

Recurrence of an Extensive Maxillary Odontogenic Keratocyst: A Rare Case Report

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Abstract

Odontogenic keratocyst is a developmental odontogenic cyst arising from dental lamina. It is unique amongst cysts due to its aggressive clinical behavior, invasion to surrounding tissues, and increased tendency to recurrence. It is usually found in the mandibular posterior region, but on rare occasions it can be seen in the maxillary jaw as well. The present case report shows such a case of maxillary odontogenic keratocyst in an 11-year-old male patient in relation to the right maxillary sinus region associated with canine. It was enucleated using Caldwell Luc approach under general anesthesia. The histopathological report showed the lining of the odontogenic epithelium, subepithelial zone showed keratinous debris, hemorrhage, fibrocollagenous tissue, and bony trabeculae. The same patient reported 6 months later with recurrence in the same region. The cyst was enucleated again and followed with Bismuth Iodine Paraffine Paste (BIPP) pack in situ to prevent recurrence. Due to the aggressiveness and tendency of the cyst toward malignant transformation, an early and accurate diagnosis is required for treatment and periodic follow-ups are imperative to check the recurrence of the disease.

Keywords

- ▶ keratocyst
- ▶ maxillary
- ▶ odontogenic
- ▶ rare tumor
- ▶ recurrence

Introduction

Odontogenic tumors are derived from the epithelium which is associated with the development of dental structures and may have stratified squamous epithelium or respiratory epithelium in cases of some developmental cysts. Odontogenic keratocyst (OKC) is the developmental cyst of oral cavity and is derived from the remnants of dental lamina. The lesion was reclassified by WHO as Keratocystic Odontogenic Tumor (KOT) due to several factors including its locally destructive nature, high recurrence rate, presence of daughter cysts in the underlying connective tissue and the association with the action of PTCH, a tumor suppressor gene leading to proliferation-stimulation effect of SMO

(oncogene).¹ The biological behavior is very common to benign neoplasm, due to the presence of multiple layers of cells in lining exhibiting a basal layer of palisaded cells and having a surface of parakeratin.

Keratocysts are most commonly found in mandible (70%–80%) at the angle between jaw and mandibular branch and in maxilla in the area of the third molar and canine.^{2,3} The anterior maxilla (canine region) is the most common site for the occurrence of OKC.⁴

The lesion grows mainly in the antero-posterior dimension without deforming the jaw skeleton. The rapid growth is due to the high activity of epithelial cells stimulating the osteolytic activity of prostaglandin in the cell population of the cyst lining.⁵

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The present case report describes a case of maxillary odontogenic keratocyst arising in the canine region in an 11-year-old male patient.

Case Summary

An 11-year-old male patient reported to the department of otorhinolaryngology, Balrampur Hospital, Lucknow, with a complaint of pain and swelling on the right side of face since last 1 year. The pain was dull, continuous, and non-radiating in nature. The patient also reported with blurring of vision from the right eye since two months associated with superior deviation of the right eyeball with proptosis and right side nasal obstruction. The swelling was insidious in onset and gradually progressive.

On examination, it was seen that the swelling was hard in consistency, extending from the infraorbital border to the upper lip with prominent malar eminence and intraorally, it extended from the right upper gingivobuccal sulcus to the hard palate. On anterior rhinoscopic examination, no nasal pathology was detected. On diagnostic nasal endoscopy, narrow nasal cavity was seen on the right side along with mucoid discharge in the middle meatus. Maxillary sinus osteum was obliterated.

On contrast-enhanced Computed Tomography (CT) face examination, expansible lesion approximate 54×46 mm in the right maxillary antrum filled with hypodense soft tissue attenuation/high attenuation fluid lesion with no significant post contrast enhancement, causing remodeling and thinning of walls of right maxillary antrum was observed. Superiorly, the lesion was bulging in the right orbit and ethmoidal air cells on right side compressing orbital structures and causing proptosis of the right eye ball. Lesion was involving right half of the upper alveolar arch with tooth structure in the lesion along the floor of maxillary antrum and the posterolateral wall bulging into the right infratemporal fossa and medial wall bulging in the nasal cavity on the right side.

On Magnetic Resonance Imaging (MRI) examination (**Fig. 1**), mucosal thickening was seen in bilateral frontal sinuses, ethmoid air cells, left maxillary sinus, and sphenoid sinus. Moderate size (52 [vertical] \times 52 [AP] \times 51 mm [TR]) well-defined T1 and T2 hyperintense cystic lesion was seen in

the right maxillary sinus. Marked thinning of the overlying bone was seen. No extra-osseous soft tissue component was seen. Remodeling of the right orbital floor was seen producing a bulge in the inferior part of the right orbit with mild antero-superior displacement of the right eye ball. Inferior rectus was effaced by the remodeled bone. No soft tissue component was seen in the orbit. The rest of the extra-ocular muscles and right optic nerve were normally visualized. Medially, it was bulging into the middle meatus. Right nasal turbinates were effaced. Mild deviation of the nasal septum was seen toward the left. Inferiorly, it was abutting the root of the right upper molars.

Surgical Procedure

The patient underwent enucleation using the Caldwell–Luc approach under general anesthesia (**Fig. 2**). The endoscopy of the sinus was performed to evaluate any remnants present (**Fig. 2**). The tissue was sent for histopathological examination and the histopathological report showed a cyst lined by odontogenic and epithelium palisaded basal layer. The cyst was seen filled with keratinized debris that confirmed the diagnosis of OKC. Postoperative period was symptom free and the patient was symptom free in the subsequent follow-ups as well.

After 6 months, patient presented with right side facial swelling in the cheek area rapidly growing and causing pressure symptom over the face. MRI was performed which revealed a cystic lesion in the right maxillary sinus. The patient was taken up again for surgery under general anesthesia for enucleation using the Caldwell–Luc approach. Histopathological examination revealed odontogenic epithelium with a subepithelial zone showing keratinous debris, hemorrhage, fibrocollagenous tissue, and bony trabeculae confirming odontogenic keratocyst.

Bismuth Iodine Paraffine Paste (BIPP) packing of the right maxillary antrum was done to prevent recurrence and infection in the postoperative period.

Discussion

Odontogenic keratocyst is the third most common odontogenic cyst occurring in the oral cavity after dentigerous cyst

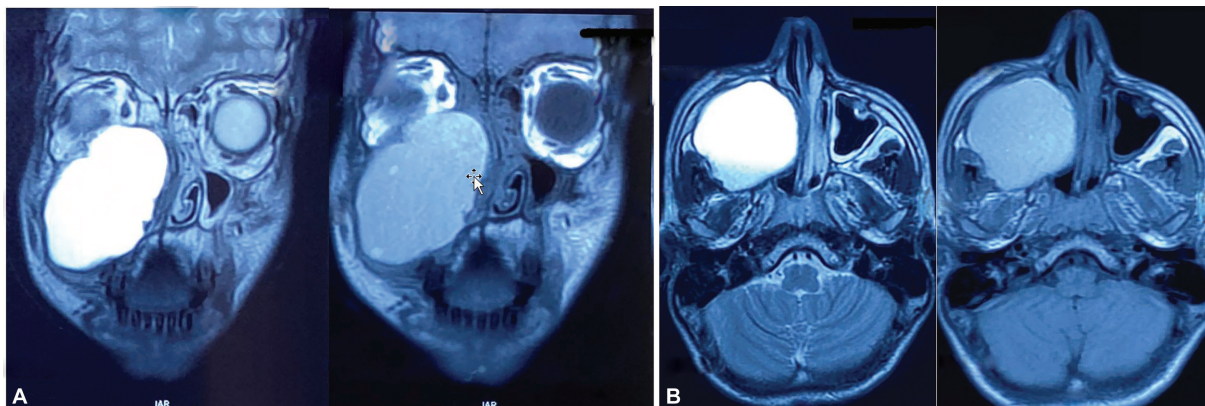


Fig. 1 MRI scan of the patient showing a cystic lesion of moderate size 52 (vertical) \times 52 (AP) \times 51 mm (TR).

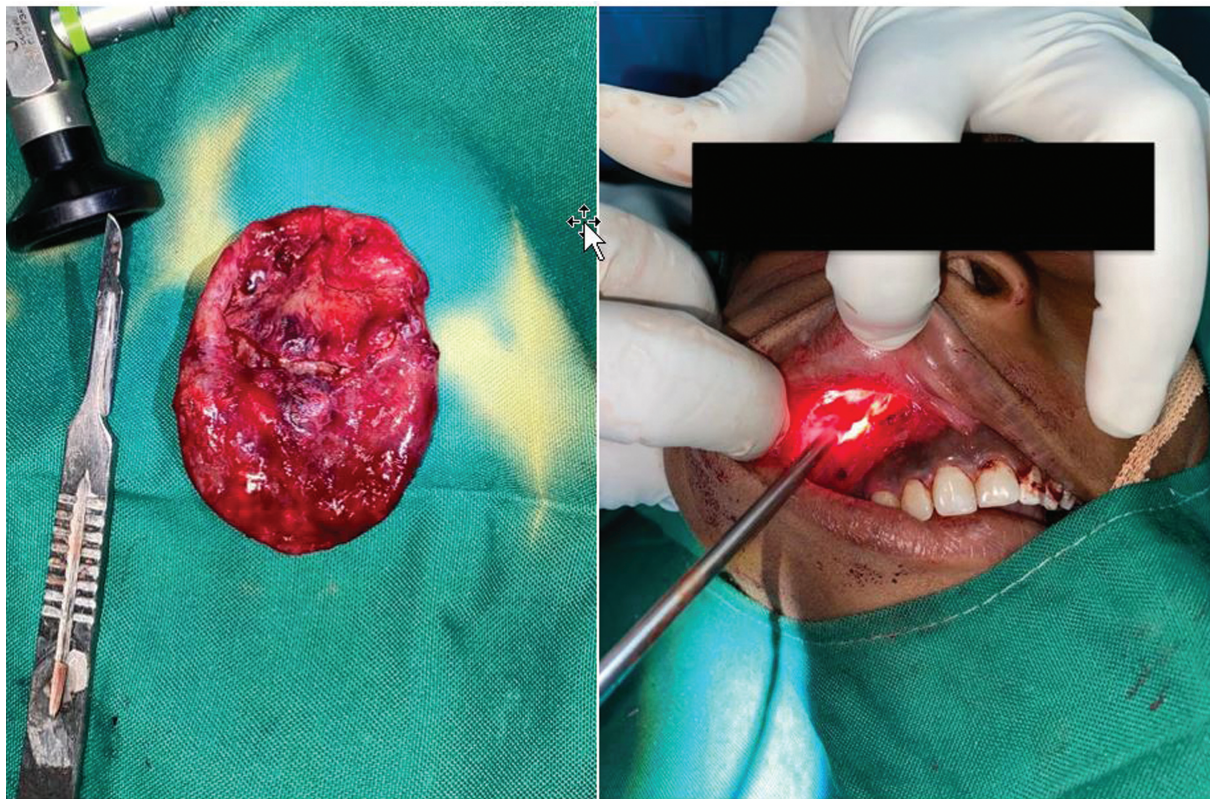


Fig. 2 Enucleation of the cyst with intact cystic walls and endoscopic examination of the maxillary sinus for any residual tissue.

and periapical cyst.⁴ The involvement of maxillary sinus and canine region by OKC is as rare as < 1%.⁶ The origin of OKC in the maxillary sinus is controversial, presumably arising from the entrapment of odontogenic epithelium within the sinus because of the close anatomic relationship between the dental lamina and developing antrum or the primordium of the canine and the floor of the sinus.⁶ OKC involves both the jaws; the mandible is more often involved than the maxilla. In the maxilla, it is seen most commonly in the canine area, followed by third molar tuberosity and anterior maxilla. In most of the cases, it presents as a periapical lesion. Here, we present a case that occurred in quite younger age, and the lesion was in the sinus similar to the cases reported by Sheethal et al⁶ and Silva et al.⁷

Radiological imaging, mainly CT and, in selected cases, MRI, plays an important role in the diagnosis and management of OKCs. Although radiological imaging does not always provide a specific diagnosis, the knowledge about typical and atypical radiological features of OKCs is essential for their diagnosis and treatment planning. In particular, the combination of clinical and radiological findings is useful in evaluating the extent of the lesions and the relationships with adjacent structures.⁸ OKCs in the maxilla more frequently present a hydraulic expansion of the alveolar bone with remodeling, thinning, scalloping, and perforation of the cortices. In addition, when OKCs originate from the alveolar bone subjacent to the maxillary sinus, its floor is lifted and lumen is reduced.⁹ Various surgical options have been considered, including enucleation alone or associated with adjunctive measures (ostectomy,

Carnoy's solution, cryotherapy), marsupialization and decompression, marginal or segmental resection.¹⁰

Recurrence rates in the literature range from 2.5% to 62.5%. The relatively high recurrence rate, especially after conservative surgery, make it necessary to perform a periodic radiographic monitoring of patients with surgically treated OKCs, at least for the first 5 years.⁸ For recurrence prevention, Carnoy's solution (a cauterizing mild penetrating agent that is used to eliminate possible vital cells left behind in the defect) is used along with endoscopy-assisted evaluation for any residual cyst particles. BIPP packing of the maxilla after enucleation can also be done.

The incidence of malignant transformation of odontogenic cysts has been estimated to be ~0.12%, with keratinizing cysts demonstrating a higher incidence than nonkeratinizing cysts.¹¹ Therefore, periodic recalls of such patients are very necessary to prevent recurrence and malignant transformation along with preventing the disability caused due to such an extensive lesion.

Conflict of Interest

None declared.

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