

# Orthokeratinized odontogenic cyst masquerading as dentigerous cyst

Devi Charan Shetty, Ajit Singh Rathore, Anshi Jain, Natasha Thokchom, Neha Khurana

Department of Oral and Maxillofacial Pathology and Microbiology, I.T.S. CDSR, Muradnagar, Ghaziabad, Uttar Pradesh, India

## ABSTRACT

The orthokeratinized odontogenic cyst (OOC) is a rare developmental odontogenic cyst that has been considered as a variant of the keratocystic odontogenic tumor until Wright (1981) defined it as a different entity. Recognition of OOC as a unique entity has long been due, yet its inexplicable clinical, radiographic presentation resembling dentigerous cyst due to its association to an impacted tooth, and its histological features makes it rather perplexing. This is the report of a case of OOC in relation to an impacted maxillary canine and its immunohistochemical analysis with Ki-67.

**Key words:** Dentigerous cyst, Ki-67, orthokeratinized odontogenic cyst

**Submission:** 03-02-2016 **Accepted:** 27-05-2016

## INTRODUCTION

Orthokeratinized odontogenic cyst (OOC) is a relatively uncommon developmental cyst comprising about 10% of cases that had been previously designated as odontogenic keratocysts (OKCs).<sup>[1]</sup> Schultz in 1927 was the first to describe it as an orthokeratinized variant of OKC and further Wright in 1981 termed it as “orthokeratinized variant of OKC,” and defines it as an independent clinical entity. Lie *et al.* is in 1998 only when he suggested the term OOC, which is now the most accepted terminology.<sup>[2]</sup>

OOC has specific histopathological features and clinical behavior. Histogenesis may be from the remains of the dental lamina or from the basal cell layer of the oral mucosal epithelium.<sup>[3]</sup> These cyst can grow large in size causing cortical expansion and presents as a swelling, along with pain, although in most cases, it can be detected incidentally during

a radiographic examination.<sup>[2]</sup> Radiographically, OOC is seen as a well-circumscribed, unilocular, or multilocular radiolucency and rarely associated with an unerupted tooth.<sup>[4]</sup>

OOC surrounds the crown of an unerupted tooth may be defined as a cyst with typical histology of an OKC.<sup>[4,5]</sup> OOC are relatively uncommon accounting 25–40% of all the OKCs.<sup>[6]</sup> An impacted tooth and its dental follicle may affect the occurrence and proliferation of an adjacent keratinizing cyst.<sup>[7]</sup>

Here, we report a case of orthokeratinized OKC, which showed the features of keratinizing dentigerous cyst in the incisional biopsy.

## CASE REPORT

A 50-year-old male reported with a complaint of swelling in the right maxillary anterior region extending from 11 to 15 from last 6 months. On intraoral examination, the swelling was oval shaped with smooth margins, soft in

Address for correspondence: Dr. Anshi Jain,  
Department of Oral and Maxillofacial Pathology and Microbiology,  
I.T.S. CDSR, Muradnagar, Ghaziabad, Uttar Pradesh, India.  
E-mail: dranshijain@gmail.com

Access this article online	
Quick Response Code:	Website: www.ijabmr.org
	DOI: 10.4103/2229-516X.192597

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

**For reprints contact:** reprints@medknow.com

**How to cite this article:** Shetty DC, Rathore AS, Jain A, Thokchom N, Khurana N. Orthokeratinized odontogenic cyst masquerading as dentigerous cyst. *Int J App Basic Med Res* 2016;6:297-9.

consistency, nonindurated, and nontender. Radiographically, radiolucency was seen in relation to an impacted I3 and a supernumerary tooth adjacent to this radiolucent area [Figure 1].

Fine-needle aspiration cytology showed keratin-like material mixed with numerous inflammatory cells mainly polymorphonuclear leukocytes. Protein estimation of aspirated fluid was 4.6 g/dl.

The incisional biopsy was performed, and histopathological diagnosis was suggestive of keratinizing dentigerous cyst.

Surgical excision of the lesion was done followed by curettage. A tissue specimen measuring 33 mm × 22 mm × 20 mm in size with multiple small bits of tissue was evaluated for histopathological examination [Figure 2].

On microscopic examination, an orthokeratinized epithelial lining of 2–6 cell layers thickness with surface corrugations, was seen along with prominent stratum granulosum and low cuboidal to flattened basal cells. The underlying fibrocellular connective tissue showed the thick bundles of collagen fibers with fibroblasts and few chronic inflammatory cells mainly lymphocytes. Subepithelial hyalinization was present at focal areas [Figure 3a].

A diagnosis of orthokeratinized OKC was made.

Immunohistochemical analysis was done using Ki-67 which showed expression in basal region and focal areas in suprabasal regions as seen commonly in cases of orthokeratinized OKC [Figure 3b].

## DISCUSSION

Incidence of OOC varies ranging from 5.2% to 16.8% few of which were previously coded as OKC.<sup>[8]</sup> Pindborg and Hansen (1963), describe “keratocyst” as any jaw cyst in which keratin was formed to a large extent lead to cases of dentigerous, radicular, and residual cyst which were included in the category of OKC. Incisional biopsy diagnosis of present case of keratinizing dentigerous cyst was similar to the study of Vuhahula *et al.* who suggested that reduced enamel epithelium can cause keratinization under opposite stimuli thus forming a true dentigerous cyst with keratinization.<sup>[9]</sup> However, other studies by Gillette and Weinmann (1958) describe these lesions are frequently misdiagnosed as dentigerous cysts with keratinized epithelial linings similar to those found in OKCs. It was suggested by Browne (1969) that this occurred when an enlarging OKC involved the follicle of an unerupted tooth and fused with the reduced enamel epithelium.<sup>[4]</sup>



Figure 1: Orthopantomogram showing radiolucency in relation to an impacted I3 and a supernumerary tooth adjacent to this radiolucent area was seen

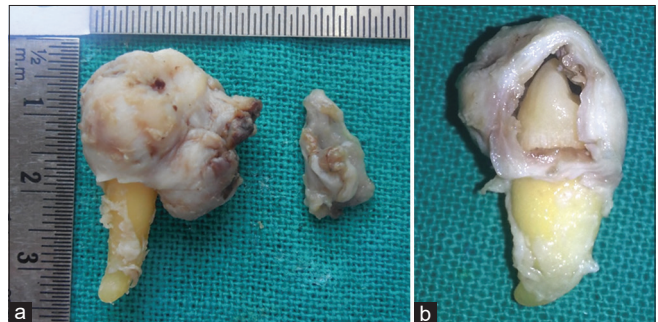


Figure 2: (a) Gross specimen measuring 33 mm × 22 mm × 20 mm in size; (b) the cyst lining enveloping the crown of the tooth at the cement-enamel junction

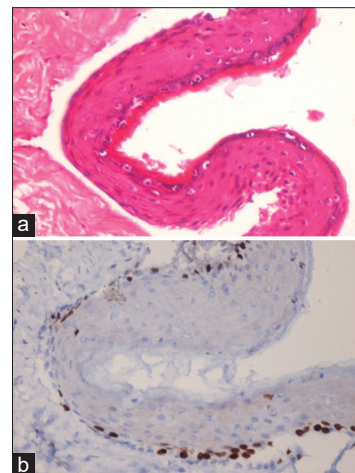


Figure 3: Orthokeratinized odontogenic cyst lining: (a) H and E; (b) Ki-67 expression in the basal layer

The World Health Organization has designated OOC as a distinct clinicopathologic entity as it has peculiar clinicoradiological and pathologic aspects when compared to other developmental odontogenic cysts, especially OKCs. Eighty-seven percent of OOC radiographically are present as unilocular radiolucencies with 60.8% of cases are associated with impacted tooth as seen in our case.<sup>[8]</sup> Histopathologically, OOC is characterized by a 4–8-cell layer thick orthokeratinized epithelial lining, with prominent granular layer and low cuboidal basal cells as seen in excisional biopsy.<sup>[8]</sup> According

to Wright (1981), OOC has uniformly thin epithelial lining with a luminal surface of orthokeratin and a well-developed granular layer. The basal cells of the OOC are much less developed. They tend to be cuboidal or squamous and show little tendency to polarize or palisade. It was recommended that care must be taken to distinguish between keratin metaplasia in otherwise nonkeratinized odontogenic cysts and the OOC.<sup>[8]</sup>

Cell proliferation molecules and related factors including Ki67, proliferating cell nuclear antigen, p53, and argyrophilic nucleolar organizer regions have previously been used to indicate biological behavior of odontogenic cysts and tumors.<sup>[10]</sup> Baghaei *et al.* in 2004 in their study showed that the mean positively stained cells/mm BM for Ki-67 antigen is more in parakeratotic OKC than that of orthokeratotic type. Findings of our case showed the positive staining for Ki-67 in the basal layer and also in suprabasal layer in focal area.

As the clinicoradiographic diagnosis of any circumscribed radiolucency around the crown of an impacted tooth is usually a dentigerous cyst, it was not unusual in the present case as well. The attachment of the cystic lining to the neck of the tooth also prompted to contemplate the origin in reduced enamel epithelium. The microscopic examination of the cyst fitted well in favor of OOC, thus, the possibility of considering this as a DC with a keratinized lining epithelium was ruled out and moreover, the mystery with regard of origin and nature of OOC was untangled.

Therefore, it was concluded that diagnosing keratinized odontogenic cyst are challenging as these cysts give an erroneous radiographic appearance of other nonkeratinizing odontogenic cyst due to its distinct location around the crown of unerupted tooth. A thorough histopathological and radiographical correlation is required along with advanced molecular analysis

to reach up to the final diagnosis of keratinized OKC which can help in understanding the pathogenesis of OOC and can help in accurate treatment planning.

### Financial support and sponsorship

Nil.

### Conflicts of interest

There are no conflicts of interest.

## REFERENCES

1. Dong Q, Pan S, Sun LS, Li TJ. Orthokeratinized odontogenic cyst: A clinicopathologic study of 61 cases. *Arch Pathol Lab Med* 2010;134:271-5.
2. González Galván Mdel C, García-García A, Anitua-Aldecoa E, Martínez-Conde Llamas R, Aguirre-Urizar JM. Orthokeratinized odontogenic cyst: A report of three clinical cases. *Case Rep Dent* 2013;2013:672383.
3. el-Hajj G, Anneroth G. Odontogenic keratocysts – A retrospective clinical and histologic study. *Int J Oral Maxillofac Surg* 1996;25:124-9.
4. Shear M, Speight P. Cyst of oral and maxillofacial region. 4<sup>th</sup> ed. Blackwell munksgaards publications. 2007. Chapter 3.
5. Altini M, Cohen M. The follicular primordial cyst-odontogenic keratocyst. *Int J Oral Surg* 1982;11:175-82.
6. Kim DK, Ahn SG, Kim J, Yoon JH. Comparative Ki-67 expression and apoptosis in the odontogenic keratocyst associated with or without an impacted tooth in addition to unilocular and multilocular varieties. *Yonsei Med J* 2003;44:841-6.
7. Tsukamoto G, Makino T, Kikuchi T, Kishimoto K, Nishiyama A, Sasaki A, *et al.* A comparative study of odontogenic keratocyst associated with and not associated with an impacted mandibular third molar. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2002;94:272-5.
8. Wright JM. The odontogenic keratocyst: Orthokeratinized variant. *Oral Surg Oral Med Oral Pathol* 1981;51:609-18.
9. Swain N, Patel S, Punja J, Dekate K. Orthokeratinized odontogenic cyst. *J Contemp Dent* 2012;2:31-3.
10. Thosaporn W, Iamaroon A, Pongsiriwet S, Ng KH. A comparative study of epithelial cell proliferation between the odontogenic keratocyst, orthokeratinized odontogenic cyst, dentigerous cyst, and ameloblastoma. *Oral Dis* 2004;10:22-6.