

Non -Syndromic Multiple Odontogenic Keratocyst – A Case Report And Literature Review

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ABSTRACT

Odontogenic Keratocystic Tumour was reclassified as Odontogenic keratocyst (OKC) in the recent edition of the World Health Organization (WHO) Classification of Head and Neck Tumors, published in January 2017. OKCs makes up 10% of all odontogenic cysts and are benign in nature. Commonly localized to the posterior mandibular area, they are characterized by aggressive behavior with a rather high recurrence rate. These are linked to syndromes as well like the nevoid basal cell carcinoma syndrome. The diagnosis is confirmed through FNAC, advanced radiographic imaging and biopsy. This case report presents a rare manifestation of multiple OKC in a non-syndromic patient.

Key words: Odontogenic Keratocyst, Odontogenic Keratocystic Tumour, Odontogenic cyst, Keratocyst, Jaw cyst, Jaw diseases.

INTRODUCTION

A benign uni- or multicystic intraosseous lesion of odontogenic origin, keratocystic odontogenic tumor (KCOT) is known as Odontogenic Keratocyst (OKC) as per the World Health Organization categorization (2017).^{8,12} It clinically presents as a swelling originating in the mandibular posterior region and is aggressive in nature with high probability of recurrence rate. Multiple OKCs are uncommon, and when they do manifest, they present in tandem with syndromes such as orofacial digital syndrome, nevoid basal cell carcinoma syndrome, Ehler-Danlos syndrome, and Noonan syndrome. According to various systematic reviews, 8.1% of multiple OKCs were in association with nevoid basal cell carcinoma syndrome alone. This case report elucidates on a presentation of multiple OKCs in a non-syndromic patient.

CASE REPORT:

A 17 years old female patient reported to the Department of oral medicine and radiology OPD with a chief complaint of pain in lower right posterior teeth region for the past 8 months. The history of associated pain was moderate in intensity, intermittent in nature and pricking in character. The associated pain radiated to the forehead in the side of interest on chewing. The patient did not give a history of associated swelling in the region of complaint. The patient had referred a local dental practitioner for the same complaint two months prior, for which she was prescribed medication and relief in symptoms achieved. Her past medical history, dental history and family history were non-contributory. On intra oral examination, full complement of twenty eight teeth was observed. Additionally, discoloration owing to dental caries was noted on 27,36 and 37. In the region of complaint, 46 demonstrated dental caries and was positive for tenderness on percussion. A retained deciduous dentition 85 was noted. Bi-cortical expansion was noted bilaterally in the mandibular posterior region. It extended from the premolar region and second molar region, right and left side respectively. The expansion manifested more on the buccal aspect bilaterally with no break in the cortication. There was no displacement or mobility of teeth. A provisional diagnosis of impaction with dentigerous cyst was eluded. Under investigatory panel, as a screening radiograph, an OPG was advised. This radiograph demonstrated bilateral radiolucent lesions in posterior mandible. On the right side, it extended from the distal surface of 44 to the ramus, involving the entire ramus measuring

approximately 4x3 cm, antero-posteriorly. Superior-inferiorly, it extended from the alveolar ridge height to 0.5cm above the border of mandible. For three-dimensional advanced imaging, computed tomography was advised.



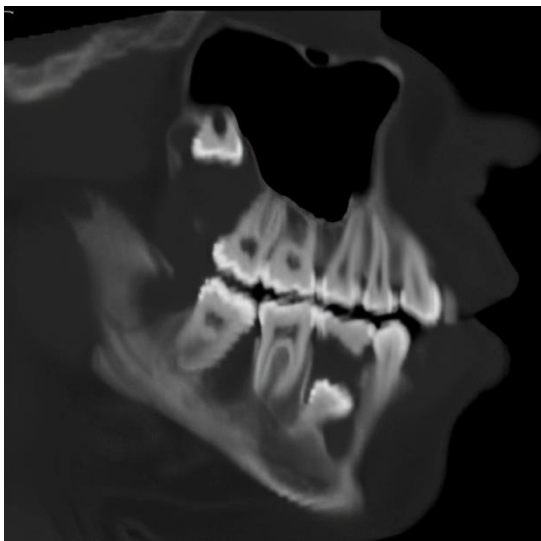
Figure 1 : Extra Oral picture



Figure 2 : Intra oral swelling



Figure 3 : orthopantomogram revealing multiple radiolucencies.



DISCUSSION:

As an aggressive intra-osseous lesion; odontogenic keratocyst manifests in the maxillofacial region and originates from the basal cell layer of the oral epithelium or the dental lamina.^{12,10} Compared to male patients (47%), female patients (53%) had a higher prevalence of OKC. In terms of anatomical localization, the angle and body of the mandible had the highest prevalence of OKC (32%), followed by the ramus and angle (24%), mandibular body (22%), maxillary posterior area (10%), maxillary anterior region (7%) and mandibular anterior region (5%). Histologically there was high prevalence of okc with parakeratinized pattern (87%), whereas only (13%) had orthokeratinized pattern.⁵ An unerupted tooth is present in conjunction with the lesion in roughly 40% of instances.⁷ It commonly presents during the second and fourth decades of life. However, when presenting in the child population, it is often seen in multiples and in association with NBCC.¹⁵ In terms

of ethnicity, it is prevalent in the Caucasian with a ratio of 1:6:1. Morphologically, addition to crossing the midline, mandibular cysts typically develop in the angle-ascending ramus region (69 to 83%) of the mouth. Whereas the pre-maxilla, nasal floor, upper wisdom teeth region, and maxillary sinus may be affected by maxillary OKCs.^{4,13}

Clinically, majority of OKC cases are asymptomatic unless they become secondarily infected. OKC's can grow in large sizes without any prominent bone swelling. It is usually identified through standard radiographic investigations (i.e.) orthopantomogram.¹⁴ Radiographically, OKC frequently extends into the mandibular ramus in both the horizontal and vertical axes, giving rise to a profile resembling a dumbbell. OKC may be unilocular, but it commonly appears multilocular.¹ Multiple case series report OKCs discovered in an apparent dentigerous connection with impacted wisdom teeth. Yet, despite the pericoronal tissues being continuous with the cyst capsule, the associated teeth's crowns are frequently detached from the cystic lumen.⁸

OKC grows from the surrounding dental lamina in the jawbones or could come from the basilar cell layer lining the oral epithelium. High proliferation rates, which exhibit significantly higher expression of proliferating cell nuclear antigen (PCNA), Ki-67, overexpression of (the antiapoptotic protein) Bcl-2, and MMPs 2 and 9 are characteristics that may contribute to the etiopathogenesis of OKCs.³

The management of OKC has always been debatable since it depends on a number of variables, including the patient's age, the size and location of the cyst, radiological characteristics, histological characteristics, and the likelihood of recurrence.¹⁰ The management of OKC can be done in one of two ways: conservatively or aggressively. Enucleation and marsupialization are part of the aggressive method, whereas peripheral ostectomy, chemical curettage with Carnoy's solution, cryotherapy, and electrocautery and resection are part of the aggressive strategy.² Liquid nitrogen is used in cryosurgery, which has the unusual ability to devitalize the bone in place while maintaining the inorganic skeleton. Both of these methods are recommended for eliminating any epithelial remains and dental lamina found within osseous keratocysts in order to lower the frequency of recurrences. In particular for retromandibular trigone, posterior maxillary, and cortically perforated lesions, excision of overlying mucosa when combined with enucleation followed by chemical cauterization using Carnoy's solution or liquid nitrogen yields the lowest recurrence rate.¹⁰

CONCLUSION:

In conclusion the authors would like to reiterate that, if a patient presents with multiple OKCs it is mandate to evaluate for other syndrome-related symptoms. Non-syndromic patients should continue to receive regular follow-ups because the symptoms of a syndrome often don't manifest until later in life. The management of young patients as conservatively as possible in keeping with the safety strictures is the key to remember.

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