



Unusual Orthokeratinized Odontogenic Cyst-A Case Report and Review

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Abstract:

Odontogenic keratocyst is a developmental odontogenic cyst which has two variants parakeratinized odontogenic keratocyst (OKC) and orthokeratinized odontogenic keratocyst (OOC). Orthokeratinized odontogenic cyst is an uncommon odontogenic cyst exhibiting less aggressive nature, low recurrence, no association with Nevoid basal cell carcinoma syndrome (NBCCS) and different clinical & histopathologic characteristics compared to parakeratinized variant. Identifying OOC as a quirky lesion is obligatory to avoid inessential over treatment. The present paper strives to report and discuss the extensive growth of OOC in posterior region of the mandible associated with impacted 38 along with different concepts of histogenesis.

Keywords: *Odontogenic cyst, Orthokeratinized odontogenic cyst, Nevoid, Carcinoma Syndrome*

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1. Introduction

Orthokeratinized odontogenic cyst is an unusual developmental odontogenic cyst thought to arise from cell rests of Malassez.^[1, 2,3] Li et al proposed the term Orthokeratinized Odontogenic Cyst, most accepted terminology as yet.^[1, 2,3,4] The World Health Organization (WHO) reclassified Head and Neck tumors in 2017 and designated Kerato Cystic Odontogenic tumor (KCOT) as Odontogenic Kerato Cyst (OKC) as it is not exhibiting malignant behaviour and Orthokeratinized odontogenic cyst (OOC) is considered as an independent entity under developmental odontogenic

cysts due to its different clinical, histopathologic behaviour and often less aggressive and low recurrence.^[1,5] Here we report a case of OOC with more aggressive behaviour associated with impacted 38, it is important to distinguish OOC from OKC & Dentigerous cyst especially when it is in relation to an impacted tooth.

2. Case Description

A 40y old male patient presented with pain & swelling in lower left back tooth region since 3 months which was gradually increasing in size and attained to the present size. History revealed that pain was severe and intermittent in nature. Extra-oral examination revealed mild facial



asymmetry. On intraoral examination slight lingual expansion was seen in relation to 37. On palpation the swelling was not tender and there was no mobility of tooth associated. Orthopantomograph showed the presence of large well-defined unilocular radiolucency. Measuring approximately 3.5 x 2cm extending from distal half of the 38 and angle to the sigmoid notch of ramus of the mandible. Thinning of buccal and lingual cortical plates were observed and there was no resorption of roots in relation to 38[Figure-1].

The case was taken under GA, nasotracheal incubation done. LA infiltrated in lower buccal vestibule. An incision is made starting from crevices of 36 extending posteriorly and superiorly over anterior border of ramus of mandible. Mucoperiosteal flap raised. The cystic lining exposed and separated from underlying bone. The entire lesion was enucleated and cystic lining was collected along with 38. Carnoy's solution was placed, irrigated and closed the flap [Figure -2].

On microscopic examination, it showed a cystic lesion with thick laminated orthokeratinized, stratified squamous epithelium of 4-6 layers with prominent granular layer. The lumen was filled with keratin flakes in certain areas. Basal cell layer showed flat or low cuboidal cells without palisading pattern. The epithelial and connective tissue interface was flat and detached from the underlying connective tissue wall at areas. The cystic capsule was fibrous and showed patchy areas of inflammatory infiltrate. Based on clinical, radiographic and histopathologic findings the final diagnosis of Orthokeratinized Odontogenic cyst was confirmed [Figure 3].

Post-operative follow-up examination was done after a month. The size of the

lesion was gradually reduced to some extent [Figure 4].

3. Discussion

OOC occur at the age of 30y-40y and shows male predilection. The prime site will be posterior region of the mandible and ascending ramus, often associated with an impacted tooth (75%).^[2] OOC constitutes 7-17% of all keratinizing jaw cysts^[2,6] and 5.2 to 16.2% of all OKCs.^[1] Whilst the etiopathogenesis of OOC is quite unclear, Zhu et al suggested that OOC arise from oral epithelium under the influence of dental papilla or only the oral epithelium. While OKC may arise from the dental lamina with the presence of dental papilla required for its development.^[5, 7, 8] Dong et al suggested that OOCs showed fully differentiated, mature keratinocytes, while OKCs lacked mature keratinocytes. Also, OOCs showed a pattern of normal cellular differentiation, while OKCs showed certain alterations in the differentiation process. With regard to the capsular tissue, OOCs seemed to be more stable than the OKCs.^[9, 10] Vuhahula et al suggested a different pathogenesis for OOC associated with impacted tooth. According to him, it arises due to the pluripotentiality of odontogenic cyst epithelium. After their function of tooth development, the reduced enamel epithelium may keratinize under appropriate stimuli, thus forming a true dentigerous cyst with orthokeratinization^[5,8,11] but Vuhahula hypothesis is still questionable as the immune-histochemical expression of CK10, CK17 which were typically expressed in a keratinized epithelium were weakly expressed in dentigerous cyst. moderately, or strongly expressed in OOC and OKC. On the other hand, CK 18,19 which were the markers of non-keratinizing epithelium, were positive in dentigerous cyst and negative in OOC and OKC. The markers of



dental lamina and enamel organ, namely CK 7 and CK 13, expressed weakly in dentigerous cyst, OOC expressed CK 7 negatively and CK 13 strongly positive and OKC expressed CK7 weakly and CK13 strongly positive. This implied that OOC and OKC might have a common inception in the rests of dental lamina. Despite of substantial research on CK expression in these three developmental odontogenic cysts, nothing tangible could be illuminated, as the results were often astonishing about the origin of OOC. This has left the investigators unglued about the definitive cell of origin in the histogenesis of any of the developmental odontogenic cysts. Positive expression of K2 and LOR in OOC indicated that the cells were in a completely differentiated state, and thus not aggressive in its behavior^[10]. In present case it showed an aggressive growth, what is the reason for its extensive growth is still questionable.

Ultrastructurally the surface morphology of OOC is more uniform & is covered by a layer of keratin squames which shows loose attachment between superficial shreds of orthokeratin and a compact layer of underlying keratin. There is an increase in tonofilaments as cells mature and the granular cell consists of compact layer of degenerated cells having keratohyaline granules. In contrast OKC shows the presence of cytoplasmic interdigitations and desmosomal junctions which give rise to complex surface morphology.^[1, 6, 12] The differences are illustrated in the table below.

Table 1: Dissimilarities between Orthokeratinized Odontogenic Cyst & Parakeratinized Odontogenic Cyst

FEATURES	OOC	OKC
CLINICAL FEATURES^[1,2,4,6]		
Age	3-4 th decades	2-3 rd decades
Gender	Male predilection	Male predilection
Location	Posterior mandible and ascending ramus	Posterior mandible and ascending ramus
Associated with impacted tooth	46-75.7%	7-48%
Associated with Nevoid basal cell carcinoma syndrome.	No	Yes
Multiple cysts	Rare	Seen
Histogenesis	Remnants of dental lamina, Basal cell halfshoots, Keratinization of reduced enamel epithelium after completion of tooth development when associated with impacted tooth.	Remnants of dental lamina, Basal cell half shoots
RADIOGRAPHIC FEATURES^[1,6,7,8]		
Locularity	Usually unilocular (or) rarely multinodular (87%)	Unilocular (or) multinodular 69.4%
Root resorption	Not seen	Seen
Bone expansion	Cortical expansion	Not seen unless it is a huge lesion as it shows intra medullary expansion.
HISTOPATHOLOGIC FEATURES^[1,6,8]		
Surface corrugations	Not seen	Seen
Basal cell palisading	Not seen	Seen
Daughter cysts	Not seen	Seen
Recurrence rate%	2.2%	42.6%
Treatment	Enucleation	Wide excision
IMMUNOHISTOCHEMICAL STUDIES^[1,2,6,8,13]		
Proliferative markers –increased expression indicates higher proliferative potential		
Ki 67 ^[13]	Decreased expression (7.8%)	Increased expression (11.2%)
P53, P63	Decreased expression	Increased expression
Bcl2(Anti apoptotic marker)	Absent in basal layer	Present in basal layer- More Aggressive
Cytokeratins ^[13]		
CK 10(Early marker of keratin differentiation)	Absent	Present in upper spinous cells
CK 13	Weekly positive in surface keratinizing cells & granular cells	Positive in upper suprabasal cells and parakeratinized cells
CK17	Weakly or locally positive in keratinocytes	Strong positive (Aggressive nature) in whole epithelium, predominant in daughter cysts.
Ck 19 (Odontogenic epithelial marker)	Negative (Origin from other cell rests such as gingival cells and mucosal cells)	Intense expression
Epithelial membrane antigen	Negative	Strongly positive
Cell surface glycoproteins	Negative	Present in basal and parabasal layers
Cell migration & tumor invasion	Low expression	Intense expression
Podoplanin		
Fibronectin	Scantly scattered	Increased expression
TREATMENT^[10]		
	Simple enucleation	Peripheral osteotomy Chemical curettage Enbloc resection

4. Conclusion

Orthokeratinized odontogenic cysts presents a completely different biologic behaviour compared to OKCs. IHC showed that OKC had higher cellular proliferation and relatively low differentiation as compared with OOC, which may be the reason for aggressive behaviour of OKC. In contrast the present case showed huge lesion involving impacted tooth with no resorption of roots. Consequently, OOC can be considered as differential diagnosis of radiolucent lesions of the jaws, especially when associated with impacted tooth. Colossal evaluations are necessary to unfold

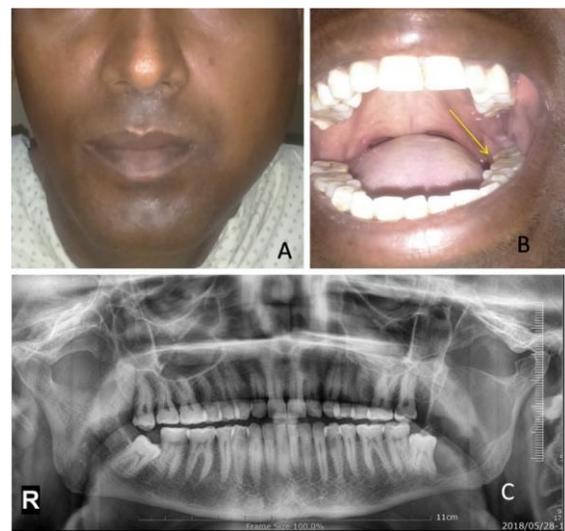


the definitive origin of OOC, as it can help in accurate treatment planning

References

1. ShubhangiJedheMhaske, RaginiMulchandani, Swati Saawarn, SonalSubhashMandale. Unusual aggressive presentation of orthokeratinized odontogenic cyst - a case report with systematic review of literature. *International Journal of Contemporary Medicine Surgery and Radiology*.2017;2(3):97-101.
2. Bharathi R, Santiago G, Nallaiyan P. Orthokeratinized odontogenic cyst. *SRM J Res Dent Sci* 2014; 5:287-9.
3. González GalvánMdel C, Garcia-Garcia A, Anitua-Aldecoa E, Martinez-Conde Llamosas R, Aguirre-Urizar JM. Orthokeratinized odontogenic cyst: A report of three clinical cases. *Case Rep Dent* 2013; 2013:672383.
4. 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.
5. Nishana M, Mohtesham I, Prabhu V et al. Orthokeratinized odontogenic cyst: a case report- a milder variant of OKC or an independent entity. *Int J Health Sci Res*. 2017, 7(9):350-354.
6. Kamat M, Kanitkar S, Datar U, Byakodi S. Orthokeratinized odontogenic cyst with calcification:
A rare case report of a distinct entity. *J Oral MaxillofacPathol* 2018; 22:S20-3.
7. Sarkar S, Khursheed O, Bansal R, Burza WK. Aggressive orthokeratinized odontogenic cyst: A rare case report and review. *J Adv Oral Res* 2015;6(2):40-43.
8. Bhasin N, Sreedevi, Pathak S, Puttalingaiah VD. Orthokeratinized odontogenic cyst: A rare presentation. *J Indian Acad Oral Med Radiol* 2014;26 (1):119-23.
9. 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.
10. Byatnal A, Natarajan J, Narayanaswamy V, Radhakrishnan R. Orthokeratinized odontogenic cyst-critical appraisal of a distinct entity.*Braz J Oral Sci* 2013;12:71-5.
11. Pimpalkar RD, Barpande SR, Bhavthankar JD, Mandale MS. Bilateral orthokeratinized odontogenic cyst: A rare case report and review. *J OralMaxillofacPathol* 2014; 18:262-6.
12. Swain N, Patel S, Poonja LS, Pathak J, Dekate K. Orthokeratinized odontogenic cyst. *J Contemp Dent* 2012; 2:31-3.
13. Koizumi.Odontogenickeratocyst,Orthokeratinized odontogenic cyst and epidermal cyst:Animmunohistochemical study Including markers of proliferation,cytokeratin and opoptosis related factors.*Int J Oral-Med Sci* 2004;2(1):14-22.

Figures with Legends:



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Figure 1: (A) Facial asymmetry on left lower back tooth region. (B) Mild lingual expansion distal to 37. (C) Large unilocular Radiolucency involving the impacted 38.

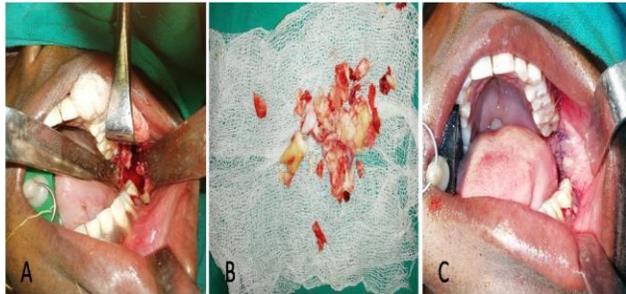


Figure 2: (A) Intra-operative image showing opened cystic lesion. (B) Enucleated cystic lining, Keratin material & extracted 38. (C) Closure of wound by placing sutures.

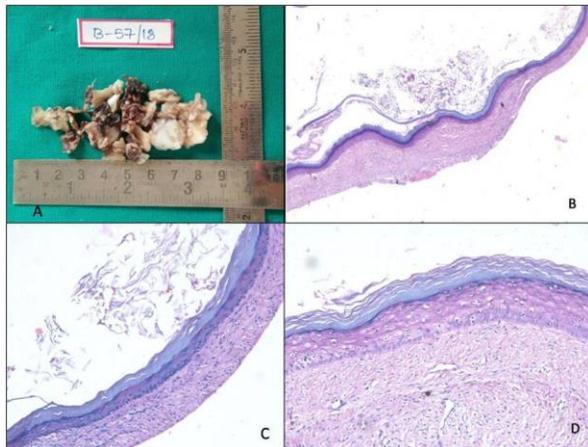


Figure 3: (A) Gross specimen showing cystic lining and keratin material. (B) Scanner view showing Orthokeratinized stratified squamous epithelium & cystic capsule. Lumen showing keratin flakes. (C&D) Showing thick Orthokeratinized stratified squamous epithelium with prominent granular cell layer.

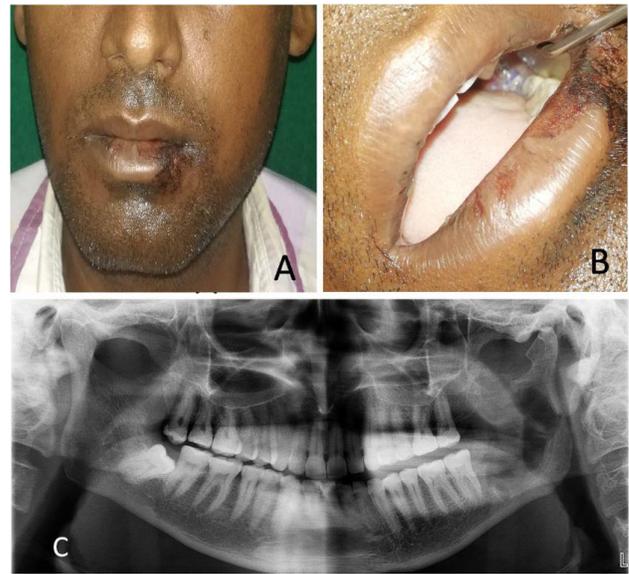


Figure 4: (A, B & C) Post-operative images showing regression of lesion clinically and radio graphically.