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Orthokeratinised Odontogenic Cyst of the Jaw- Report of Two Cases and Review of Literature

Shiladitya Sil, Rudra Pyne, Subhankar Ghosh, Nilanjana Saha.

1. Dept of Oral Medicine and Radiology, North Bengal Dental College and Hospital.
2. Dept. Of Oral and Maxillofacial pathology, Dr R Ahmed Dental College and Hospital.
3. Dept of Dentistry, Burdwan Medical College and Hospital.
4. Dept of Dentistry, Deben Mahato Government Medical College and Hospital.

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KEYWORDS

impacted tooth, root resorption, unilocular radiolucensy, ortho-keratinisation, palisaded epithelium.

ABSTRACT:

Introduction-

Orthokeratinized Odontogenic Cyst (OOC) is a rare developmental cyst with distinct histopathological characteristics. Initially classified as a variant of odontogenic keratocyst (OKC), OOC was later recognized as a separate entity by WHO classification (2017) due to its unique features.

Case Report-

A 17-year-old male presented with painful swelling in the lower jaw, progressively increasing over 2-3 months. Clinical examination revealed bony hard growth in the posterior mandible, confirmed as OOC through imaging and histopathology. Surgical enucleation resulted in uneventful healing and no recurrence over 12 months.

A 29-year-old male reported pain and reduced mouth opening in the lower jaw. Clinical and radiographic evaluations showed unilocular homogenous radiolucensy with root resorption in 36 37 and impacted 38, confirmed as OOC histo-pathologically. Surgical enucleation led to successful recovery with no recurrence over 7 months.

Conclusion-

These cases illustrate rare occurrences of OOCs in the posterior mandible that were clinically diagnosed as OKC/ Dentigerous cyst.

Introduction-

Orthokeratinized Odontogenic Cyst (OOC) is a relatively rare developmental odontogenic cyst, recognized for its distinct histo-pathological features. First identified by Schultz in 1927[1] and later classified by Wright in 1981 as a variant of the odontogenic keratocyst (OKC),[2] OOC has since undergone several changes in its classification. In 2005, the World Health Organization (WHO) reclassified OKC as a keratocystic odontogenic tumor (KCOT) due to its aggressive nature and high recurrence rates. However, this reclassification excluded the orthokeratinized variant from the KCOT spectrum, leaving OOC without a clear categorization. This gap was addressed in the 2017 WHO classification, which reintroduced odontogenic cysts and reinstated OKC under its original terminology while recognizing OOC as a distinct entity separate from OKC.[3]

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OOC primarily affects males and is commonly found in the molar and posterior regions of the mandible. It is usually asymptomatic, with most cases being incidentally discovered during routine radiographic examinations.[2] However, in some instances, the cyst can grow large enough to cause cortical expansion, leading to swelling and pain.[4] Radiographically, OOC typically appears as a well-defined unilocular or multilocular radiolucency, often associated with unerupted teeth but without causing root resorption.[4, 5]

Histologically, OOC is characterized by a thin, uniform lining of orthokeratinized stratified squamous epithelium, usually 5-8 cell layers thick. Unlike OKC, the keratin surface in OOC is thick and lamellated, without the corrugated appearance seen in OKC. The basal cells in OOC are typically cuboidal and lack the palisading of nuclei that is characteristic of OKC. Additionally, OOC has a prominent granular cell layer, which further distinguishes it from OKC. In cases of inflammation, the epithelial lining may become non-keratinized or parakeratinized.[6]

One of the most significant differences between OOC and OKC is their behavior and recurrence rates. OOC is less aggressive and has a significantly lower recurrence rate of less than 2% compared to OKC, which can recur in up to 28% of cases.[7] This lower recurrence rate makes surgical enucleation the preferred treatment for OOC, often involving the removal of the associated teeth. The prognosis for OOC after enucleation is generally favorable, with minimal risk of recurrence.[8]

In contrast, OKC's aggressive behavior and higher recurrence rates often necessitate more extensive surgical interventions, especially for larger lesions. Furthermore, while about 5% of OKC cases are associated with the Nevoid Basal Cell Carcinoma Syndrome (NBCCS) and may present with multiple lesions, such associations have not been observed with OOC. Multiple occurrences of OOC are rare, and no link to NBCCS has been established.[3]

It is crucial to differentiate OOC from OKC due to their distinct clinical and histopathological features, as well as their differing behaviors and recurrence risks. Accurate diagnosis and appropriate treatment are essential to managing these cystic lesions effectively. Herein we report two cases of OOC and the challenges faced in diagnosis.

Case 1-

A 17 year old male patient reported to the OPD with chief complaint of painful swelling in the lower left back tooth region that has been gradually increasing in size since last 2-3 months. The pain was dull, continuous type, not aggravated by lying down or extremes of temperature. It was relieved by analgesics. The patient gave no H/O fever or associated signs.

Gross facial asymmetry was appreciated on extra-oral examination owing to a swelling in the lower third of the face on the left side. Intra-oral examination revealed pathological migration in relation to 36 37. Mild expansion of the buccal cortical plate could be elicited. No surface ulceration or discharging sinus was present. Clinically 38 was not visible.

On palpation, the lesion was tender, bony hard in consistency with no evidence of de-cortication and no local rise in temperature. Paresthesia was not elicited. Buccal cortical plate expansion was appreciated in 36 37 region with no area of decortication. Tooth mobility was absent in 35 36 37.

Cone Beam Computed Tomography (CBCT) was advised that revealed a well defined well corticated homogenous radiolucensy in the left body of the mandible measuring 3 X 3.5 cm in diameter causing the pathological migration of 36 37 without any evidence of root resorption. Inferior displacement of inferior alveolar canal was noted. 38 was in developmental stage and not impacted. A differential diagnosis of odontogenic keratocyst and unicystic ameloblastoma was given.

Surgical enucleation was done from 36 37 region, taking care not to disturb the developing tooth germ of 38 and the specimen was sent for histo-pathological evaluation. H & E stained section of the specimen revealed the presence of multiple bits of tissue composed of fragmented orthokeratinised stratified squamous epithelial lining with prominent granular cell layer. Focal areas of palisaded basal cells are noted in the epithelial lining. Laminated sheets of keratin are present in the lumen. The fibrous capsule shows patchy inflammatory cell infiltration. A final diagnosis of Ortho-keratinised Odontogenic Cyst was given. A 12 month follow up revealed no recurrence.

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Case 2-

A 29 year old male patient reported to the OPD with chief complaint of pain in the lower left back tooth since last 3 weeks with reduced mouth opening since last 7 days. The pain is localized in the lower left region and is of dull, continuous type, with no aggravating or relieving factors. No relevant medical or habit history could be elicited.

Mild facial asymmetry was present on extra-oral examination, on the left side due to a swelling in the lower third of the face. Intra-oral examination revealed the presence of 36, 37 with slight buccal cortical plate expansion. There was no evidence any surface ulceration or pus discharging sinus. The mouth opening was initially measured at 7.33 mm. 38 was clinically missing, raising suspicion of impaction.

On palpation, the swelling was tender, bony hard in consistency with no evidence of de-cortication and no local rise in temperature. Paresthesia was present on the left side, but 36 37 were not mobile. Buccal cortical plate expansion was appreciated in the 36 37 region with de-cortication of the alveolar bone distal to 37 that representing soft, smooth-surfaced, non-tender area with no discharging sinus.

Orthopantomogram (OPG) was advised that revealed a diffuse well corticated homogenously radiolucent area extending antero-posteriorly from mesial root of 36, involving the ascending ramus and terminating 3-4 mm short of the massateric notch. The inferior alveolar canal appeared obliterated in that region. Root resorption was evident in the distal root of 36 and 37 with 38 being displaced to the left angle of the mandible. The inter-dental bone in relation to 36 37 appeared intact, presence of a bony septa was noted in the homogenous radiolucensy in between 36 37.

Differential diagnosis of Dentigerous cyst, OKC and Unicystic variant of Ameloblastoma was given. Incissional biopsy was performed from the left alveolar ridge and sent for histo-pathological evaluation. H & E stained section of the specimen revealed the presence of a cystic wall composed of odontogenic cystic liningbacked by fibro-collagenase capsule. The odontogenic cystic lining shows corrugated superficial ortho-keratinisation and variable thickness in areas of keratinisation. Mature keratin protein fibres can be seen extruded into the cystic lumen. A final diagnosis of Ortho-keratinised Odontogenic Cyst was given. The patient underwent complete surgical excision of the lesion with partial hemi-mandibulectomy and reconstruction. A 7 month follow up yielded no evidence of recurrence.

Discussion-

OOC once considered a variant of the OKC, is now recognized as a distinct entity due to its unique clinicopathologic characteristics. A comprehensive study conducted by Dong et al. in 2010 on the Chinese population examined 61 cases of OOC, revealing that the majority of patients were male with an average age of 39.[5] Most cases presented as jaw swellings, with a significant portion associated with impacted teeth, and radiologically, all were radiolucent with a predominance of unilocular lesions. Notably, none of these cases recurred during a follow-up period of 282 months. This study's findings align with previous research, indicating a male predominance and mandibular involvement in most cases, and all being radiolucent on radiographic evaluation. Similar findings were noted in the present case series, where both were males in their second and third decade of life having mandibular manifestation with similar radiographic presentation.

OOCs typically present as solitary cysts, though rare bilateral occurrences have been reported. For instance, Pimpalkar et al. described a case involving a 22-year-old male with bilateral OOCs,[6] consistent with other case series such as those reported by Oh et al.[9] Histologically, OOCs are distinguishable from OKCs by their orthokeratinized squamous epithelium with a prominent granular cell layer and low cuboidal basal cells, unlike the palisaded nuclei and parakeratinized epithelium characteristic of OKCs.[10] The present study confirmed these features, with cases exhibiting uniform thickness of 6-8 layers, absence of rete ridges, and a cystic lumen filled with keratin flakes.

Additionally, some OOCs show unique histopathologic traits such as partial histiocytic lining, sebaceous differentiation, and in rare cases, dystrophic calcification.[11] The need to distinguish between OOC and OKC in clinical practice is critical due to their different behaviors. OKC is more aggressive with a higher recurrence rate, while OOC has a lower

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recurrence rate and is less aggressive.[10] This distinction is essential in determining the appropriate treatment approach. For example, Selvamani et al.'s 2014 study on the South Indian population emphasized the necessity to differentiate between the two entities to tailor treatment strategies effectively. They found that while the clinical presentations of OOC and OKC were similar, their histologic features were markedly different.[11] Interestingly, OOC can sometimes mimic other odontogenic cysts, such as dentigerous cysts, as reported in a case study by Shetty et al.,[7] where an OOC presented radiographically similar to a dentigerous cyst. The study highlighted the diagnostic challenge posed by OOCs, especially when inflammation obscures typical histologic features. The second case reported in the present case series was associated with impacted 38, clinically and radiographically mimicking dentigerous cyst.

The literature also notes rare instances where OOCs have transformed into malignant lesions, although this is uncommon. MacDonald-Jankowski's study, which reviewed multiple OOC cases, found only a 4% recurrence rate and two cases of malignant transformation.[12] Further studies have explored the potential for malignancy in OOCs, with reports of squamous cell carcinoma (SCC) developing in long-standing OOCs.[13] Such cases underline the importance of careful follow-up after treatment, as delayed malignant transformation can occur. Oral surgeons are advised to distinguish OOC from other odontogenic cysts, recognize its malignant potential, and encourage patients to attend regular follow-ups.[14] The current understanding of OOCs is still evolving, with more research needed to fully comprehend its histogenesis and potential for malignancy.[15] In the present case series, both the patients showed complete recovery with no evidence of recurrence or malignant transformation. Table 1 shows a compilation of the recent reported cases of OOC and their associated features.

Immunohistochemical studies have shown lower proliferation markers in OOCs compared to OKCs, suggesting different biological behaviors. However, the clinical implications of these findings remain uncertain.[16] The literature also highlights the absence of an association between multiple OOCs and syndromes like NBCCS, contrasting with the known association between multiple OKCs and this condition.[3, 17]

Root resorption is usually associated with aggressive lesions like OKC, Dentigerous cyst, ameloblastoma etc.[18] It is evidenced from existing literature that OOCs rarely shows root resorption (Table 1). One unique presentation of this case series is that the second patient had root resorption. Literature suggests that the lack of space and presence of dense compact bones in the posterior mandible accounts for OOCs to extend into the tooth bearing areas of the mandible, thereby causing tooth root resorption.[19]

Table 1 showing the list of previously reported OOCs in the literature-

SL NO	Authors	Year	Location	Clinical features	Radiological features	Histological features	Treatment outcomes	Recurrences
1.	Dong et al[4]	2010	China	Males, avg. age 39, jaw swelling, impacted teeth	Radiolucent, mostly unilocular	Orthokeratinized epithelium, granular layer	Enucleation	No recurrence observed in 282 month follow up
2.	Pimpalkar et al[5]	2014	India	22-year-old male with bilateral OOCs	Not specified	Orthokeratinized epithelium, keratin-filled lumen	Surgical enucleation with peripheral ostectomy	Not specified

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3.	Selvamani et al[11]	2014	South India	Emphasized need to differentiate OOC from OKC	Similar to OKC but distinct in histology	Orthokeratinized epithelium, lack of rete ridges, keratin flakes in lumen	Tailored treatment strategies	Not specified
4.	Shetty et al.[7]	2016	South India	Painless, asymptomatic	Similar to dentigerous cyst	Orthokeratinized epithelium, inflamed areas	Surgical excision and re-contouring	No recurrence after 18 month follow up
5.	MacDonald-Jankowski[12]	2010		Mean age- 35 years, more common among males in the lower jaw	Radiolucent, mostly unilocular	Orthokeratinised with palisaded alignment of basal cells	Surgical enucleation	4% recurrence,malignancy in 2 cases
6.	Mahdavi et al[13]	2021	Iran	Third to fifth decade, M>F, More in mandible.	Radiolucent, mostly unilocular	Orthokeratinised with prominent granular cell layer	Surgical excision/ enucleation	Recurred as malignancy
7.	Mehdizadeh et al[16]	2022	Iran	26 year old, male, right posterior mandible, painful swelling	Unilocularradiolucensy with impacted 48	orthokeratinized stratified squamous epithelium with prominent granular cell layer	Complete enucleation and curettage	No recurrence after 12 month follow up
8.	Dineshkumar et al[9]	2024	South India	Painful swelling, M>F, Second to third decade, More in mandible	Well circumscribed radiolucensy, mostly associated with impacted tooth	orthokeratinized stratified squamous epithelium with prominent granular cell layer	Enucleation	No recurrence after 18 month follow up
9.	Crane et	2020	Lei	Second decade,	Unilocularc	Orthokerat	Enucleation	No

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	al[14]		cester, UK.	male, more in mandible.	ircumscribed radiolucency with impacted teeth	inized stratified squamous epithelium with prominent granular cell layer	with osteotomy and curettage	recurrence after 24-48 month follow up
10.	Mehdizadeh et al[7]	2022	Iran	14 year old male, painless swelling, maxilla, regional tooth displacement	Well-defined, corticated unilocular radiolucent with floor of maxillary sinus pushed upwards	cystic lesion, lined by an orthokeratinized stratified squamous epithelium with a prominent granular cell layer	Enucleation	No recurrence after 6 month follow up

Conclusion-

In conclusion, OOC is a rare but distinct odontogenic cyst with specific clinical and histologic features. It presents predominantly in males, often in the mandible, and is usually asymptomatic, though it can occasionally cause swelling or other symptoms. Radiographically, OOCs are typically unilocular and associated with impacted teeth. Treatment primarily involves enucleation with or without peripheral ostectomy, and the prognosis is generally favorable, with a low recurrence rate. However, due to the potential for malignant transformation, long-term follow-up is recommended.

Future direction-

The malignant potential of OOCs is poorly understood, long term follow up of treated OOCs is necessary to clearly understand the pathogenesis.

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Figures

Figure 1 showing the extra-oral profile of the patient (Case I).

Figure 2 showing the intra-oral presentation of the patient (Case I) that reveals Buccal cortical plate expansion with pathological migration of 36 37.

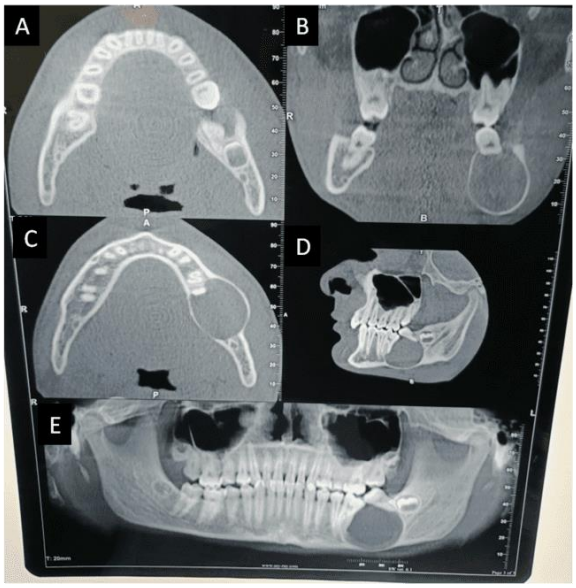

Figure 3 showing the axial section of the CBCT (A, C), coronal section (B) and saggital section (D) that reveals well defined well corticated uni-locular homogenous radiolucensy with displacement of 36 37, without any root resorption. Inferior displacement of the inferior alveolar canal is evident in the two dimensional reconstructed image (E), without any evidence of tooth impaction. (Case I)

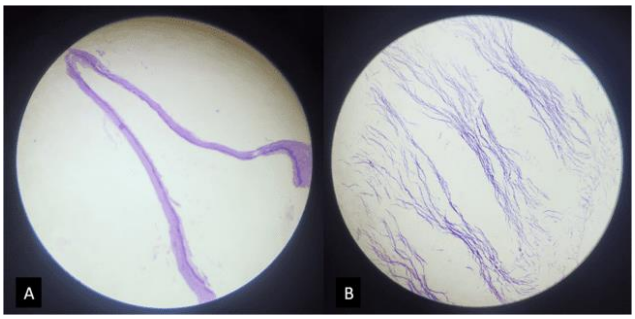

Figure 4 showing the photomicrograph of the H & E stained specimen (Case I) that reveals the presence of multiple bits of tissue composed of fragmented orthokeratinised stratified squamous epithelial lining with prominent granular cell layer. Focal areas of palisaded basal cells are noted in the epithelial lining (A). Laminated sheets of keratin with bundles of collagen fibres are present in the lumen. The fibrous capsule shows patchy inflammatory cell infiltration. (B)

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Figure 5 showing the extra-oral presentation of the patient (Case II), displaying the frontal (A) and lateral profile (B).

Figure 6 showing the intra-oral presentation of the patient (Case II) that reveals the presence of 36 37 with Buccal cortical plate expansion. 38 is clinically missing.

Figure 7 reveals the orthopantomogram of the patient (Case II) that reveals a well defined well cortical unilocular homogenous radiolucensy in the left mandible with impacted 38, and causing root resorption of 36 37. Obliteration of the inferior alveolar canals is appreciated in that region.

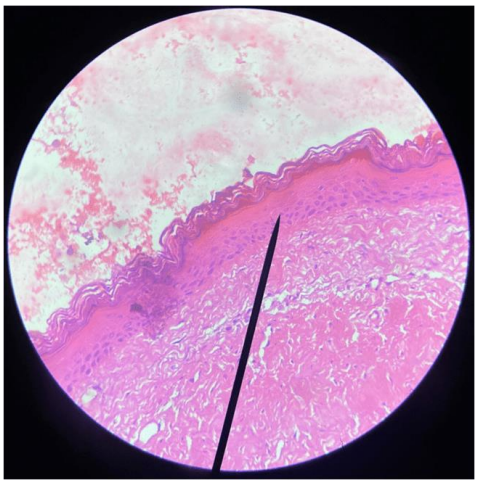

Figure 8 showing the photo-micrograph of the H & E stained specimen (Case II) that reveals the presence of a cystic wall composed of odontogenic cystic liningbacked by fibro-collagenase capsule. The odontogenic cystic lining shows corrugated superficial ortho-keratinisation and variable thickness in areas of keratinisation. Mature keratin protein fibres can be seen extruded into the cystic lumen.