pain and food impaction in the left lower back tooth region for the past 2 weeks along with a hole in the gum in the same region for the past 6 months. The hole was progressively increasing in size. On general examination patient was of normal built and moderately nourished with no relevant past medical history. Examination of the oral cavity revealed a defect in the gingival distal to 37. 38 tooth was missing. Radiological examination presented a well definedunilocular radiolucency measuring 2.5cm *2.5cm which was well circumscribed with a sclerotic border. Surgical enucleation under general anaesthesia was done

and the tissue specimen was sent for histopathological examination. The patient was lost to follow up. Differential diagnosis of ameloblastoma, KCOT and dentigerous cyst was given.

A single soft tissue specimen was fixed in 10% formalin. The specimen measured about 2*2.5cm with a cystic cavity and was soft in consistency (fig 1). Microscopic examination revealed a cystic epithelial lining which was orthokeratinized and of 4-6 cell layer thickness in most of the areas with prominent granular cell layer (fig 2) along with corrugated keratin flakes (fig 3). The basal cells in most of the areas are prominent and arranged in a single layer with nuclear palisading. There were certain areas that revealed thin rete ridges (fig 4). There were areas in the section showed hyperplastic epithelium which was of stratified squamous type with prominent granular

cell layer and orthokeratin with broad rete pegs. (fig 5). The underlying connective tissue capsule was made up of mature collagen bundles with a few areas showing sub-epithelial foci of chronic inflammatory cells where the overlying epithelium exhibited detachment at the basal layer (fig 6). In one area of the section the transition from hyperplastic epithelium to odontogenic epithelium was observed. (fig 7)

Orthokeratinized odontogenic cyst (OOC) is a developmental odontogenic cyst which has an uncommon occurrence⁶. OOC was first described in 1927 as orthokeratinised variant of odontogenic keratocyst which is now renamed as keratocystic odontogenic tumor (KCOT)^{4,5}. WHO in 2005 classified the parakeratinized variant as KCOT and since then OOC forms a separate entity^{5,9}. The



Fig.1. Gross specimen of OOC

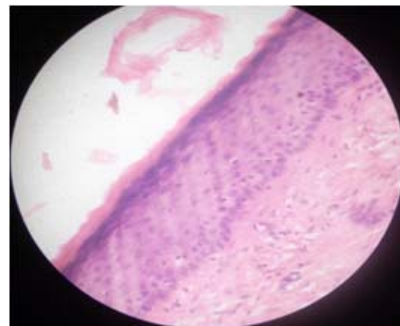


Fig.2. Area of cyst showing orthokeratinized epithelium with prominent granular cell layer and basal cell palisading. 10 X

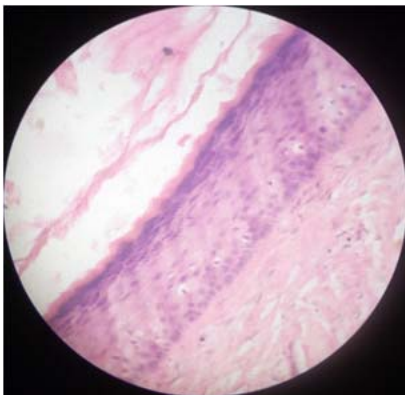


Fig. 3 10X corrugation of OOC

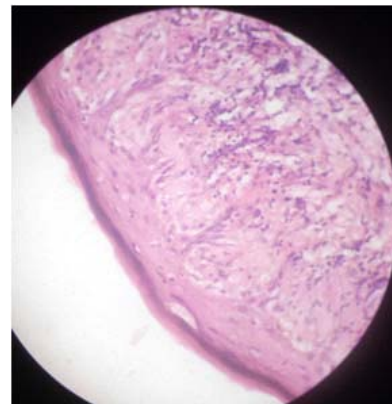


Fig.4. Areas showing thin rete ridges 10 X

occurrence of KCOT is 8 times more common than that of OOC as stated by the WHO. There is considerable difference between OOC and other developmental odontogenic cysts such as dentigerous cyst and OKC⁷. Apart from OKC's there are many other odontogenic cyst which produce orthokeratin as well⁸.

It usually occurs in the third and fourth decades and shows male predilection as reported by Mac Donald *et al.*, and Li *et al.*,^{11,12,13}. OOC was found to be occurring in about 5.2% to 16.8% of cases that have been previously diagnosed as odontogenic keratocyst in different case series¹². A female in the second decade was affected in the present case which is in contrast to what was stated in the literature. Mac Donald *et al.*, put forth that if the lesion appears in a female it is usually in the second decade of life¹³ as like in our case. The

possible reason could be due to the hormonal influence due to the onset of menarche and the fluctuations associated with it which is commonly seen in the early twenties⁴.

Clinically OOC presents as a painless slow growing swelling in the posterior mandible. Sometimes the swelling may be associated with pain. Radiographically it appears as a well defined unilocular or multilocular radiolucent lesion of size in the range of 2-7cm with a mean of 4.8cm [11,12]. Histopathological findings of OOC include a cystic cavity that is lined by a uniform 4-8 cell layer thick regular stratified squamous epithelium, with a basal layer of flat to cuboidal cells that have hyperchromatic nucleus arranged in a palisading pattern. The intermediate layer is made up of polyhedral cells with eosinophilic cytoplasm. The granular layer is usually prominent with keratohyaline granules and the superficial layer is made up of a thick layer of orthokeratin. On the other hand, KCOT shows 5-10 cell layers of thick epithelium with the basal cells having an elongated nucleus and the presence of a characteristic superficial corrugated layer of parakeratin^{3,5}

In the case reported the typical histopathological features of OOC was observed providing an easy diagnosis. But there were certain areas in the section which was unusual and caused a diagnostic dilemma. The area of hyperplastic stratified squamous orthokeratinized epithelium with no inflammatory cells in the underlying connective tissue was atypical. The epithelium resembled the surface epithelium in all its characteristics. The chances of surface epithelium

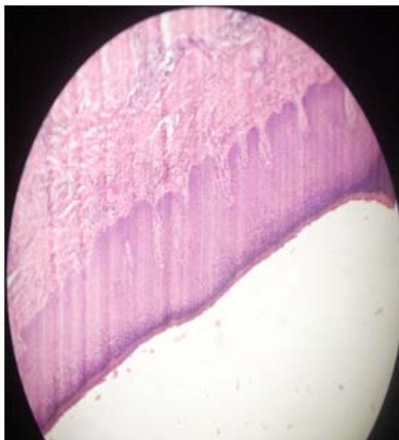


Fig. 5. Areas of cyst showing hyperplastic epithelium with broad rete peg - 4 X

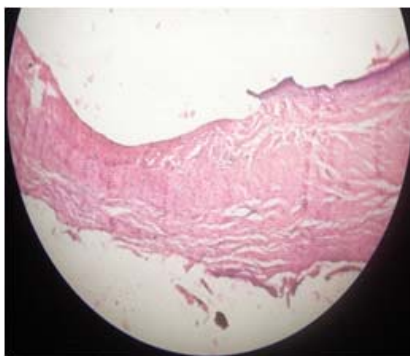


Fig. 6. 4X - discontinuity of epithelium

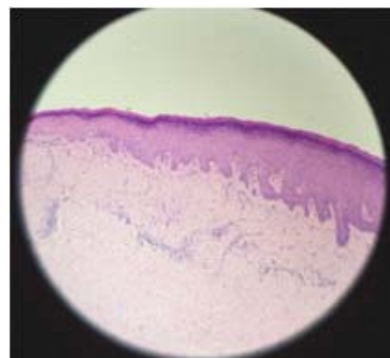


Fig. 7. Transitional area of the epithelium

getting incorporated into the cystic lining was remote as the radiograph revealed a well defined lesion with a sclerotic border and the cyst was enucleated in toto and there was an overlying defect in the gum clinically. The epithelium did not show any dysplastic changes as against cases reported in the literature where there was dysplastic changes in the epithelial lining of the cyst¹⁶.

In some areas there was the presence of thin rete ridges along with the OOC epithelial lining. In the present case there seems to be the presence of 2 different epithelial linings with a smooth transition. One side there is an OOC lining and on the other side epithelium resembling the surface epithelium. Variations are common in OOC. Kasat *et al.* reported a case of OOC with multiple supernumerary teeth[14]. Rare cases of bilateral involvement of the mandible, dysplastic changes in the epithelial lining of the OOC, association of the OOC with the calcifying odontogenic cyst and complex odontoma have additionally been reported^{15,16,17}. Bolbaran *et al.* reported a case of OOC in a patient with nevoid basal cell carcinoma syndrome¹⁸.

Though variations have been reported this type of unusual histopathological presentation has not been reported. This kind of presentation warrants for further research in determining the direction of the transition (odontogenic to non

odontogenic or vice versa) and the possible lesions that can arise and the precautions that needs to be taken. The recurrence rate reported is about 0-4% after complete enucleation during an average period of 6.4-7.8 years of follow up^{9,11,13}. This case could not be followed up as the patient did not report back for review.

CONCLUSION

OOC is a distinct entity which exhibits typical clinical, histopathological features and a biological behavior that varies considerably from KCOT. This cyst has a better prognosis and lower recurrence rate compared to the aggressive KCOT. Usually the histopathological features are typical and enable the pathologist to give a diagnosis. Sometimes such unusual features may be present. These have to be carefully analyzed before arriving at a diagnosis. It is imperative that such areas have to be explained as it can have a bearing on the treatment aspect. Many a times developmental odontogeniccyst have got secondarily infected leading to threatening complications, malignant lesions have been present in association with an odontogenic cyst. So multiple sections have to be made and carefully studied. In the present case due to the presence of unusual features we made multiple sections and then arrived at a diagnosis of OOC with atypical findings. Malignancy was ruled out as there was no dysplastic features.

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