Case Report

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A Rare Case Report of Odontogenic Keratocyst in a 2-year-old Child

Dentistry Section

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ABSTRACT

Odontogenic Keratocyst (OKC) is a distinct developmental odontogenic cyst with an aggressive behaviour, a high recurrence potential and association with Nevoid Basal Cell Carcinoma Syndrome (NBCCS). The present case involves a two-year-old male who presented with a progressive, painless swelling in the left anterior mandible, first noticed at one month of age. Clinical examination revealed a firm, non tender swelling and Computed Tomography (CT) demonstrated a well-defined unilocular radiolucency with buccal cortical expansion but no root resorption or perforation. Aspiration yielded straw-coloured keratinaceous fluid and subsequent histopathological analysis confirmed the diagnosis of a parakeratinised OKC. The lesion was managed with complete enucleation under general anaesthesia, while preserving adjacent developing tooth buds and avoiding adjunctive chemical cauterisation to minimise potential harm due to the child's young age. The present case is rare primarily due to the exceptionally early age of presentation—a two-year-old—which is well below the typical age range reported for OKCs in the paediatric population, generally 6 to 12 years or older. Additionally, the anterior mandibular location is less commonly reported compared with the more frequent posterior mandibular involvement.

Keywords: Infant, Jaw neoplasms, Mandibular diseases, Paediatric oral pathology

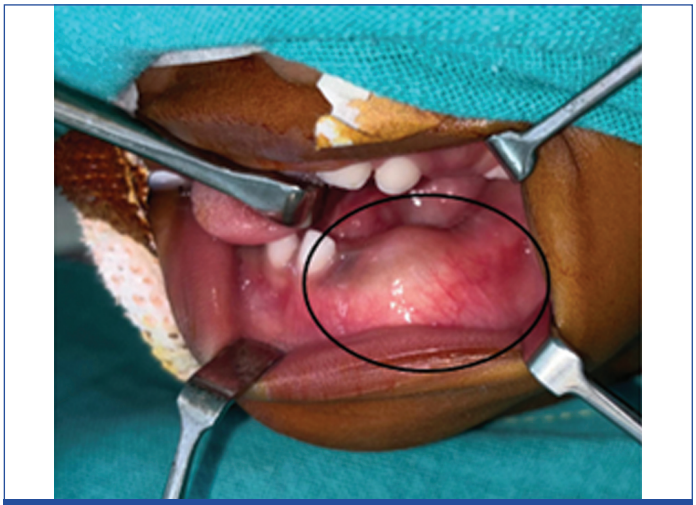
CASE REPORT

A two-year-old male presented to the department of Oral and Maxillofacial Surgery with swelling in the left anterior mandible. The patient had no relevant past medical history. There was a history of unerupted left mandibular teeth. The parents reported noticing the swelling at one month of age and it gradually increased in size over two years.

Extraoral examination revealed facial asymmetry due to a diffuse swelling over the left chin region measuring approximately 3×2 cm. The swelling was oval, with the skin colour similar to adjacent skin, a smooth surface and diffuse margins; pulsations were absent. On palpation, the local temperature was not raised; the swelling was hard in consistency, non tender, with no fluctuations, non compressible, fixed to the underlying bone and nonpulsatile. The overlying skin was free [Table/Fig-1].

[Table/Fig-1]: Extraoral swelling left side of anterior mandible region.

Intraoral examination revealed a stony hard, non tender swelling in the left anterior mandible extending from the midline to the left molar region, measuring 3×2 cm. The adjacent right mandibular deciduous incisors were intact, non mobile and with a mesial tilt of the crown. The overlying mucosa was intact, with no crackling, translucency, ulceration, or infection [Table/Fig-2]. Cervical lymph nodes were non palpable and there were no systemic symptoms.

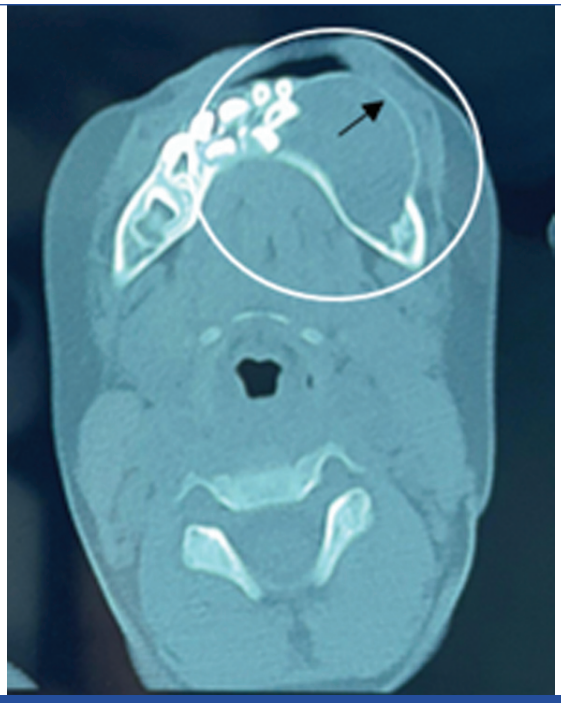


[Table/Fig-2]: Intraoral swelling

Basic laboratory investigations, including Complete Blood Count (CBC), Kidney Function Test (KFT), Liver Function Test (LFT), coagulation profile, Random Blood Sugar (RBS) and testing for Human Immunodeficiency Virus (HIV), Hepatitis B surface Antigen (HBsAg) and Hepatitis C Virus (HCV), with Electrocardiogram (ECG) and chest X-ray, were performed to assess paediatric and preanaesthetic fitness. Orthopantomogram (OPG) was not performed because the CT scan demonstrated a well-defined unilocular radiolucency in the anterior mandible, measuring approximately 2.7 cm in diameter, with adjoining teeth displaced medially and buccal cortical expansion but without perforation or root resorption [Table/Fig-3].

Preoperative aspiration yielded clear, straw-coloured, keratinaceous material and was sent for cytologic evaluation, which showed squamous epithelial cells in a keratinaceous background with a protein content of 3.2 g/dL (less than 4 g/dL).

Based on clinical, radiological and cytological findings, differential diagnoses included dentigerous cyst, eruption cyst, orthokeratinised

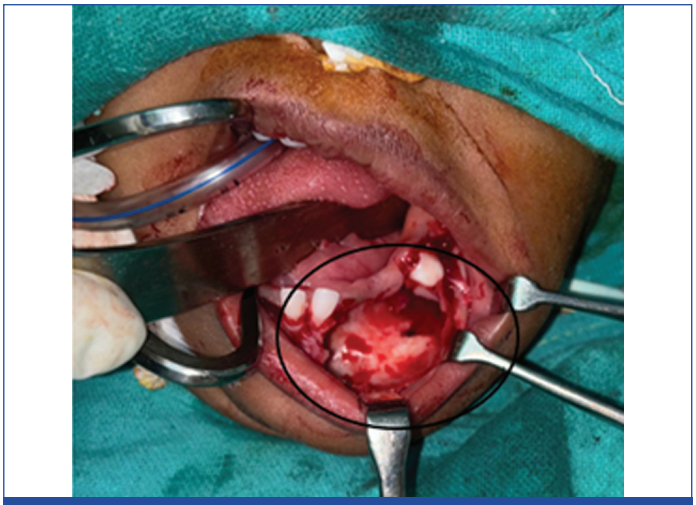


[Table/Fig-3]: Axial CT scan cut showing cystic lesion in left mandibular region with arrow showing intact buccal cortical plate.

odontogenic cyst, gingival (alveolar) cyst of the newborn, lateral periodontal cyst, calcifying odontogenic cyst and glandular odontogenic cyst. Because the patient was uncooperative for biopsy, the plan proceeded under general anaesthesia with the intention of complete excision of the lesion.

Intraorally, a mandibular crestal and crevicular incision was made over the left side from the deciduous right canine to the deciduous left first molar region. The cystic lining over the mandibular alveolar region was exposed. After reflection of a full-thickness mucoperiosteal flap, the cystic lining was exposed. Enucleation of the cyst was performed in toto, carefully without breaching the cystic lining to minimise the risk of recurrence [Table/Fig-4,5]. Peripheral ostectomy or chemical cauterisation was not performed, given the patient's growing age and the concern to preserve vital anatomical structures and developing dentition. The enucleated cyst was sent for histopathological examination. Thorough irrigation was carried out with betadine and saline solution. Closure was achieved by a horizontal mattress suturing technique using absorbable 3-0 Vicryl sutures [Table/Fig-6].

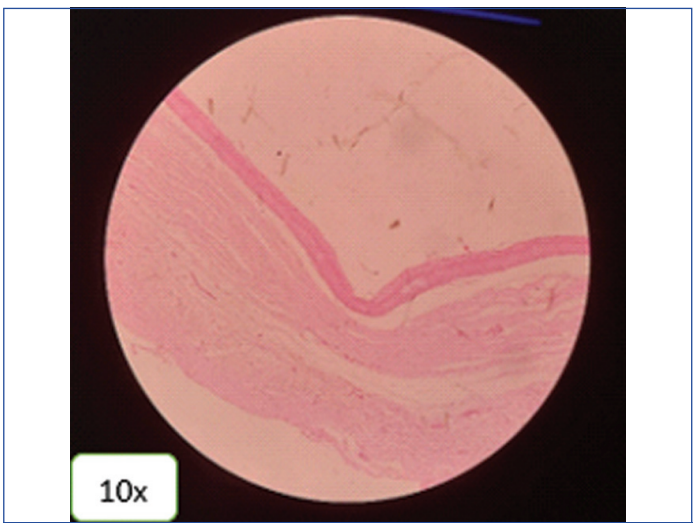
[Table/Fig-4]: In toto excised cystic lesion



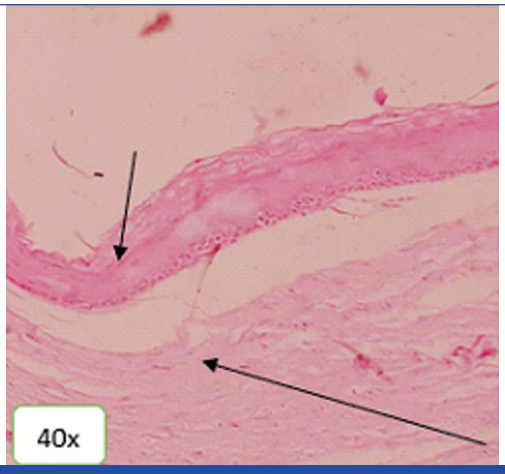
[Table/Fig-5]: Surgical defect in mandible after excision of lesion preserving tooth buds and vital structures

[Table/Fig-6]: Closure by horizontal mattress suturing technique using absorbable 3-0 Vicryl sutures

Histopathology {Haematoxylin and Eosin (H&E)} indicated a parakeratinised stratified squamous epithelial lining with a corrugated surface, palisaded basal cell layer with uniform, hyperchromatic nuclei and a fibrous connective tissue capsule with no inflammation, which was suggestive of a parakeratinised OKC [Table/Fig-7,8]. Postoperatively, no significant complications were observed, including nerve paraesthesia. The patient was followed-up regularly and after six months there was no sign of recurrence and facial asymmetry had improved [Table/Fig-9]; the left mandibular anteriors and molar teeth erupted normally [Table/Fig-10].



[Table/Fig-7]: Microscopic view in H&E, 1x.



[Table/Fig-8]: High power microscopic view with small arrow showing parakeratinised stratified squamous epithelial lining and large arrow showing fibrous connective tissue capsule with no inflammation (H&E, 4x).

[Table/Fig-9]: 6-month postoperative follow-up extraoral picture

[Table/Fig-10]: 6-month postoperative follow-up intraoral picture.

DISCUSSION

The OKC is a developmental odontogenic cyst with aggressive behaviour, a high recurrence rate and potentially considerable bony destruction [1]. It has been argued that the OKC should be classified as a benign cystic neoplasm, renaming it a Keratocystic Odontogenic Tumour (KCOT) due to genetic alterations and increased cell proliferation [1]. Although there is supporting evidence, the term "OKC" continues to be debated but remains widely used, with both names considered synonymous [1]. While small OKCs are usually asymptomatic, larger ones may cause pain, swelling, or drainage. These cysts generally expand intramedullary without appreciable peripheral expansion and are distinguished from other cysts such as dentigerous or radicular cysts [1,2]. They most commonly occur in the posterior mandible and are typically found in individuals between 10 and 40 years of age, with a slightly higher male predilection and a peak incidence in the second to fourth decades of life (54.2%) [2], though rare reports exist as early as the first decade and as late as the ninth decade of life [3,4]. OKCs are radiographically well-defined radiolucent areas with smooth borders and can mimic other cystic lesions such as dentigerous or radicular cysts. They may be multilocular in adults and unilocular in paediatric cases and have been reported to rarely exhibit root resorption as seen with other cysts [1,2].

The present case is particularly notable for its occurrence in the anterior mandible of a young child—an uncommon site and age group for OKCs. The aetiology may involve local irritants causing proliferation of epithelial remnants or genetic predisposition. Impacted teeth or trauma causing chronic irritation may also contribute. CT evaluation and histopathological examination, which showed parakeratinised stratified squamous epithelium with a palisaded basal cell layer and the lesion's characteristic scalloped borders, supported the diagnosis of OKC. Given its high recurrence potential and aggressiveness, surgical enucleation was performed in an attempt to completely remove the lesion. The use of Carnoy's solution was considered but deemed unnecessary due to the lesion's size and location.

Presentations of OKCs in paediatric patients are uncommon and careful evaluation and management are necessary to prevent potential complications [1,2]. The radiographic appearance and absence of root resorption or cortical perforation supported the initial clinical impression of a benign process [1,2]. Ultimately, histopathological analysis remains the definitive diagnostic tool for OKCs, used to establish their characteristic histology and to guide appropriate treatment strategies [1,2]. The cyst wall is usually thin and friable, rendering enucleation difficult [1]. Either clear, serum-like fluid or keratinaceous debris may be present in the lumen [1]. Microscopically, OKCs show stratified squamous epithelium, usually six to eight cells thick, with a corrugated parakeratinised luminal surface and a palisaded basal cell layer of hyperchromatic cuboidal or columnar cells [1,2].

Occasionally, satellite cysts and odontogenic epithelial islands are found in the fibrous wall in 7-26% of cases, which may explain the high recurrence rate of OKCs [1,2]. Epithelial features can be distorted by inflammation, with rete ridges and loss of the palisaded basal layer, thus complicating diagnosis unless sections elsewhere retain the features characteristic of the classic histology [1,2]. The morphologically distinct orthokeratotic variant of OKCs, which lacks the hyperchromatic palisaded basal cells, demonstrates markedly different clinical behaviour and should no longer be classified as a variant of OKCs [1,2]. Understanding these histopathologic nuances guarantees accurate diagnosis and guides treatment to prevent recurrences.

Management of OKC in paediatric patients is particularly challenging due to the need to balance effective treatment with preservation of vital anatomical structures and developing dentition. Recurrence is often attributed to satellite cysts within the fibrous capsule or remnants

of cystic epithelium left behind during surgery. To minimise this risk, complete removal of the cyst lining and careful management of peripheral osseous tissue are essential. Enucleation with adjunctive techniques, such as peripheral ostectomy or chemical cauterisation using Carnoy's solution, has effectively reduced recurrence rates. More aggressive strategies, including resection with or without continuity defects, are typically reserved for extensive or recurrent lesions. Conservative methods, such as marsupialisation followed by enucleation, while less invasive, carry a higher recurrence risk due to the potential persistence of pathological epithelium. Additional adjunctive therapies, like cryosurgery and open packing, have shown mixed results [5].

Paediatric cases tend to have more favourable outcomes than adults, where recurrence rates often exceed 30% [6,7]. Most paediatric OKCs are typically diagnosed in children aged six years and older, as noted by Deboni MC et al., Kumar V (6 years) and Singh M and Gupta KC (11 years) [8-10]. Compared with these cases, the patient in the present case was significantly younger, emphasising the rarity of this presentation and the diagnostic challenges in infants and toddlers. While enucleation with adjunctive procedures such as Carnoy's solution or peripheral ostectomy is commonly employed in older children to reduce recurrence risk, these methods carry potential risks to developing tooth buds and nearby vital structures [11]. In the present case, complete enucleation without adjunctive chemical cauterisation was chosen to minimise morbidity and preserve the developing dentition, guided by radiographic evidence and cyst accessibility. Long-term follow-up is necessary to monitor for recurrence and ensure a successful outcome.

CONCLUSION(S)

From this rare case of an OKC in a two-year-old patient, the need for early identification of the cyst, accurate evaluation and appropriate surgical treatment in the paediatric population is underscored. This lesion was atypical in occurring in the anterior mandible of a child; however, enucleation proved effective with no relapse after six months. The present case highlights the importance of clinical, radiographic and histologic examination and long-term follow-up to address the possibility of relapse and to improve the child's health and development.

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