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Non Syndromic Odontogenic Keratocyst: A Case Report

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Abstract: *Odontogenic keratocyst (OKC) is common odontogenic cyst with an aggressive clinical behavior and high recurrence rate. OKC is known for its rapid growth and its tendency to invade the adjacent tissues including bone. Multiple OKCs are usually seen in association with nevoid basal cell carcinoma syndrome (NBCCS) but approximately 5% of patients with OKC have multiple cysts without concomitant syndromic presentation. We report rare case of multiple odontogenic keratocyst involving all four quadrants with more emphasize on the clinical, radiological, and histopathological features of this cyst and its surgical management.*
Keywords: *Odontogenic keratocyst (OKC); Keratocystic odontogenic tumor (KCOT); nevoid basal cell carcinoma syndrome(NBCCS); Gorlin-Goltz syndrome.*

I. Introduction

The odontogenic keratocyst is a distinctive form of developmental odontogenic cyst that deserves special consideration because of its specific histopathologic features and clinical behavior. This cyst shows a different growth mechanism and biologic behavior from the more common Dentigerous cyst and Radicular cyst.[1] There is general agreement that the odontogenic keratocyst arises from cell rests of the dental lamina.[1-3] Therefore the term ‘‘laminal cysts’’ was even suggested by Toller.[3] Odontogenic keratocysts (OKCs) first identified and described in 1876 and further characterized by Phillipsen in 1956.[2,4-6] Hansen designated ‘keratocyst’ to describe any jaw cyst in which keratin was formed to a large extent and also suggested the histologic criteria necessary to diagnose OKC in 1963.[7] Several investigators suggested that OKCs must be regarded as a benign cystic neoplasm rather than a cyst.[8,9] In 2005 the WHO working group considered odontogenic keratocyst (OKC) to be tumor and recommended the term Keratocystic odontogenic tumor (KCOT), separating lesion from the orthokeratinizing variant. Clinically the parakeratinizing lesions are characterized by aggressive growth and tendency to recur after surgical treatment.[1, 7-10] It occurs as solitary or multiple especially in association with nevoid basal cell carcinoma syndrome, In 1960, Gorlin and Goltz initially described the simultaneous existence of multiple basal cell carcinomas, multiple OKCs of the mandible and the maxilla, bifid ribs, and other variable manifestations that comprise the basal cell nevus bifid rib syndrome.[6,11] OKCs constitute about 3% - 21.5% of odontogenic cysts. The peak incidence is during the second to fourth decades of life. [2,12-14] Odontogenic keratocysts may occur in any part of the upper and lower jaw with the majority occurring in the mandible, most commonly in the angle of the mandible and ramus.[2,5,13,14]A localized asymptomatic swelling is the most common sign; spontaneous drainage of the cyst into the oral cavity and teeth mobility are also common. Radiographic appearances of OKCs are variable from a unilocular or a multilocular radiolucency with scalloped and well-defined margins to soap-bubble or honeycomb radiolucency.[2, 15] Considerable controversy exists regarding the proper management of these lesions. There are proponents of “conservative” or “aggressive” methods of treatment. Meiselman et al consider conservative therapies to include “enucleation, curettage, and marsupialization.”[16, 17] Williams et al defines aggressive treatment as “that which is used in addition to enucleation, and includes curettage (mechanical, physical and/or chemical) and /or resection with or without loss of jaw continuity. [16, 18]

As mentioned, there are most of the cases of solitary OKC without association of any syndrome. Whereas multiple odontogenic keratocysts are often presented with associated syndrome. We reported rare case of multiple odontogenic keratocyst involving all four quadrants in non syndromic patient and which was managed conservatively.

II. Report Of Case

A 20 years old male patient reported with swelling over the right lower facial region and left mandibular body region which was noticed by the patient approximately three month back. Swelling was painless, slow growing without any discomfort. Patient also complains of pus discharge from right side of mandible with foul breathing.

Clinical examination showed buccolingual expansion of the left mandible. The lesion was sensitive to pressure, but the overlying skin appeared normal. Intra orally obliteration of left and right buccal vestibule with partially impacted and carious 38 and unerupted 18, 28 and 48. Right retromolar region was associated with draining sinus, which on application of pressure gives off the thick, foul smelling grayish pus. Aspiration from mandibular lesions reveled blood tinged fluid where as aspiration from 28 region reveled clear liquid. Systemic

signs and symptoms, past medical history and hematologic tests were within normal limits and no cutaneous abnormality was revealed.

Orthopantomagraphic examination revels well defined unilocular radiolucency in left body of mandible, large unilocular radiolucency in right posterior mandible involving the complete ramus and irregular radiolucency with respected to impacted 18 and 28. The subsequent axial and coronal computed tomography was done to confirm the extent and site of a lesion. The radiographies from chest and skull were unremarkable.

The patient was admitted to hospital, and complete enucleation of the cystic lesions was performed under general anesthesia all the specimens were sent for histopathologic examination. During the surgery, white cheesy material was found extruding from the mandibular cystic lesion. Curettage was carried out in both the sinus walls. An uneventful healing of the treated area was observed during a close follow up of six weeks which was satisfactorily assisted with surgical stent .

The histopathologic report revealed that the cystic lining of all 4 lesions was parakeratinized stratified squamous epithelium of uniform 5–10 cell thickness. The lining epithelium consisted of well-defined columnar basal cells in a palisade arrangement and with polarized nuclei. The maxillary lesion showed an inflammatory odontogenic cyst appearance with inflammatory cells infiltration in fibro-vascular connective tissue wall.

The patient made a rapid and uncomplicated recovery. To date, more than 1 ½ yr postoperatively, there has been no further pain or swelling and no clinical or radiographic evidence of cyst recurrence.

III. Discussion

OKC has been theme of investigation and study motivated by its tendency of recurrence and potential aggressive nature. Some authors support that it should be considered a benign cystic neoplasm,[8–10] due to its growth capacity and development characteristics related to the mutation of a suppressor tumor gene, PTCH, found in sporadic and in associated to basal cell nevus syndrome keratocysts.[3,8,9,19] The OKC has a predilection for men, occurs significantly more in the posterior region of the mandibule and affect more people from second and third decades of life.[2,5,12-14,19] in our case 20 years male patient had two OKC in posterior mandible and two in posterior maxilla. It seems that less than 1% of all cases of OKC occur in the maxilla with sinus involvement, [19,20] in our presented cases the left maxillary OKC was completely restricted in the maxillary sinus with unerupted teeth association, while the right maxillary OKC developed in the maxillary bone with impacted tooth and expanded to the sinus. In 25 to 40% of cases, an unerupted tooth is seen in association with the lesion[7,21] in our case three out of four OKCs are associated with unerupted teeth.

Multiple OKCs usually occur as a component of NBCCS or Gorlin-Goltz syndrome,[1,11,22] orofacial digital syndrome,[7,23] Noonan syndrome,[7,24]Ehler-Danlos syndrome,[7,25]Simpson-Golabi-Behmel syndrome[7,26] or other syndromes. Our patient was apparently healthy except had history of solitary sebaceous cyst over the scalp which had been operated at the age of 5 years and now no features suggestive of these syndromes, such as basal cell carcinoma, skeletal or orofacial defects, stunted growth, bleeding diathesis, hyperextensible skin and hypermobile hyperextensible skin and hypermobile joints or other congenital anomalies associated with overgrowth.

NBCCS can also include concomitant skeletal features, such as bifid rib, frontal and parietal bossing and mandibular prognathism, and cutaneous abnormalities, such as multiple basal cell carcinomas and palmar and plantar keratosis. Hypertelorism, mental retardation, strabismus, calcification of the falx cerebri and medulloblastoma have also been reported.[11,12,27] In our patient, none of these features indicative of NBCCS was present. Multiple KCOTs might be the first and the only manifestation of NBCCS without any other features associated with syndrome. However, other symptoms can occur in later decades of life.[7]

Radiographic appearances of OKCs are variable from a unilocular or a multilocular radiolucency with scalloped and well-defined margins to soap-bubble or honeycomb radiolucency.[2, 5,15,

The radiographic presentation was divided in four categories[21]:

- Unilocular, i.e. a more or less round configuration, with or without a well defined radiopaque margin.
- Scalloped, i.e. radiolucency with a festooned margin.
- Multilobular, i.e. two or more lobes were seen with no bony septae dividing the lobes.
- Multilocular, i.e. separate locules were seen seemingly divided by bony septae.

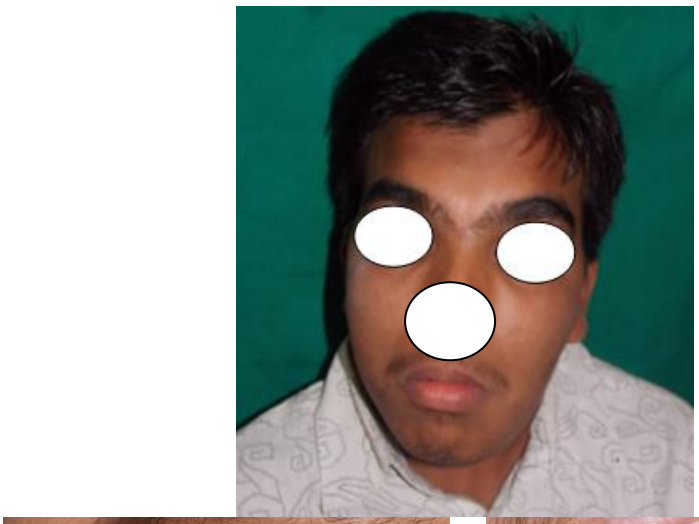
The histological diagnosis was based on the criteria as defined by the WHO ‘Characterised by a thin fibrous capsule and a lining of keratinized stratified squamous epithelium, usually about five to eight cells in thickness and generally without rete pegs.[21] Histologically OKCs have been classified into three categories: parakeratinised, Orthokeratinised, or a combination of the two types. Mostly (86.2%) were parakeratinised, 12.2% were Orthokeratinised, and 1.6% had features of both orthokeratin and parakeratin.[6,15,21,28] Orthokeratinised OKCs have a substantially lower recurrence rate than parakeratinized as parakeratinized variety has more aggressive nature and often associated with NBCCS.[6,21,27] In this case the cystic lining of all 4

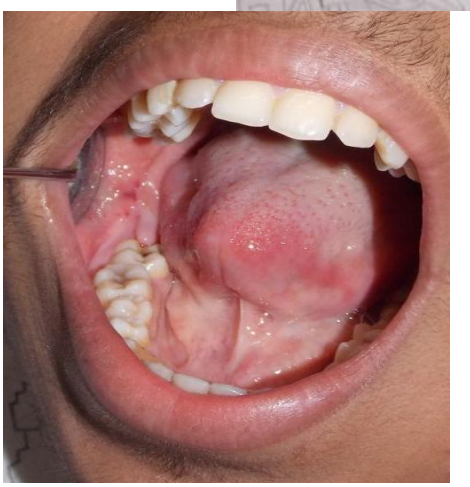
lesions was parakeratinized stratified squamous epithelium of uniform 5–10 cell thickness. The lining epithelium consisted of well-defined columnar basal cells in a palisade arrangement and with polarized nuclei.

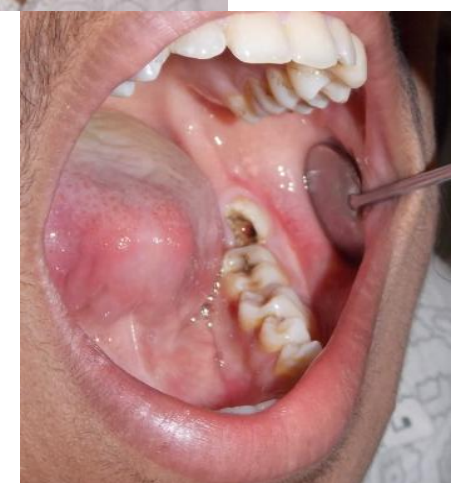
Multiple treatments for the Keratocystic odontogenic tumor have been proposed and debated. There are proponents of "conservative"[16,17, 22,29-31]or "aggressive"[16,18,32] methods of treatment. The challenge lies in minimizing both the risk of recurrence and morbidity of an extensive resection. In this patient complete enucleation was performed for all the four cystic lesions under general anesthesia to reduce the post operative morbidity as patient is of very young age group. Without tearing of cystic lining undue care had been taken for complete elimination. The rates of recurrence vary enormously, from a maximum of 62% to a minimum of 0%.The majority of recurrent cases occur within the first 5 years after treatment. The enucleation alone is associated with the highest recurrence rates (range, 17% to 56%), usually when the cyst is removed in a fragmented fashion. [31] for this reason patient is under the regular follow up for more than 1 ½ year.

IV. Conclusion

The odontogenic keratocyst is a distinctive form of developmental odontogenic cyst that deserves special consideration because of its specific histopathologic features and clinical behavior. Multiple OKCs may or may not be associated with syndrome; it might be the first & only manifestation of NBCCS. In any case irrespective of type of treatment rendered, clinical and radiographic follow-up is mandatory for years after surgery, because recurrence of this lesion may occur even years later.







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