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BEYOND SOLITARY OOC: SYNCHRONOUS MULTI-QUADRANT INVOLVEMENT

Oral & Maxillofacial Pathology

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ABSTRACT

Orthokeratinized odontogenic cyst (OOC) is a distinct developmental odontogenic cyst characterized by its low aggressiveness and low recurrence rate. Multifocal involvement is exceedingly rare. We report a case of a 17-year-old male presenting with cystic lesions involving three quadrants of the jaws. Clinical and radiographic evaluation suggested odontogenic keratocyst and infected dentigerous cyst. However, histopathological examination confirmed the diagnosis of OOC. This case highlights the rare multifocal presentation of OOC and emphasizes the importance of histopathological evaluation in establishing a definitive diagnosis and guiding appropriate management.

KEYWORDS

Orthokeratinized Odontogenic Cyst, Odontogenic Keratocyst, Dentigerous Cyst, Orthokeratinization

INTRODUCTION

Orthokeratinized Odontogenic Cyst (OOC) is a rare benign developmental odontogenic cyst, accounting for approximately 1% of all odontogenic cysts and 7–17% of keratinizing jaw cysts.[1] First described by Schultz (1927), it was later established as a distinct clinicopathological entity by Wright (1981) [2] and classified as a separate cystic lesion by WHO (2017).[3] Histopathologically, lined by orthokeratinized stratified squamous epithelium with a prominent granular cell layer and non-palisaded basal cells. It commonly involves posterior mandible and often associated with impacted teeth. We report a rare case of multifocal OOC involving three separate jaw sites in a patient.

CASE REPORT

A 17-year-old male patient reported to the department of Oral and Maxillofacial Pathology and Oral Microbiology, Best Dental Science College, Madurai and presented with a complaint of swelling in the posterior regions of the jaws that had gradually increased in size over a period of one month. There was no history of pain, discharge, trismus or associated systemic symptoms. Extraoral examination revealed no facial asymmetry. Intraoral examination revealed swelling involving bilateral posterior maxilla and left posterior mandible with mild buccal cortical expansion. The overlying mucosa appeared normal without evidence of infection or discharge.

Orthopantomographic examination revealed multilocular radiolucent lesions associated with impacted third molars in the right maxillary and left mandibular posterior regions, suggestive of Odontogenic keratocyst. A unilocular radiolucency associated with an impacted third molar was observed in the left maxillary posterior region, suggestive of dentigerous cyst. Displacement of 36 and 37 with root resorption were evident. Based on the clinical and radiographic findings, provisional diagnoses of odontogenic keratocyst in the left mandibular and right maxillary regions and dentigerous cyst in the left maxillary region were made.

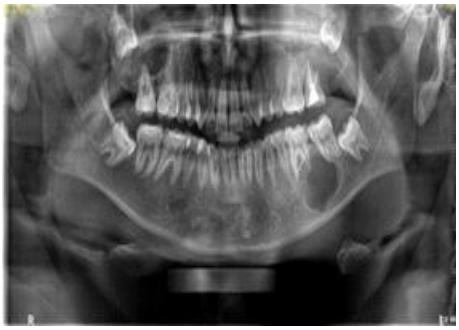

Figure 1: Orthopantomogram showing radiolucency involving bilateral posterior regions of the jaws

Differential diagnosis of dentigerous cyst, unicystic ameloblastoma and odontogenic keratocyst were made. Then the patient underwent surgical enucleation of all three lesions along with extraction of the corresponding third molars. Peripheral curettage was performed to reduce the risk of recurrence. Gross examination revealed three separate hard and soft-tissue specimens of varying dimensions, labelled A, B and C, corresponding to lesions from different quadrants of the jaws.

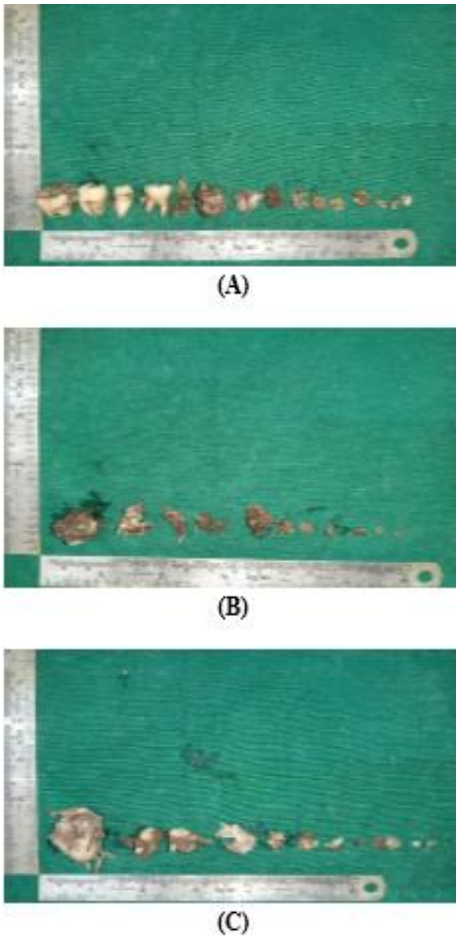

Figure 2: Photographs showing excised hard and soft-tissue biopsy specimens. A-Left maxillary posterior region, B-Right maxillary posterior region, C- Left mandibular posterior region

Histopathological evaluation of hematoxylin and eosin–stained (H&E) sections from all three specimens (A, B and C) demonstrated similar features diagnostic of orthokeratinized odontogenic Cyst (OOC). The cystic lining was orthokeratinized and composed of 2–4 cell layers of uniform thickness with surface corrugations and absence of rete ridges (Figure 3). The cystic lumen contained abundant lamellated keratin flakes (Figure 4). The connective tissue capsule was hyalinized and composed of fibroblasts, dense collagen bundles and odontogenic islands. Few blood vessels engorged with red blood cells and focal haemorrhagic areas were evident. Absence of inflammatory cell infiltrate.

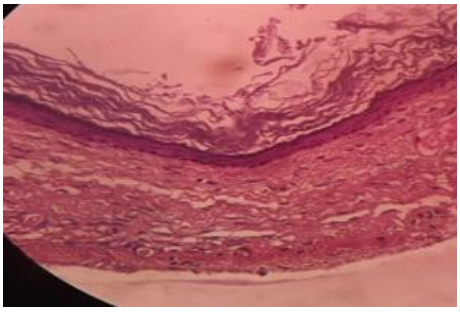

Figure 3: Photomicrograph showing cystic lumen, lining epithelium and connective tissue capsule (H&E,40×)

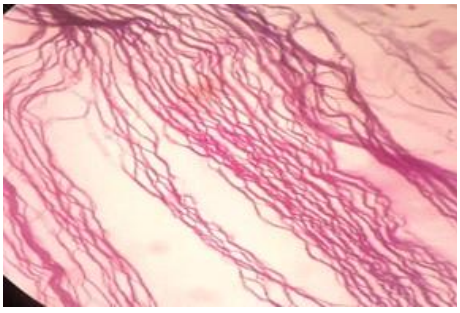

Figure 4: Photomicrograph showing lamellated keratin flakes (H&E, 40×)

Immunohistochemical (IHC) analysis was performed using Ki-67 marker to assess the proliferative index of the orthokeratinized odontogenic cyst. The sections demonstrated low Ki-67 immunoreactivity under both low and high power magnification, with a proliferative index of 2.6% (Figures 5). Long-term periodic follow up is recommended due to risk of recurrence.

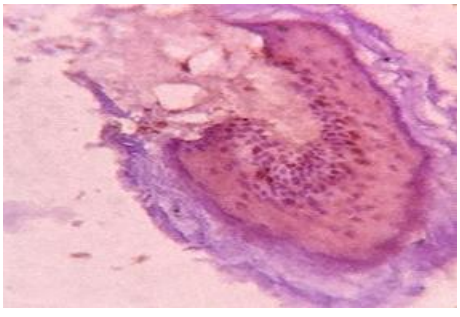

Figure 5: Photomicrograph of IHC section showing Ki-67 reactivity at (10×)

DISCUSSION

Orthokeratinized odontogenic cyst (OOC) is a distinct developmental odontogenic cyst that differs from the more aggressive parakeratinized odontogenic keratocyst (OKC).[4] Wright (1981) established OOC as a separate clinicopathological entity based on its characteristic histopathological features, less aggressive biological behavior, and lower recurrence rate. Subsequent studies have confirmed OOC as an independent lesion with low proliferative potential and a favorable prognosis.[5]

OOC is uncommon, accounting for approximately 1–11% of odontogenic cysts and 5–16% of lesions previously diagnosed as OKCs. MacDonald-Jankowski (2010) reported a male predilection (2.3:1), peak occurrence in the third and fourth decades and predominant involvement of the posterior mandible.[6] Clinically, OOCs are usually asymptomatic, slow-growing lesions frequently associated with impacted teeth, particularly third molars.[7] Radiographically, they typically present as well-circumscribed unilocular radiolucencies, whereas multifocal or bilateral presentations are rare, comprising less than 5% of reported cases.[8]The present case is unusual because three separate lesions were identified in different jaw quadrants of a 17-year-old male. Uddin et al. (2019) reported 10 cases of OOC, with only one demonstrating bilateral involvement, highlighting the rarity of multifocal presentations.[3]Such lesions may clinically and radiographically mimic dentigerous cysts, parakeratinized OKCs or unicystic ameloblastomas, making definitive diagnosis challenging.

Histopathologically, OOC is characterized by a thin orthokeratinized stratified squamous epithelial lining with a prominent granular cell layer and absence of basal palisading. The lumen typically contains keratin flakes, while the connective tissue wall may show minimal inflammation. Immunohistochemical studies, including Ki-67, demonstrate low proliferative activity, consistent with its low recurrence rate of approximately 2%.[9] In contrast, parakeratinized OKCs exhibit higher proliferative indices and recurrence rates ranging from 10–30%, emphasizing the importance of histopathological differentiation. The etiology of OOC remains uncertain, although derivation from dental lamina remnants or reduced enamel epithelium has been proposed. While multifocal lesions may raise suspicion of syndromic conditions such as Gorlin-Goltz syndrome, Gardner syndrome or Noonan syndrome, the present patient showed no evidence of syndromic association, supporting a sporadic multifocal occurrence.

Management is predominantly conservative, consisting of enucleation with or without peripheral curettage and is associated with an excellent prognosis. Zhang et al. (2010) reported only 2 multifocal cases among 61 OOCs, while Li et al. (2012) demonstrated molecular differences from syndromic OKCs, including infrequent PTCH1 alterations.[10] Crowley et al. (1992) further emphasized the clinicopathological differences between orthokeratinized and parakeratinized variants.[11] The present case contributes to the limited literature on multifocal OOC and highlights the importance of thorough clinicoradiographic and histopathological evaluation for accurate diagnosis and appropriate management.

CONCLUSION

Orthokeratinized odontogenic cyst (OOC) is a rare developmental odontogenic cyst. Despite its distinctive histopathological features, knowledge regarding its etiopathogenesis and biological behavior remains inadequate, particularly at the molecular level. Future research focusing on larger case series, molecular profiling and long-term follow-up is essential for establishing standardized diagnostic and treatment protocols, ultimately improving patient care and outcomes.

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