

A rare case of multiple odontogenic keratocyst in a pediatric patient

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Abstract

The odontogenic keratocyst is a developmental cyst of maxillofacial region. It has characteristic histopathological and clinical features. The cyst is very famous for its aggressive behavior and high recurrence rate. Recurrence rates reduced when more meticulous surgical treatment is done. The occurrence of multiple odontogenic cysts in jaw bones of a non syndromic pediatric patient is not a very common entity.

We hereby report an unusual case of multiple OKCs in an otherwise healthy pediatric patient of mixed dentition period associated with no other dental anomalies. Management approach consisted of enucleation with curettage. Follow-up of 1 year is completed with repeated clinical and radiographic examinations in 6 months interval with no evidence of recurrence.

Keywords: Cyst, Odontogenic Keratocyst, Keratocystic Odontogenic Tumor, Recurrence

Introduction

Odontogenic keratocysts (OKCs) are benign odontogenic cystic neoplasms occurring intraosseously, usually in the posterior part of the mandible. Odontogenic keratocyst (OKC) is a developmental odontogenic cyst. It is thought to originate from the dental lamina. It was first noted in 1956 by Phillipsen ^[1]. OKC is well recognized by its aggressive behavior, rapid growth and high tendency to invade the adjacent tissues including bone. It tends to recur and occasionally is associated with the basal cell nevus syndrome ^[2]. Despite the benign appearance of OKCs, they can behave aggressively and grow infiltratively. If not removed completely, they tend to recur in up to 62.5% of cases. ^[3,4]

Redesignation of the OKC as the KCOT (Keratocystic Odontogenic Tumor) by the World Health Organization (WHO 2005) is based on the well-known aggressive behaviour of this lesion, its histology and new information

regarding its genetics. But again WHO has categorized OKC into Odontogenic and non-odontogenic developmental cysts (2017). This new WHO classification of Head and Neck pathology re-classified OKC back into the cystic category. It is no longer considered a neoplasm as the evidence supporting that hypothesis like clonality is considered insufficient.^{5,6}

Odontogenic keratocyst (OKC) have low prevalence in children. It usually appears in the second, third and fourth decades of life (54.2%) and rare cases reported as early as the first, and as late as the ninth decade of life^[7]. Treatment of OKCs is still a debatable subject because of their great tendency to recur^[7]. Occasionally decompression or marsupialization is the first surgical manipulations in OKC and when the volume becomes smaller in size, enucleation has to be performed.^[5,6]

It has been proposed that difficult operative access, leaving affected teeth in place after treatment, cystic satellite formation, and the association with Gorlin-Goltz syndrome are correlated with a higher recurrence of OKCs⁵. Studies reveal that 5.8% of multiple OKCs presented without any features of a syndrome, 8.1% were associated with Nevoid basal cell carcinoma syndrome, and 7.6% of them had recurrences¹⁰

Case report

A 12 year old boy reported to our centre with complaints of pain and swelling on the left side of the face since 2 weeks. He had consulted in some other centre and took parenteral antibiotics and analgesics, but only temporary relief. So he was referred to our centre for proper diagnosis and management.

On extra oral examination there was a single diffused swelling in the left side of the face extending from the left infraorbital region to upper lip. The swelling was firm in consistency and was tender on palpation. Intraorally swelling extended from upper left lateral incisor region to

second molar region with obliteration of the left buccal sulcus. There were no carious teeth. On aspiration pus was present. A provisional diagnosis of infected cyst was made and patient was asked to take OPG.

OPG revealed well defined radiolucency in the left maxilla extending from distal side of lateral incisor to the mesial side of first permanent molar and superiorly up to left maxillary sinus. The unerupted canine and first premolar was very close to the lesion and second premolar was inside the lesion. OPG also showed well defined radiolucent lesion in the bilateral angle of mandible, pushing the tooth bud of mandibular third molars.

CT scan was advised to assess the extent of lesion. A well defined cystic area with water density measuring 3x 2.2 cm, expanding the left maxilla is found in relation to a unerupted premolar tooth. Two well defined cystic areas with water density measuring 1x 1.4 cm in the left and 1.1x 1.2 cm in the right angle of mandible in relation to unerupted wisdom tooth were noted. There was no evidence of sclerosis of adjacent bone.

Blood investigation reports were within normal limits and there was no family history of any syndromes

Based on the clinical and radiological findings a provisional diagnosis of Odontogenic Cyst was made and the patient was advised to undergo Surgical management under GA. Since the bilateral angle lesions were asymptomatic, patient's parents were not willing to do surgery in the mandible at first. Due to the patient's parent's personnel constraints, surgery was planned under Local anesthesia in three stages.

Surgical Procedure

Under Local Anaesthesia the maxillary lesion was exposed by trapezoidal incision. Mucoperiosteal flap was raised. Lesion exposed. Cyst enucleation done. The unerupted tooth bud of second premolar was found inside the lesion and it also came out along with the cyst.

Hemostasis achieved and the closure was done with 3-0 black silk. The specimen was sent for Histopathological examination and was reported as “Odontogenic Keratocyst”. Healing was uneventful.

Cyst enucleation from the left angle of mandible was done 2 months after the first surgery and from the right angle 4 months after the first surgery under LA. In both sides the tooth bud of third molars were also removed along with the cyst. Healing was uneventful in both the sides and the Histopathology report was ‘Odontogenic Keratocyst’.

Six months after the maxillary cyst enucleation an OPG was taken which showed no recurrence and healing of bone from all sides. Patient is kept under regular follow ups due to the high recurrent nature of OKC.

Fig 1: Pre op OPG



Fig. 2: Pre op Maxillary Lesion



Fig.3: Pre op CT-1

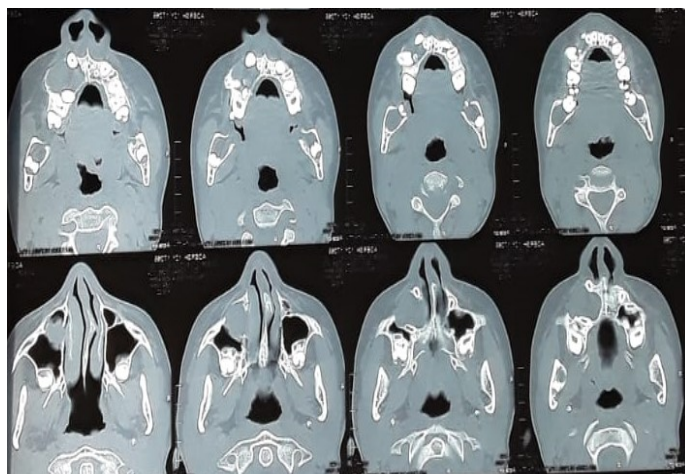


Fig. 4: Pre op CT-2

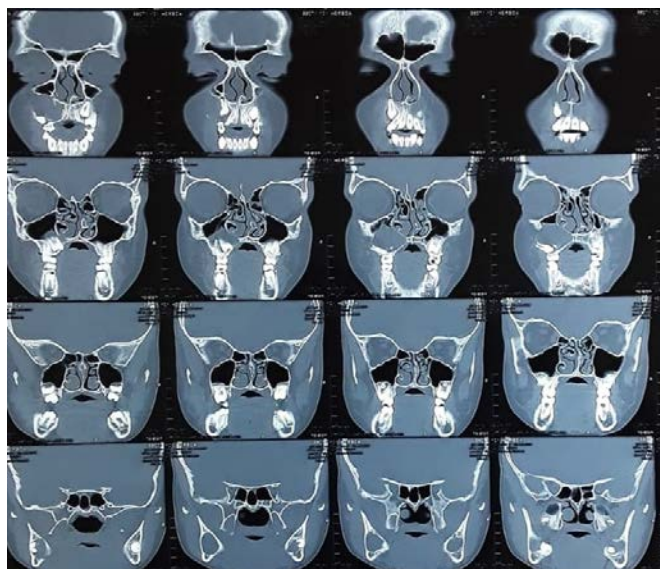


Fig. 5: Intra Op Maxilla



Fig. 6: Enucleated lesion - Maxillary



Fig.7: Intra op- Left angle of Mandible



Fig. 8: Enucleated lesion – mandibular Left Angle

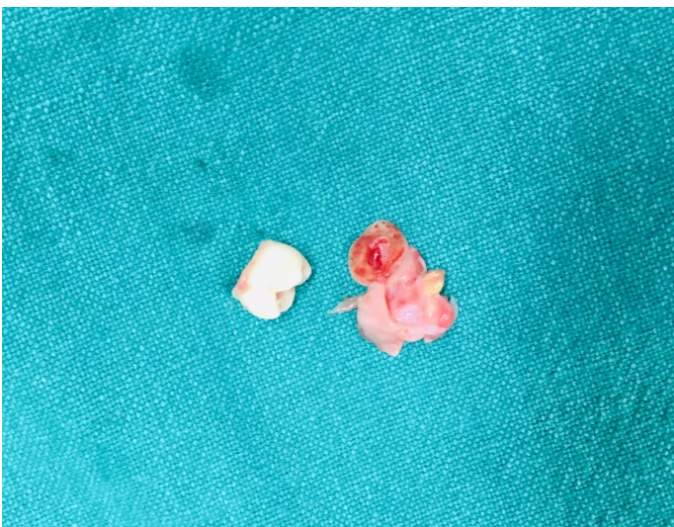


Fig. 9: Histopathology

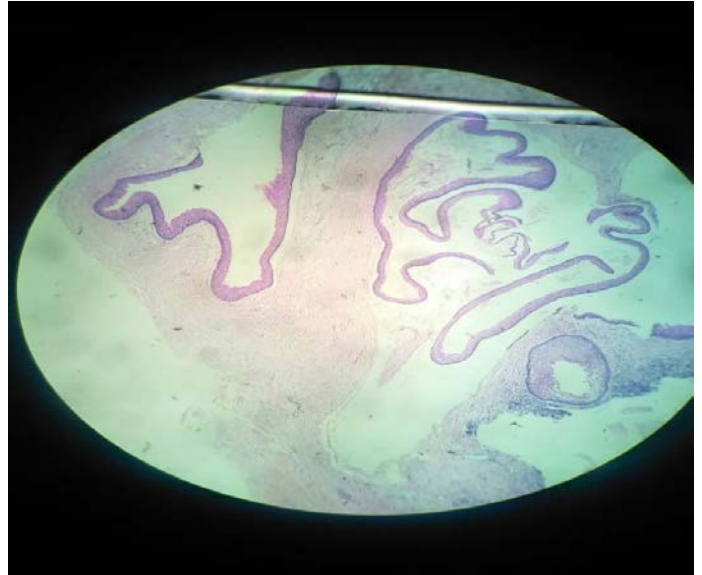


Fig. 10: Post op OPG



Discussion

Management of odontogenic keratocyst particularly in children still remains a subject of debate. The decision on the treatment option should be established on the size and site of the lesion, recurrence status and radiographic evidence of cortical destruction, presence of associated syndromes and histologic variety as it is well known that parakeratotic type is more common in young age. Guided by the aforementioned characteristics, there is a general accord in the literature, for aggressive surgical approaches with complete lesion eradication, such as resection with or without reconstruction. These aggressive manipulations

may lead to deformities, which may lead to serious psychosocial outcomes, especially in adolescents. Hence, the priority has been given to the reduction of complications as much as possible [6,7]

Odontogenic keratocysts require dental follow-up visits, including a periodic radiographical evaluation every 6 months, especially in childhood and early adolescence due to its high recurrence rate.¹¹

Conclusions

Although pediatric jaw lesions are uncommon, symptoms such as swelling could indicate potential pathologic findings and require panoramic examination. Management of pediatric jaw lesions should consider the biologic behavior of the lesion, maxillofacial development, and growth. Enucleation combined with pharmacologic therapy is a promising strategy for the management of aggressive central giant cell tumors in children.¹²

The suspected diagnosis of an OKC is possible with radiological images and should be confirmed histopathologically. Due to the high recurrence rate, regular radiological follow-ups is important for treatment success. A possible presence of Gorlin-Goltz syndrome should be suspected in case of multilocular cysts in pediatric patients, even if further signs of the syndrome are not discernible at first sight and all pediatric patients with OKC diagnosis

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