

Orthokeratinized Odontogenic Cyst

Niharika Swain, Shilpa Patel, LS Poonja, Jigna Pathak, Kamlesh Dekate

ABSTRACT

Orthokeratinized odontogenic cyst (OOC) is a developmental cyst of odontogenic origin and was initially defined as the uncommon orthokeratinized variant of odontogenic keratocyst (OKC). However, recently World Health Organization has designated OOC as a distinct clinicopathologic entity as it has peculiar clinicopathologic aspects when compared to other developmental odontogenic cysts, especially OKCs. The orthokeratinized odontogenic cyst is histologically characterized by a thin, uniform, epithelial lining with orthokeratinization and a subjacent prominent granular cell layer. The purpose of the article is to present a case of OOC arising in the anterior mandible, an unusual site for the lesion and also highlights the importance of distinguishing it from the more commonly occurring keratocystic odontogenic tumor (KCOT).

Keywords: Odontogenic cyst, Orthokeratinization, Parakeratinization.

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INTRODUCTION

Orthokeratinized odontogenic cyst (OOC), a developmental cyst was first described as a dermoid cyst by Schultz in 1927. In 1945, Philipsen considered this entity as a variant of odontogenic keratocyst (OKC). OOC gained individuality in 1981 as Wright described its clinicopathological features after observing for 30-year period.¹ As recent World Health Organization classification (2005) has considered the odontogenic keratocyst as a neoplasm and designated it as keratocystic odontogenic tumor (KCOT), it is necessary that both oral surgeons and pathologists should possess a thorough knowledge of the clinicopathologic differences between the more aggressive KCOT and the less aggressive OOC so that patients receive the most appropriate treatment.²

Here, we report a case of orthokeratinized odontogenic cyst in 42-year-old male patient.

CASE REPORT

A 42-year-old man presented with a 6-month history of discomfort in the anterior region of mandible following extraction of regional teeth. On intraoral examination, extraction wound of 41 was observed (Fig. 1). No obvious swelling was observed in the area of concern. On palpation, slight obliteration of the buccal vestibule extending from

35 to 45 with areas of fluctuation was assessed. Orthopantomograph view showed well-defined unilocular radiolucency with sclerotic margins extending from 37 to 47 and an impacted 33 within (Fig. 2). After clinical and radiographic evaluation, a preliminary diagnosis of odontogenic keratocyst was made. An incisional biopsy was performed under local anesthesia. Tissue sample was submitted for histopathological examination. Microscopic examinations showed a thick cyst wall, lined by the orthokeratinized squamous epithelium (Fig. 3). The flattened basal cell layer lacked the palisading and the prominent granular cell layer was apparent (Fig. 4). The lesion was finally diagnosed as OOC. Patient has undergone segmental mandibulectomy followed by reconstructive surgery.



Fig. 1: Intraoral view



Fig. 2: Orthopantomograph view showed well-defined unilocular radiolucency with impacted 33

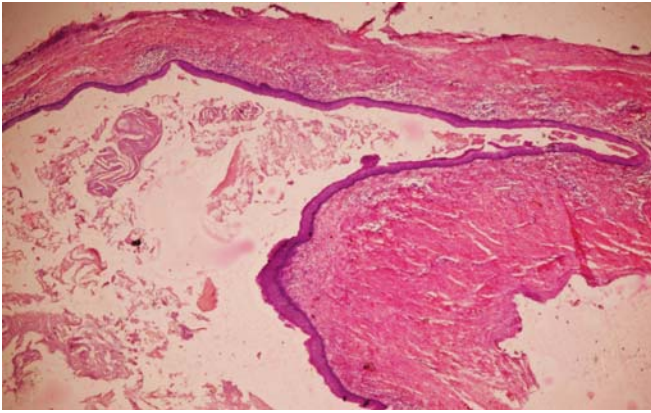


Fig. 3: H & E stained section shows orthokeratinized squamous epithelial lining (4x)

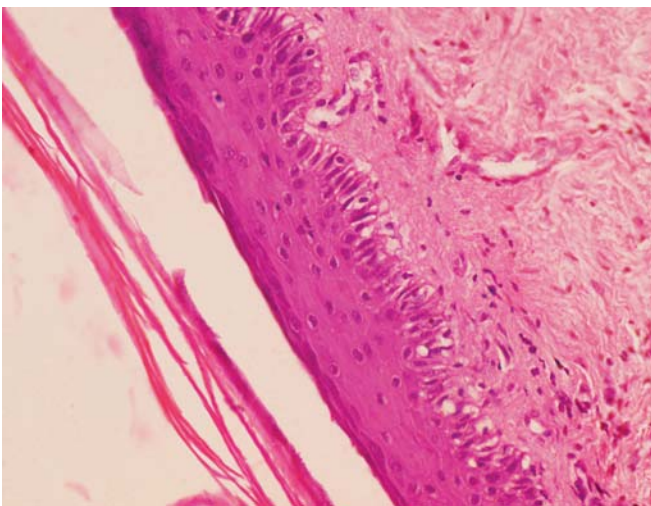


Fig. 4: H & E stained section shows prominent granular cell layer (40x)

DISCUSSION

Despite some similarities in microscopic features of OKC and OOC, i.e. equal epithelial lining thickness, stratified squamous epithelium with keratinization, these two cysts are different in various aspects. Previous documented series have reported the varied incidence of OOC ranging from 5.2 to 16.8% among cases which had been previously coded as KCOT. Orthokeratinized odontogenic cyst has a male predominance with a male to female ratio of 2.59:1, which is higher than that reported for KCOT (ranging from 1.42:1 to 1.76:1). The mandible was far more commonly involved than the maxilla with the most common location being the mandibular molar and ramus region. The mandible-maxilla ratio of OOC is 9.17:1, higher than that reported for KCOTs (ranging from 2.08:1 to 4.4:1). The size can vary from less than 1 cm to large lesions greater than 7 cm in diameter.^{3,4} Radiographically, OOCs more frequently presented as unilocular radiolucencies (87.0%) in comparison with KCOTs (69.4 to 73.3%). OOC is more often associated with an impacted tooth (60.8%) than the OKC (7-48%), and the

OKC recurred in at least 42.6% compared with only a 2.2% recurrence rate for the OOC. In addition, OKC, unlike OOC, may be a part of nevoid basal cell carcinoma syndrome (NBCCS) and may transform into a neoplasm.^{5,6} Histologically, the basal cells of the OOC are much less developed than those in the OKC. They tend to be cuboidal or squamous and show little tendency to be polarized or palisaded. In addition, OOC has a luminal surface of orthokeratin and a well-developed granular layer.⁷

Recent immunohistochemical studies that compared OOCs with parakeratinized OKC have shown distinct differences in the expression of Ki-67 proliferative index, p53, p63 and bcl-2. Reduced expression of all these markers in OOC reflect the variations in epithelial cell maturation and proliferation between the two types of lining epithelia; namely, those of OOC seem to assume a different cell differentiation and exhibit a lower cellular activity than those of KCOT. Wysocki and Sapp showed that there are distinct ultrastructural differences between the OKC and OOC. The surface morphology of the OOC is more uniform and is entirely covered with a layer of keratin squames. There is an increase in tonofilaments as the cells mature, and the granular cell layer consists of a compact layer of degenerated cells that contain large amounts of keratohyaline granules. The luminal surface consists of a compact layer of shreds of orthokeratin.^{8,9}

Enucleation with curettage is the usual treatment for OOCs.¹⁰ Recurrence has rarely been noted. In the present case, more aggressive treatment was carried out as the extension of lesion had included the lower border of mandible.

CONCLUSION

Through the experience of the present case, we reconfirmed the importance of precise clinicopathological observation of an individual case. As in the present case, the size and location of the lesion did not corroborate with the usual clinical finding of OOC, the histopathological evaluation was helpful in proper treatment planning. Though OOC exhibits a number of distinctive clinical, pathologic, and behavioral features that varied substantially from KCOTs, variation in clinical presentation of OOC requires meticulous review of literature.

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ABOUT THE AUTHORS

Niharika Swain (Corresponding Author)

Senior Lecturer, Department of Oral Pathology, MGM Dental College and Hospital, Navi Mumbai, Maharashtra, India
Phone: +919324701036, e-mail: niharikadec30@gmail.com

Shilpa Patel

Professor and Head, Department of Oral Pathology, MGM Dental College and Hospital, Navi Mumbai, Maharashtra, India

LS Poonja

Professor, Department of Oral Pathology, MGM Dental College and Hospital, Navi Mumbai, Maharashtra, India

Jigna Pathak

Professor, Department of Oral Pathology, MGM Dental College and Hospital, Navi Mumbai, Maharashtra, India

Kamlesh Dekate

Reader, Department of Oral Pathology, MGM Dental College and Hospital, Navi Mumbai, Maharashtra, India