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Journal of Oral Medicine, Oral Surgery, Oral Pathology and Oral Radiology

Journal homepage: www.joooo.org

Case Series

Non-syndromic, synchronous multiple odontogenic keratocysts: A rare case-series

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Abstract

Odontogenic keratocysts have retained an enigmatic nature since their emergence, with their multifocal occurrence in both jaws complicating diagnosis and management. While OKCs are frequently linked to syndromic conditions such as Nevoid Basal Cell Carcinoma Syndrome (NBCCS), their occurrence in non-syndromic patients is rare and demands a distinct clinical approach. This case series analyzes non-syndromic patients presenting with multifocal, multiple OKCs of maxilla and mandible, focusing on their clinical, radiological, and histopathological characteristics and its aggressive behaviour necessitating long-term follow-up.

Keywords: Non-syndromic OKCs, Multiple sites, Enucleation.

Received: 07-03-2025; **Accepted:** 17-03-2025; **Available Online**: 03-04-2025

1. Introduction

Odontogenic keratocysts (OKCs), reclassified as such by WHO in 2018 from their interim classification as keratocystic odontogenic tumours, are benign, developmental, locally aggressive odontogenic cystic lesions exhibiting high risk of recurrence.[1] There was and still is a general belief that all OKCs derive from remnants of the dental lamina, as was first suggested by Hjörting-Hansen et al. in 1969 and reiterated by Toller. A study on syndromic cysts that were removed with inclusion of the attached oral mucosa or gingiva, showed that OKCs were derived from hamartias in the submucosa, which are derived from offshoots of the basal layer of the epithelium covering the mucosa, since in many cases they were connected to this basal layer. This theory was endorsed by Gorlin et al.[2] OKCs occur either as multiple (syndromic) or solitary (non-syndromic). Multiple OKCs typically occur as one of the findings in Gorlin–Goltz syndrome along with other abnormalities of the ribs, eyes, nervous system, and skin carcinomas.[3] Multiple OKCs occurring in non-syndromic patients prompt intriguing investigations into their etiology, including the potential role of unidentified genetic mutations or environmental influences. We hereby present a case series on multiple OKCs reported in our institution, managed with enucleation in conjunction with adjuvant therapies with no evidence of recurrence during subsequent reviews.

2. Case Series

2.1. Case 1

A fourteen-year-old boy presented with the chief complaint of occasional swelling and pus discharge from both the left lower and upper back teeth region for 6 months. There was no history of any associated toothache, numbness, or fever. He had a history of mild global developmental delay and macrocephaly when he was one and a half years old. He was also suspected of having Alexander disease. Systemic evaluation and genetic analysis ruled out the syndromic association.

On examination, a diffuse swelling noted on the left lower one-third of the face, which was soft in consistency, non-fluctuant, and tender on palpation. There was no localized rise in temperature, and the overlying skin was apparently normal. Ipsilateral level Ib lymph node was palpable—firm, mobile, and tender.

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https://doi.org/10.18231/j.jooo.2025.005

Intraorally, buccal and palatal cortical plate expansion noted in bilateral posterior maxilla from mesial aspect of 15 to maxillary tuberosity and mesial aspect of 25 to tuberosity. Minimal buccal and lingual cortical plate expansion were evident in the mandible along bilateral ascending ramus and from distal aspect of 33 to distal aspect of 45, with tenderness on palpation. Displacement of teeth noted from 33 to 42.

Radiographs revealed well-defined multiple radiolucencies on bilateral posterior maxilla, bilateral mandibular body-ramus region, and anterior mandible. The lesion extends along the entire left maxillary sinus, pushing the orbital floor superiorly. Multiple impacted teeth were noted in both jaws. Bicortical expansion and erosion were seen in the maxilla. Minimal buccal and lingual cortical expansion and breach noted at multiple levels in the mandible. Extraneous, replacement, envelopmental, and collateral variants were noted.

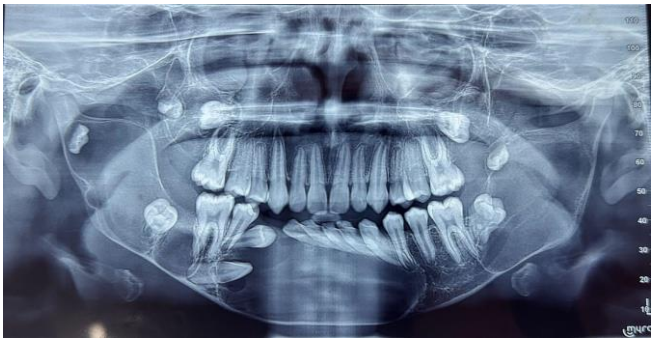

Figure 1: OPG demonstrating multiple radiolucent lesions involving bilateral posterior maxilla and entire mandible except 35, 36 region and bilateral condyles; multiple impacted and displaced teeth

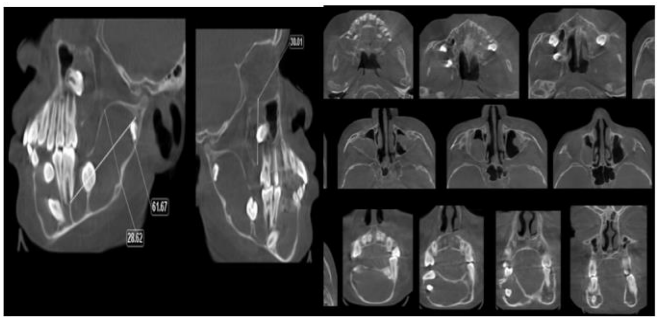

Figure 2: Uniformly hypodense areas with well-defined and scalloped borders; Bicortical expansion, thinning and breach, along with obliteration of sigmoid notch in the mandible and bilateral superior displacement of floor of maxillary sinus. Multiple impacted teeth within lesions in both jaws.

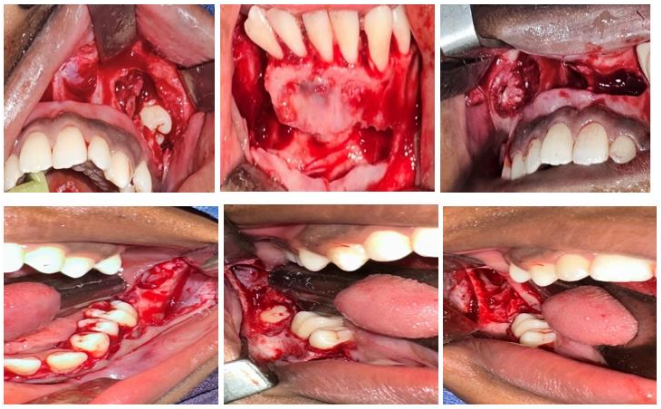

Figure 3: Intraoperative images showing intraoral exposure of cyst lining and multiple impacted teeth

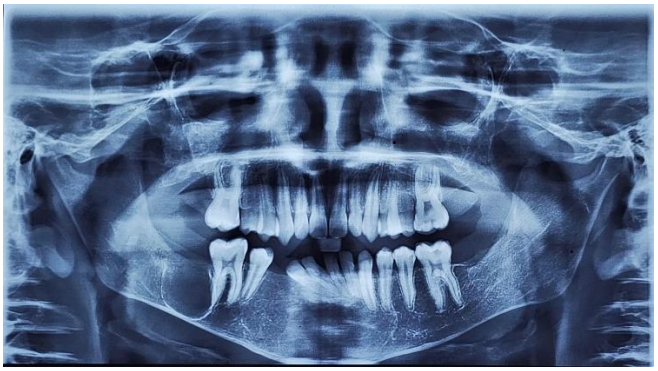

Figure 4: Post-operative OPG demonstrating significant bone regeneration and progress in healing

2.2. Case 2

A seventeen-year-old girl was referred to our department in view of cystic lesions on routine radiographic examination. No history of trauma, pain, swelling, or any discharge. No pertinent medical history and all laboratory investigations were within normal limits.

On examination, extraorally, no swelling was evident. Intraorally minimal buccal cortical plate expansion noted extending from 33 to 47. No visible pus discharge or bleeding and no tenderness on palpation.

Radiographs revealed two well-defined radiolucent lesions – one on the anterior mandible in the periapical region from the distal aspect of 32 to distal aspect of 43 and another one on the right body of the mandible with scalloped borders, extending from the mesial aspect of 44 to distal aspect of 47 and superoinferiorly from about 6-8mm below the alveolar crest to the lower border of the mandible. These are collateral variants. Thinning and expansion of labial cortical plate noted with anteroposterior expansion, more than buccolingual expansion. All teeth were vital except 46. RCT done in caries exposed 45.

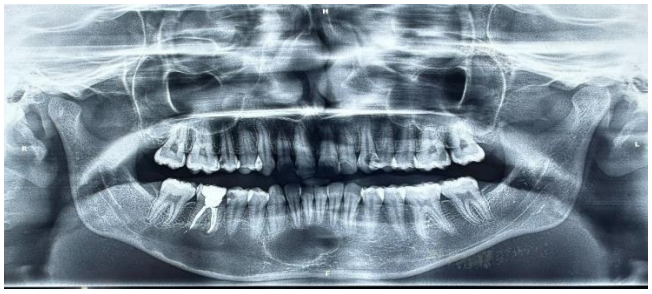

Figure 5: OPG demonstrating two well-defined radiolucent lesions in the right body and anterior mandible crossing the midline

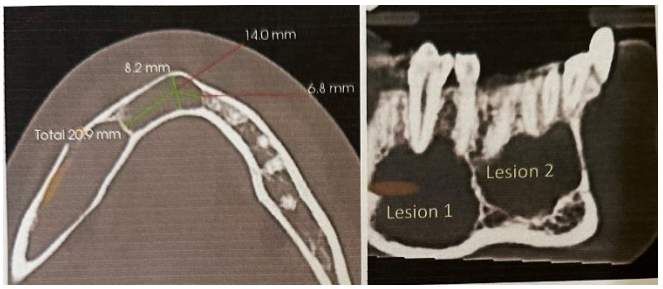

Figure 6: Individual cross-sections demonstrating two separate lesions with more anteroposterior expansion and superoinferior extent of the lesion from below the alveolar crest to 1cm above the inferior border of the mandible

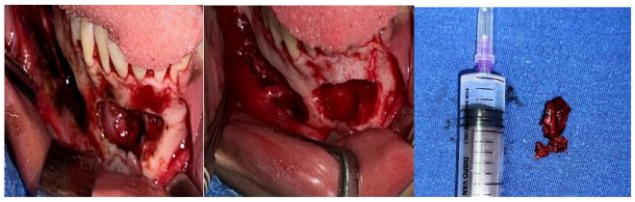

Figure 7: Intraoperative images demonstrating intraoral exposure of cyst lining and bony defect following enucleation; surgical specimen

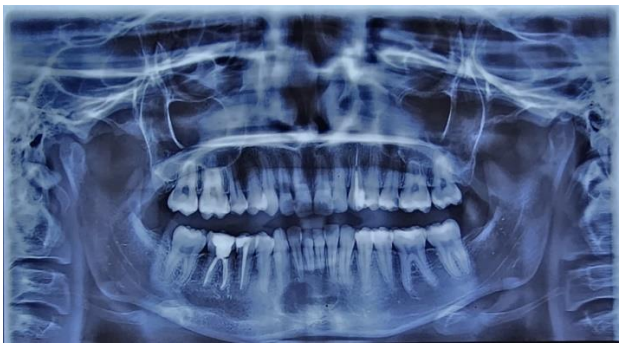

Figure 8: Post-operative OPG demonstrating effective bone regeneration

2.3. Case 3

A 37-year-old female reported with the chief complaint of nasal congestion and headache for 10 years. History revealed that the patient had undergone an MRI for the same and noted cystic lesions in the maxilla and mandible. The patient also complaints of occasional pus discharge from the left mandible. There was no relevant medical history, and routine hematological investigations were normal.

On examination, extraorally, no swelling was evident. Intraorally, mild buccal cortical plate expansion noted on bilateral posterior maxilla and left body and ramus of the mandible.

Radiographs revealed three well-defined unilocular radiolucencies involving bilateral posterior maxilla and left mandibular body and ramus with incomplete septae. Horizontally impacted 18 and 38. Root resorption was noted in 16, 17, and 27. Envelopmental and collateral variants were noted.

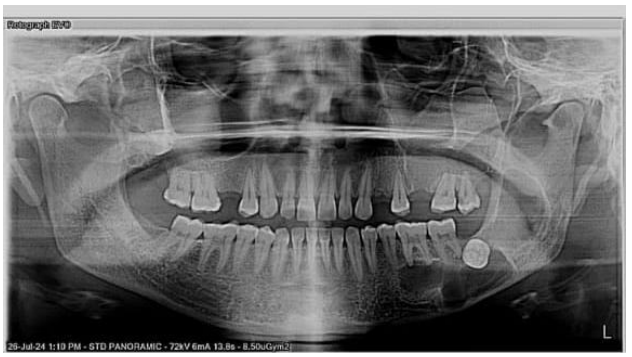

Figure 9: OPG showing multiple well-defined radiolucent lesions involving bilateral posterior maxilla and left mandibular body-ramus region along with multiple impacted teeth

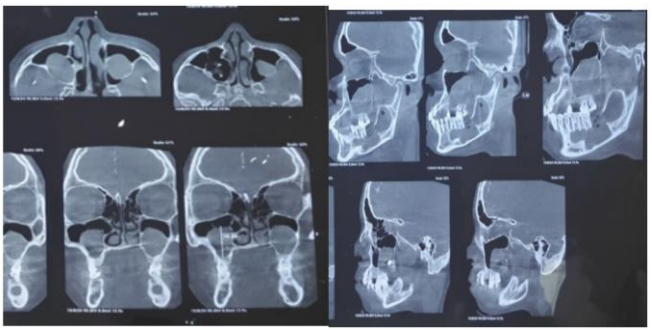

Figure 10: CBCT demonstrating bilateral uniformly hypodense lesions with superior displacement of floor of maxillary sinus and a well-defined radiolucent lesion with incomplete septae involving the left body-ramus region

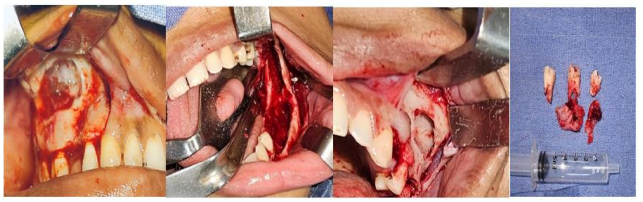

Figure 11: Intraoperative images showing cyst lining and bony defect; extracted teeth with root resorption and surgical specimen

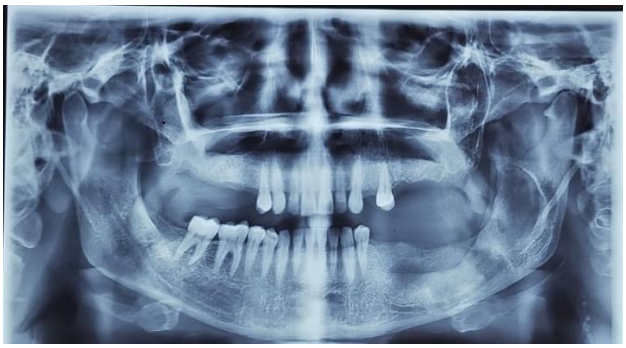

Figure 12: Post-operative OPG demonstrating favourable bone formation

In all cases, provisional diagnosis of multiple OKC was made, later confirmed by histopathological examination.

Given the patient's young age, size, location, and occurrence of primary OKCs, conservative approach was chosen as the treatment modality. Enucleation followed by peripheral ostectomy and chemical cauterization with modified Carnoy's solution and either open or closed packing with iodoform was performed. Impacted and resorbed teeth were extracted. Staged enucleation was done at six-month interval in Case-1.

3. Discussion

Epidemiologically, OKCs account for approximately 7.8% of jaw cysts, and incidence varies from 4 -16.5%. They can occur at any age, with peak incidence between second and fourth decades of life. They can occur spontaneously or as manifestation of a syndrome and can be found incidentally.[1] In our case series, two patients presented at the second decade and one at the third decade, which corresponds to peak incidence. However, in case of syndromic OKCs, the cysts occur at an early age (first decade of life), are more frequently located in posterior sextants of maxilla, exhibit more aggressive behaviour, and have a higher recurrence rate than non-syndromic OKCs. OKCs are seen twice in the mandible, commonly in angle-ascending ramus region (69-83%) and may cross the midline, whereas in the maxilla anterior region is common followed by third molar region.[4]

Multiple OKCs are usually associated with NBCCS. Genetic screening revealed that PCNA and TP53 have a pivotal role in the development of OKC. OKC may also be the result of the combined effects from dysregulation in multiple tumour suppressor genes or oncogenes.[5] Rarely, multiple OKCs are seen without syndromic manifestations, with Brannon reporting an incidence of 5.8%.[6]

Most OKCs are flagged as abnormal on routine dental examination, and patients present early if there is evidence of delayed eruption or if associated with syndromes such as NBCCS, as is the case with 5% of all OKCs. The majority of patients, however, will show up later in life when they start exhibiting symptoms, such as pain, and pus discharge from a secondary infection, or when they are associated with teeth displacement or deformity caused by cortical expansion.[1] In our case series, one of them was asymptomatic and diagnosed on routine radiographic evaluation, and two of the patients were symptomatic, and delayed eruption was evident in one of them.

While they may erode the cortices, OKCs in the mandible tend to grow mesiodistally throughout the length of the bone, leading to minimal expansion of buccal and lingual cortical plates. Asymptomatic patients may have extensive lesions in the mandible that significantly erode cortical plates and involve surrounding structures. Rarely, it causes neurosensory deficit, which presents as lower lip parasthesia. On the contrary, hydraulic expansion of alveolar bone with remodelling, thinning, scalloping, and perforation of the cortices is more commonly seen in large OKCs of the maxilla.[7] OKCs infrequently cause root resorption with documented incidences ranging from 1.3 to 11%. About 40% of OKCs occur in conjunction with the crown of an impacted tooth, and 30% of maxillary and 50% of mandibular OKCs cause buccal plate expansion. Maxillary sinus floor elevation or inferior displacement of mandibular canal and infrequent orbital floor invasion are among effects on adjacent structures.[8,9] All these findings in our cases are in line with the literature.

The multifocal nature of OKCs may be the source of occurrence of multiple cysts, as discussed by Boyne in their study of resected mandibular OKC specimens.[10] Radiographically, an OKC usually appears as a well-defined unilocular or multilocular radiolucency bounded by corticated margins. Small unilocular OKCs may simulate other odontogenic and non-odontogenic cysts.[7]

The objective of management of OKCs is to eliminate the lesion, hence decreasing the chances of recurrence and reducing morbidity. Surgical alternatives encompass enucleation alone (recurrence rate of 60%) or in conjunction with adjuncts such as peripheral ostectomy (26.7% recurrence), Carnoy's solution (17.4% recurrence), cryotherapy, and 5 FU; marsupialization and decompression, marginal or segmental resection both associated with lowest recurrence rates but, should be reserved for multiple recurring lesions, owing to their morbidity.[1,11] Alchalabi et al. reported that extraction of involved teeth showing root resorption can ensure that no remnants of cyst are left behind, thus decreasing the incidence of recurrence.[1]

The recurrence is usually related to satellite cysts left behind, especially in multilocular lesions and permanence of the impacted tooth associated with the lesion.[7] The remaining daughter cysts beyond the lesion's osseous cavity are removed by peripheral ostectomy. This method involves mechanical removal of additional bone (1–2 mm in depth) from the osseous cavity after removing visible lesion.[12] Carnoy's solution is a mixture of absolute alcohol 6 ml, chloroform 3 ml, glacial acetic acid 1 ml, and ferric chloride 1 gm, which penetrates cancellous spaces in the bone,

devitalizing and fixing tumour cells in the cyst cavity. A modified Carnoy's solution was devised devoid of chloroform, as chloroform has been classified as a carcinogen and banned as a therapeutic agent. It penetrates bone to a predictable, time-dependent depth of 1.54 mm in 3 minutes without injuring neurovascular structures.

In large lesions, following enucleation, iodoform formulations such as Bismuth iodine paraffin paste (BIPP), Whitehead's varnish, and Alveogyl are used as packing material, which has an amalgamation of properties to prevent secondary infection and assist in healing, attributed to the release of elemental iodine.[13]

Long-term follow-up is vital for optimal patient outcomes, and all our cases are regularly evaluated, showing good bone healing and regeneration.

4. Conclusion

Although multiple OKCs are associated with syndromic conditions, only a few incidences of multiple OKCs without syndromic conditions have been reported in the literature. They can exhibit aggressive growth patterns, and cause significant bone destruction, necessitating a strategic approach to management. However, there are chances of developing symptoms of syndrome later. Therefore, the absence of the syndrome at presentation does not eliminate the need for vigilant monitoring. Long-term follow-up with periodic imaging is essential for early detection and timely intervention.

5. Source of Funding

None.

6. Conflict of Interest

None.

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Cite this article: Akhilesh AV, Amrutha S, Navyasree KK, Ravikumar A. Non-syndromic, synchronous multiple odontogenic keratocysts: A rare case-series. *J Oral Med Oral Surg Oral Pathol Oral Radiol*. 2025;11(1):21–25.