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Case Report

Non-Syndromic Multiple Odontogenic Keratocyst

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ABSTRACT

Odontogenic keratocyst (OKC) is a common developmental odontogenic cyst affecting maxillofacial region. It shows peak incidence in second and third decade . Presence of multiple odontogenic keratocyst without syndromic association is very uncommon. Multiple Odontogenic Keratocysts are principle features of nevoid basal cell carcinoma syndrome (NBCCS; Gorlin-Goltz syndrome). A case of non syndromic multiple odontogenic keratocysts is reported here so as to add to the growing number of such cases in the literature. As odontogenic keratocyst spreads by way of bone marrow, results in more destruction ahead of any clinical manifestation. Therefore, detection in initial stage and intervention are essential in preventing extensive destruction.
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Introduction

Odontogenic keratocysts (OKCs) are the most common form of cystic lesions affecting the maxillofacial region.[1] They are clinically aggressive lesions which are thought to arise from the dental lamina or its remnants.[2] The OKC was first described in 1876and named by Phillipsen in 1956.[3] In the latest WHO classification of odontogenic tumors in 2005, these lesions have been given the name "keratocystic odontogenic tumors" (KCOTs). [4] Multiple KCOTs are usually seen with cutaneous, skeletal, ocular and neurologic abnormalities as a component of nevoid basal cell carcinoma syndrome (NBCCS)which is also recognized as Gorlin- Goltz syndrome. [5] These multiple lesions may be the first manifestation of the NBCCS or otherwise it may be because of the multifocal nature of OKCs.[6] We report a case of multiple OKC without any syndromic manifestations.

Case Report

A 22 year old patient reported to the department with the chief complaint of swelling in mandibular left side of jaw since one month. He also gives history of intermittent fluid discharge from the swelling since 7 days. There is no history of trauma in the past. There is no relevant past dental , medical and family history and all vital signs were in normal limits.

On extraoral examination revealed a solitary diffuse sewelling with overlying normal skin colour present on left side of face extending anterio posteriorly crossing the midline on right side of symphysis region to tragus of ear on left side and superior inferiorly from ala tragus line to inferior border of mandible measuring about 9×3

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cm in size. No visible pulsations and no draining sinus present and on palpation is soft to firm in consistency with overlying skin pinchable , non tender and with slight raised temperature over surface of swelling , [figure 1].

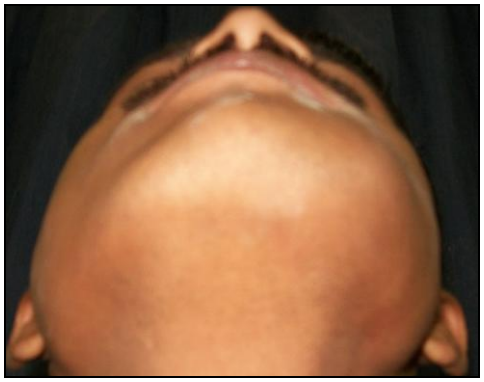


Figure 1: Extraoral extension of the swelling

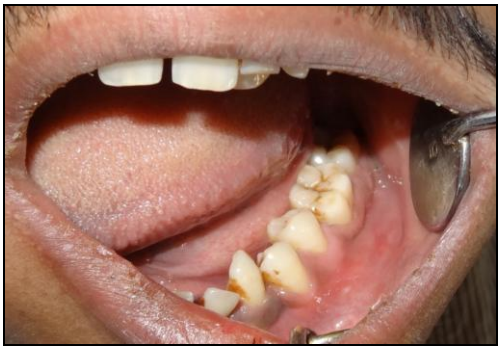
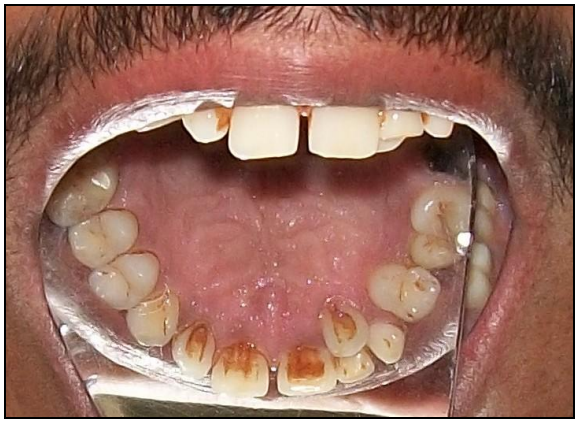


Figure 2: Intraoral examination showing swelling in relation to mandibular left side



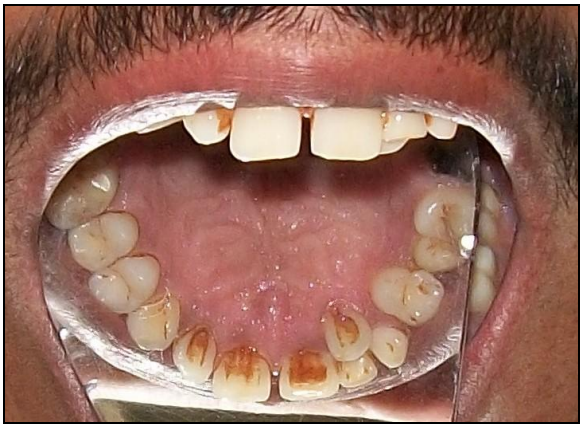


Figure 3(a,b) (intraoral examination showing retained 62 ans 63)

Intraoral examination revealed a solitary diffuse swelling in mandibular left side of jaw in relation to lingually inclined 33 ,34,35 and obliterating the buccal vestibule and non tender , soft to firm in consistency with no draining seen and over retained 62,63 and transposition with 23 and 24 seen,[figure 2 and figure 3].

Radiographic investigations OPG and CT scan were advised.OPG examination revealed multiple cysts in all quadrants. Multiple unilocular radiolucencies seen in relation to 15 ,16 on right side and on left side with 23,24,25. On mandibular left side a single oval multilocular radiolucency seen extending from left coronoid to 43 crossing the midline and another unilocular radiolucency was seen in relation to 47 and 48,[figure 4].

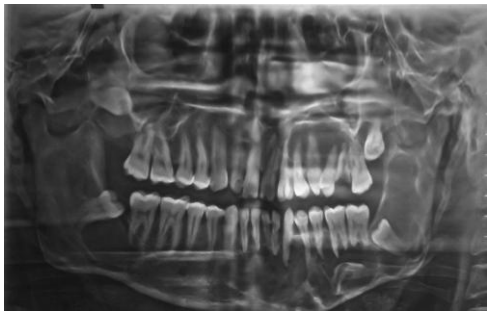


Figure 4: OPG showing multiple radioluciencies

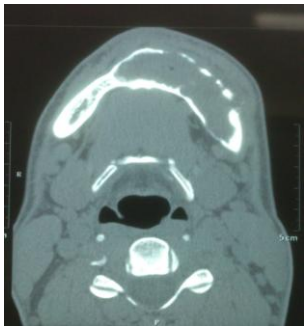


Figure 5: CT scan examination showing hypodense region on andibular left side with expansion and perforation of buccal cortical plate

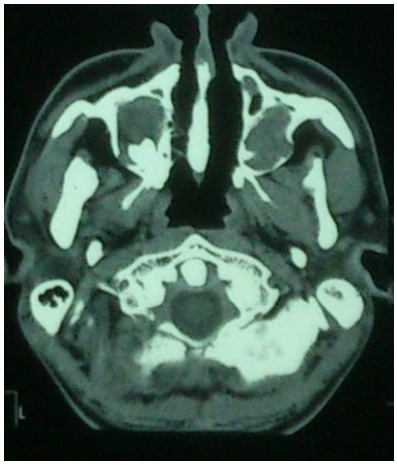


Figure 6: CT scan examination showing haziness in left and right maxillary sinus with tooth in it)

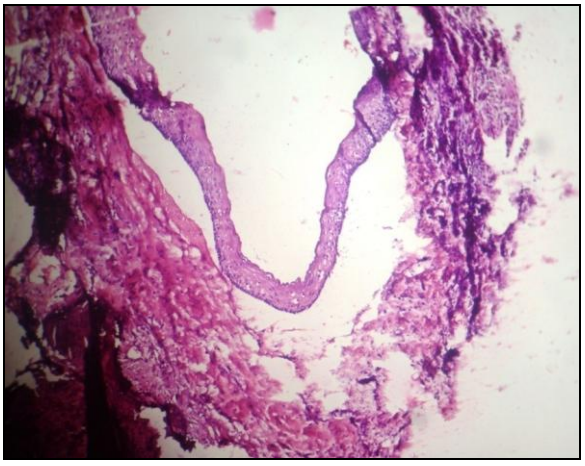


Figure 7: Histopathological report showing stratified epithelium parakeratinized with 6-8cell layered.

CT examination revealed hypodense region on mandibular left side with expansion and perforation of buccal cortical plate and haziness in left and right maxillary sinus with tooth in it. The radiographic examination gave diagnosis of odontogenic keratocyst with mandibular left side and multiple jaw cysts, [figure 5 and figure 6]. Fine needle aspiration cytology of straw coloured fluid aspirate revealed diagnosis of infected dental cyst.

Surgical management was planned under general anesthesia. The cysts in maxillary region were enucleated . The largest cyst extending from 43 to the left coronoid process was marsupialized . A cyst in the Rt mandibular molar ramus region was enucleated.

The histopatholigical report revealed epithelium is stratified parakeratinized with 6-8cell layered. The connective tissue is mostly composed of loosely arranged collagen fibres with inflammatory cells. Thus concluded to diagnosis of infected odontogenic keratocyst,[figure 7]. The patient was on follow up for 6 months and without any recurrences. Later he didn't turned up for follow up.

Discussion

Multiple OKCs usually occur as a component of NBCCS or Gorlin-Goltz syndrome, Noonan syndrome ,Orofacial digital syndrome, , Simpson-Golabi-Behmel syndrome, Ehler–Danlos syndrome, or any other syndromes. Our patient was fit and well with no significant family history and had no features suggestive of these syndromes, such as basal cell carcinoma, defects of skeletal or orofacial region , bleeding diathesis, stunted growth, hyper extensible skin and hyper mobility of joints, or other birth anomalies associated with overall growth.Multiple OKCs may be the first and the only manifestation of NBCCS without any other features associated with syndrome.[1]

NBCCS is associated with mutation in the PTCH gene 9q (22.3-q31). PTCH gene mutation occurs in sporadic OKCs as well as those associated with NBCCS. It is mentioned that a "two –hit" mechanism may underlie the variable expression of NBCCS and sporadic OKCs. In Nevoid Basal Cell Carcinoma Syndrome , the basal cell carcinomas and keratocysts arise as a consequence of a "first–hit" of allelic loss of PTCH within the predecessor cell. The development of basal cell carcinoma and Keratocysts in the absence of NBCCS reflects two somatic hits in which there are mutations of PTCH within locally susceptible cells that ultimately result in allelic loss. The absence of all the features of NBCCS may be due to variability in expression of PTCH gene. [1]

OKC may be found in any age with peak prevalence between 10 to 40 years old and mostly occuring in mandibular posterior body and ascending ramus. Radiologically, KCOTs demonstrate a well defined radiolucent area with smooth and often corticated margins and may be unilocular or multilocular.[4]

Histopathologically, two different variants might be seen even in the similar jaw. Such a case of multiple OKC with diverse histological features showing orthokeratinised and parakeratinised types were reported by Rudagi and Kiyi.[7] However when recurrence rate is concerned, it is said that OKCs without syndrome are less aggressive and have a less recurrence rate as compared to syndrome-associated OKC.[8]

Few identical cases to the current case have been reported in a few published English articles.

Brannon, Auluck , Bartake et al , Guruprasad et al reported cases of OKC , had multiple cysts without any syndromic manifestations.[4,9,10,11]

Other cyst type known to form MCs is the dentigerous variety which generally shows solitary lesion although occasional cases of multiple DCs have also been reported.

Multiple DCs also found associated with few syndromes as Mucopolysacchridiosis , Gardners syndrome and Cleido-cranial dysplasia.[12]

Tournas et al, Freist and Tempest reported a case of multiple Dentigerous cysts in non-syndromic patien.t[13,14]

Multiple radiolucencies in the jaws can also be formed by various other pathologies and should be differentiated from multiple cystic lesions. Some lesions which forms multiple jaw radiolucencies include multiple myeloma, metastatic carcinoma, langerhans disease, etc.[15]

Therapeutic interventions of OKC include marsupialisation and enucleation, combined with adjuvant cryotherapy with Carnoy's solution, and marginal or radical resection.[4]

Conclusion

In any patient with a KCOT, the presence of multiple KCOTs should be considered.

If any patient reporting with the multiple OKCs should be evaluated thoroughly for the possibility of NBCCS as OKCs may be the first and only manifestation of this syndrome. Also for the fact that OKCs associated with this syndrome have higher rate of recurrence than the isolated OKCs, a very strict follow up has to be followed for a long period of time.

The possibility of other features of NBCCS has to be explained to the patient as well as his relatives, so as to allow appropriate genetic counseling and serial screening for the development of malignancies and other complications besides OKCs.

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