Original Research Paper

Volume - 15 | Issue - 08 | August - 2025 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Oral Pathology

BILATERAL OCCURRENCE OF DENTIGEROUS CYST AND ORTHOKERATINIZED ODONTOGENIC CYST IN THE MANDIBLE: A RARE CASE REPORT

Dr. G. Kesavan	Reader.
Dr. T. Cibi Shalini	Senior Lecturer.
Dr. G. Nithyasri*	Post Graduate. *Corresponding Author
Dr. S. Sivagurudev	Post Graduate.

ABSTRACT Bilateral odontogenic cysts are rare in occurrence, often associated with syndromic conditions. This case report presents a 35-year-old female with bilateral mandibular cysts of distinct histopathological origin. The left side was diagnosed as a dentigerous cyst (DC) associated with an impacted lower third molar, while the right side presented a multilocular orthokeratinized odontogenic cyst (OOC). Both cysts were surgically enucleated, and the patient showed no recurrence during follow-up. This report highlights the importance of comprehensive radiographic and histopathological evaluation in diagnosing and managing bilateral odontogeniccysts.

KEYWORDS : Dentigerous Cyst , Impacted Teeth, Orthokeratinized Odontogenic Cyst

INTRODUCTION

The Orthokeratinized Odontogenic Cyst (OOC) is a rare developmental cyst of the jaw that originates from odontogenic epithelium. It was first identified by Schultz in 1927, who initially categorized it as an intra-osseous dermoid cyst [1]. However, further research confirmed its odontogenic nature. For many years, OOC was considered as a subtype of Odontogenic Keratocyst (OKC) due to the presence of a keratinized epithelial lining in both lesions. In 1981, Wright distinguished OOC as a separate entity based on its unique clinicopathological characteristics, such as lower recurrence rates and limited growth potential compared to OKC. He referred it as the "odontogenic keratocyst-orthokeratinized variant", setting the foundation for its classification as an independent lesion. Further developments in terminology occurred in 1993, when Vuhahula et al introduced the term "jaw cyst with orthokeratinization", reinforcing its distinct nature[2]. Later in 1998, Li et al. formally coined the term "Orthokeratinized Odontogenic Cyst (OOC)," which has been widely accepted in oral pathology. A major shift in the classification took place in 2005, when the World Health Organization (WHO) redefined OKC as a Keratocystic Odontogenic Tumor (KCOT) due to its aggressive behavior, neoplastic potential, and higher recurrence rates[3]. However, OOC was recognized as a separate, non-neoplastic entity, characterized by lower recurrence and less invasive behavior. This reclassification solidified OOC's status as a distinct pathological condition rather than a mere variant of KCOT[4]. (Schultz, 1927; Wright, 1981; Vuhahula et al., 1993; Li et al., 1998; WHO, 2005). (Philipsen, 1956; MacDonald et al., 2010).

Odontogenic cysts are common jaw lesions, but bilateral presentation is rare and may indicate an underlying syndrome. Dentigerous cysts are typically associated with impacted teeth and has distinct histopathological characteristics[5]. The simultaneous occurrence of these two cysts on either sides of the mandible is extremely uncommon. This report aims to describe the clinical, radiographical and histopathological features of this unique case[6].

CASE REPORT:

A 35-year-old female patient reported to the department with a complaint of swelling in the posterior region of the left lower jaw. The swelling had been gradually increasing in size for past one month and was asymptomatic, with no associated pain or discharge.

Extraoral Examination revealed no facial asymmetry.

Intraoral Examination revealed bilateral swellings in the posterior mandible, more prominent on the left side. There was mild buccal cortical expansion bilaterally. The overlying mucosa was intact, and there were no signs of infection, discharge, or trismus.

Panoramic radiograph and **Cone Beam Computed Tomography** was taken. (Fig 1) shows a well-defined **unilocular radiolucency** associated with an impacted mandibular third molar on the **left side**, suggesting a dentigerous cyst. On the **right side**, a **well-defined multilocular radiolucency** was observed in the posterior mandibular ramus with an associated impacted tooth 48. A **supernumerary tooth** surrounded by a radiolucent area was noted beneath the roots of the erupted mandibular right second premolar and first molar. No signs of root resorption or tooth displacement were evident. Additionally, a radiolucency was seen around the impacted maxillary right third molar. CBCT image showing lesions on either side of jaw (Fig 2)

Fig 1: OPG showing lesions on either side of jaw

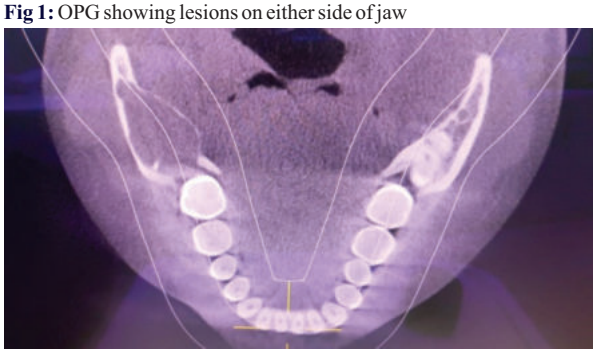

Fig 2: CBCT showing lesions on either side of jaw

Based on clinical and radiographic findings, the **provisional diagnosis** included:

Left Side: Dentigerous cyst

Right Side: Odontogenic keratocyst

Based on clinical and radiographic findings, the **differential diagnosis** included:

Left Side: Unicystic ameloblastoma

Right Side: Ameloblastoma, Dentigerous cyst, Odontogenic myxoma.

The patient underwent **surgical enucleation** of both lesions and extraction of corresponding third molars under local anesthesia (Fig 3). **Curettage** was performed to minimize recurrence. Postoperative

healing was uneventful and she is followed up for **six months** without any signs of recurrence on radiographic evaluation.

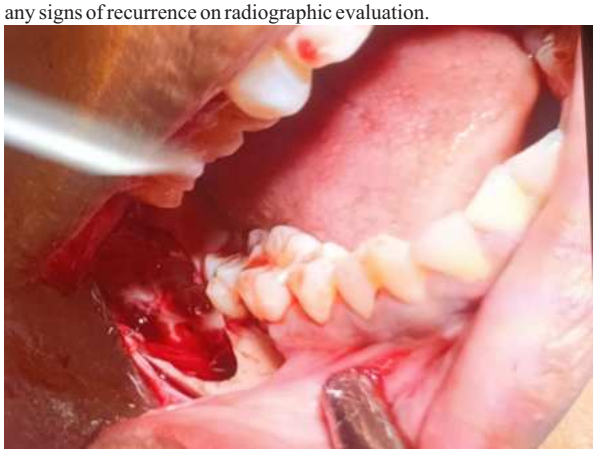

Fig 3: Surgical Removal Of The Lesion

Histopathological examination revealed the following:

Left Side (Dentigerous Cyst):

- The cyst lining was 2–3 cell layers thick, resembling reduced enamel epithelium. (Fig 4)
- No rete ridges was observed.(Fig 5)
- The connective tissue capsule was hyalinized, with bundles of collagen fibers.
- Islands of odontogenic epithelium were present.
- There was no evidence of inflammation.

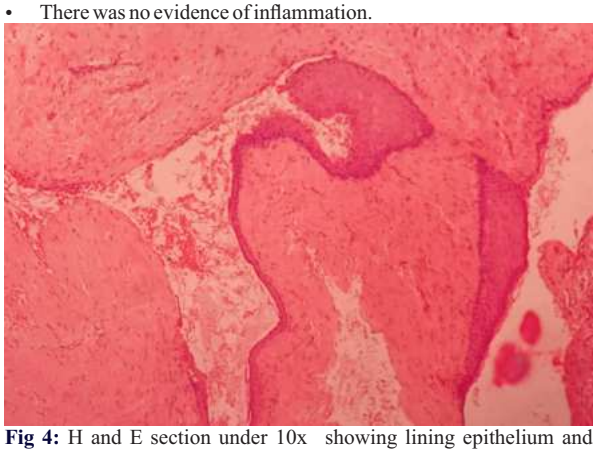

Fig 4: H and E section under 10x showing lining epithelium and connective tissue capsule

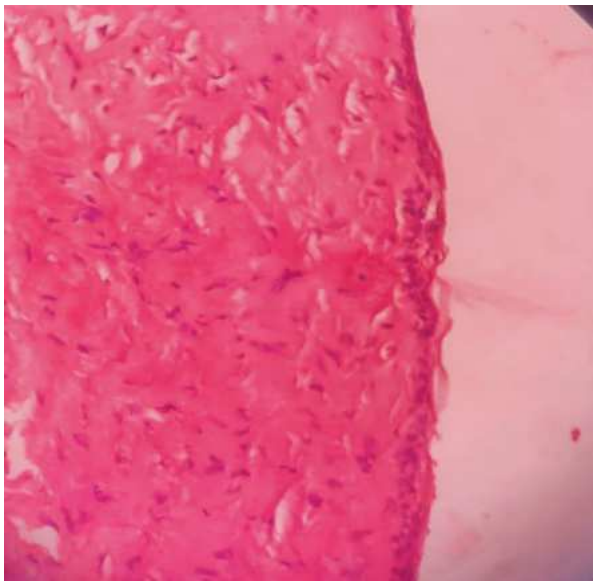

Fig 5: H and E section under 40x showing absence of rete ridges

Right Side (Orthokeratinized Odontogenic Cyst):

- The cyst lining exhibited 3–4 uniform cell layers with orthokeratinization and surface corrugations.(Fig 6)
- Absence of rete ridges.
- The cystic lumen showed keratin flakes resembling an onion-skin appearance.
- The fibrous capsule was hyalinized, with odontogenic epithelial islands.
- Few blood vessels were engorged with RBCs, and areas of hemorrhage were observed.
- The final diagnosis was given as a **dentigerous cyst** on the left and an **orthokeratinized odontogenic cyst (OOC)** on the right side of the mandible.

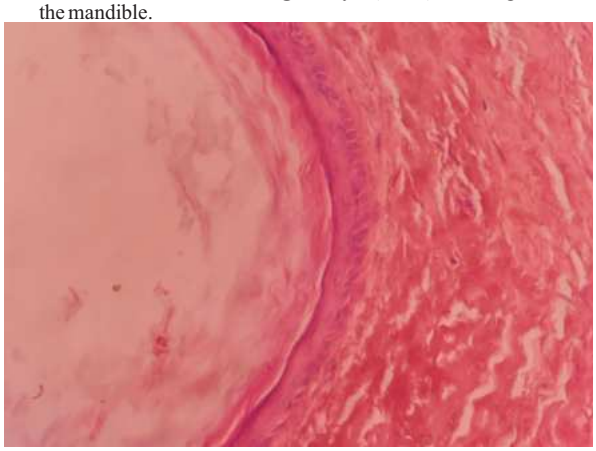

Fig 6: H and E section under 40x showing lining epithelium and connective tissue capsule

Immuno histochemical study done using Ki67 marker to check for proliferative index in OOC (right side). (Fig 7) and (Fig 8) showing Ki67 positivity under 10x and 40x respectively.

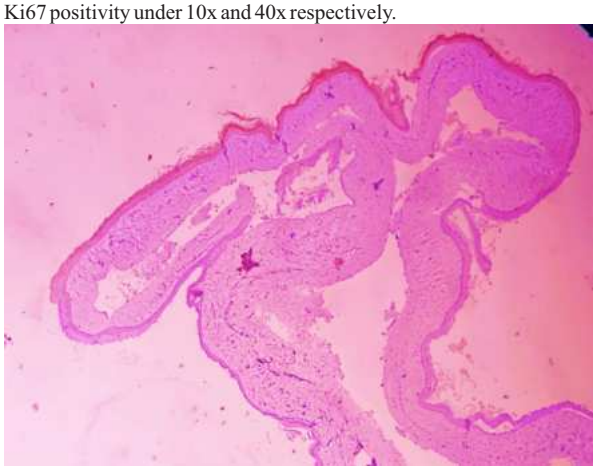

Fig 7:IHC section in 10x showing Ki67 reactivity

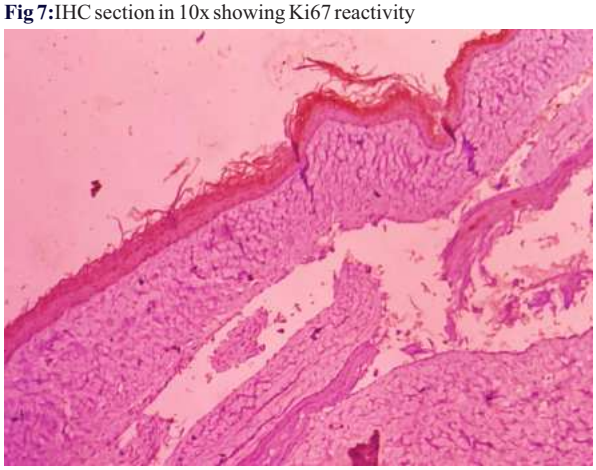

Fig 8: IHC section in 40x showing Ki67 marker with proliferative index of 2.6%

DISCUSSION

Philipsen in 1956 coined the term "odontogenic keratocyst" (OKC).

Since then, OKC has become the frequent diagnosis for both parakeratinized and orthokeratinized lining jaw cysts. However, a number of studies conducted throughout time have demonstrated that these two entities were different from one another[7]. While the orthokeratinized variant of OKC is now regarded as a separate entity, the parakeratinized variant was reclassified as keratocystic odontogenic tumor (KCOT) by WHO (2005) due to differences in behavior, recurrence rate, association with Nevoid Basal Cell Carcinoma Syndrome (NBCCS) and distinct histopathological and immunohistochemical features[8].

They represent around 1–11% of all odontogenic cysts. OOC accounts for approximately: 5.2% to 16.2% of cases that were previously classified as odontogenic keratocysts (OKCs) which are more aggressive and more frequently associated with NBCCS. Bilateral presentation is even rare with only 4 reported cases in the literature[9].

The exact etiology of OOC is not fully understood, but it is believed to arise from odontogenic epithelium. Several contributing factors and theories have been proposed that OOCs were derived from the dental lamina remnants or reduced enamel epithelium. They commonly occur in association with impacted teeth, particularly third molars, which supports the developmental theory. The epithelial lining undergoes orthokeratinization, giving the cyst its distinct histological characteristics[10]. Although OOCs are not typically associated with NBCCS, some molecular studies suggest that PTCH1 gene mutations (implicated in parakeratinized variants) may rarely be involved in sporadic OOC cases. However, OOCs generally lack the genetic alterations linked to NBCCS, making them biologically distinct from parakeratinized odontogenic keratocysts (OKCs).OOCs frequently develop around unerupted or impacted teeth, suggesting a relationship with the follicular epithelium[11]. The frequent association with mandibular third molars indicates a potential link between tooth eruption patterns and cyst formation. While OOCs are primarily developmental, local trauma or inflammation may play a secondary role in cyst formation or progression. Chronic irritation from impacted teeth could potentially stimulate cyst development.

OOCs are more frequently diagnosed in young to middle-aged adults, typically between 20–40 years. There is a slight male predilection, with a male-to-female ratio of approximately 2:1.No strong racial predilection has been identified. OOCs most commonly occur in the posterior mandible, particularly in the molar and ramus regions.These cysts are typically unilocular and well-defined radiolucency on radiographs, distinguishing them from more aggressive odontogenic lesions[12]. OOCs tend to have a less aggressive behavior and a lower recurrence rate (around 2%), compared to parakeratinized odontogenic cysts, which have a recurrence rate of 10–30%. Malignant transformation is extremely rare. (Zhang et al., 2010).

Histopathologically the epithelial lining of OOC is thin and uniform, typically composed of 4 to 9 cell layers.The surface shows orthokeratinization: The keratin layer is thick and onion-skin–like, with laminated keratin sheets devoid of pyknotic nucleus. Beneath the keratinized surface, there is a prominent granular cell layer (stratum granulosum), which is a hallmark of orthokeratinization. OOCs show little to no nuclear palisading or hyperchromatin. The basal layer lacks the characteristic polarized arrangement. The connective tissue capsule around the cyst is typically non-inflamed or only mildly inflamed[13].

In OOCs, Ki-67–positive cells are mainly localized to the basal cell layer whereas p63 expression is seen in the basal and some suprabasal layers. OOC shows strong CK10 expression while CK13 is expressed in occasional groups of cells in the suprabasal and superficial layers.CK14 gives a constant expression in both basal and suprabasal layers.

While the direct transformation of a dentigerous cyst (DC) into an orthokeratinized odontogenic cyst (OOC) is not a well-documented phenomenon, there are some plausible mechanisms and theories that may explain the occurence such as metaplastic changes in the cyst lining, orthokeratinization as a secondary change, development of OOC from the dentigerous cyst's epithelial remnants and misdiagnosis or overlapping histology.

When odontogenic cysts present bilaterally, it raises the suspicion of an underlying syndrome or genetic disorder. Here are some key syndromes associated with bilateral or multiple odontogenic cysts: NBCCS, Gorlin-Goltz Syndrome, Maroteaux-Lamy Syndrome, Gardner's Syndrome, Noonan Syndrome and Orofaciodigital Syndrome (OFD).In this case patient is non syndromic [14].

The standard treatment for OOC is enucleation with curettage. Due to its low recurrence rate, more conservative management is often sufficient. Long-term follow-up is still recommended, but the risk of recurrence is low.

OOCs have an excellent prognosis with minimal risk of recurrence Unlike parakeratinized odontogenic keratocysts, they are not linked to syndromic conditions and carry no significant risk of malignant transformation. Their biologically indolent nature makes them clinically favorable and less worrisome for patients[15].

This case highlights the importance of thorough clinical, radiographic, and histopathological evaluation in diagnosing and managing bilateral odontogenic cysts. Both cysts were successfully treated with enucleation and curettage. This case underscores the rarity of bilateral odontogenic cysts and the need to consider potential syndromic associations.

REFERENCES

1. Li, T. J., Kitano, M., & Komiyama, K. (1998). Orthokeratinized odontogenic cyst: A clinicopathological and immunohistochemical study. Journal of Oral Pathology & Medicine, 27(2), 71-75.
2. MacDonald, D. S. (2010). Lesions of the jaws. Clinical Radiology, 65(1), 2-13.
3. Ono S, Hirose K, Sukegawa S, Nakamura S, Motooka D, Iwamoto Y, Hori Y, Oya K, Fukuda Y, Toyosawa S. Multiple orthokeratinized odontogenic cysts: clinical, pathological, and genetic characteristics. Diagnostic pathology. 2022 Oct 14;17(1):82.
4. Schultz, L. H. (1927). Cysts arising from epithelial remnants of the dental lamina and their relationship to dermoids. American Journal of Orthodontics, 13, 205-213.
5. Vuhahula, E. A., Nikai, H., Ijuhin, N., Ogawa, I., Takata, T., & Kimi, K. (1993). Jaw cysts with orthokeratinization: Analysis of 12 cases. Journal of Oral Pathology & Medicine, 22(1), 35–40.
6. Wright, J. M. (1981). The odontogenic keratocyst: Orthokeratinized variant. Oral Surgery, Oral Medicine, Oral Pathology, 51(6), 609-618.
7. World Health Organization (WHO). (2005). WHO Classification of Tumours: Pathology and Genetics of Head and Neck Tumours. IARC Press.
8. Zhang LL, Yang R, Zhang L, Li W, MacDonald-Jankowski D, Poh CF. Dentigerous cyst: a retrospective clinicopathological analysis of 2082 dentigerous cysts in British Columbia, Canada. International journal of oral and maxillofacial surgery. 2010 Sep 1;39(9):878-82.
9. Gadipelly S, Reddy VB, Sudheer M, Kumar NV, Harsha G. Bilateral calcifying odontogenic cyst: a rare entity. Journal of maxillofacial and oral surgery. 2015 Sep;14(3):826-31.
10. Akkaş İ, Toptaş O, Özan F, Yılmaz F. Bilateral glandular odontogenic cyst of mandible: a rare occurrence. Journal of Maxillofacial and Oral Surgery. 2015
11. Freitas DQ, Tempest LM, Sicoli E, Lopes-Neto FC. Bilateral dentigerous cysts: review of the literature and report of an unusual case. Dentomaxillofacial Radiology. 2006 Nov 1;35(6):464-8.
12. Ustuner E, Fitoz S, Atasoy C, Erden I, Akyar S. Bilateral maxillary dentigerous cysts: a case report. Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontology. 2003 May 1;95(5):632-5.
13. Aloyouny A, Albagieh H, Mansour S, Mobarak F. Case Report: Bilateral mandibular buccal bifurcation cysts. F1000Research. 2021 Feb 19;9:1502.
14. Szaraz D, Ksinan AJ, Machacek C, Borilova Linhartova P. Developmental odontogenic cysts with special focus on the occurrence of multiple cysts and syndromic association: a single-centre cross-sectional study from the Czech Republic. Orphanet Journal of Rare Diseases. 2025 Mar 4;20(1):103.
15. Crane H, Da Forno P, Kyriakidou E, Speight PM, Hunter KD. Multiple orthokeratinized odontogenic cysts: a report of two cases and review of the literature. Head and neck pathology. 2020 Jun;14(2):381-5.