Innovations

Multiple odontogenic keratocysts of the jaws – syndromic or non - Syndromic -a case report and review

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Abstract: *Odontogenic keratocyst (OKC) is a benign unicystic or multicystic intraosseous tumor of odontogenic origin. Odontogenic keratocyst (OKC) arises from the cell rests of dental lamina and was primarily termed as "keratocystic odontogenic tumor" in the WHO 2005 conference wherein later during revision in the 2017 WHO meeting, OKC was reinstituted (1). OKCs are mostly solitary lesions commonly seen in the third molar region of the mandible, arising during the second to fourth decade of life that can present radiologically as a single lesion or with septations. OKC's are predominantly seen in males. Multiple OKCs are a rare entity but are usually seen with NBCCS or Gorlin -Goltz syndrome, orofacial digital syndrome, Noonan syndrome, Ehlers-Danlos syndrome, Simpson-Golabi-Behmel syndrome (2). Typically, multiple OKC's have been known to occur in association with NBCCS, but rarely may they be seen without concomitant syndromic manifestations. The Gorlin-Goltz syndrome also called Nevoid Basal Cell Carcinoma (NBCCS) is an autosomal dominant disease presenting with triad features of multiple basal cell carcinoma, jaw cysts and bifid rib (3). Other features include palmar and plantar pits, frontal and parietal bossing, mandibular prognathism, hypertelorism, mental retardation, strabismus, calcification of the falx cerebri and medulloblastoma have also been reported. Studies reveal that 5.8% of non-syndromic multiple OKCs, 8.1% were associated with NBCCS, and 7.6% of them had recurrences (4). We present a case report of a young male with multiple OKC's with absence of any associated syndrome.*

Keywords*: 1.Multiple keratocysts, 2.Gorlin goltz syndrome, 3.maxilla, 4.mandible*

Case report:

A 20-year-old male reported to our Department of Oral medicine and Radiology with chief complaint of persistent swelling on the right side of face for the past four months. Patient's history revealed the swelling was insidious in onset and has gradually progressed to the current state involving the ala of the nose and lower orbital region with no pain associated. Patient also gives a history of continuous pus discharge from his right upper teeth region for the past two weeks. He had already visited a dentist previously regarding the same complaint which was uneventful.

On extra oral examination, a diffuse swelling was noted on the right-side face region with raised right ala of nose and superiorly extending up to the lower orbital region. Patient exhibited increased intercanthal distance.

On intra oral examination there was presence of retained deciduous 53 with intra oral draining sinus seen in relation to 53 and 14. Mild diffuse swelling was seen extending from distal of 37 towards the raphae region, which was soft to firm in consistency, non-tender and was obliterating the buccal vestibule. The overlying mucosa was normal with no secondary changes [Figure 1, 2].

Radiographic examination revealed multiple radiolucent lesions seen in the maxillary and mandibular region [Figure 2]. Well defined unilocular radiolucency with surrounding sclerotic border measuring of about 3x5cm seen extending distal to 12 and mesial to 14 and surrounding impacted 13 superiorly and periapex of retained deciduous 53 inferiorly with root resorption of 53. Another well-defined unilocular radiolucent lesion with surrounding sclerotic border measuring approximately 5x9cm seen in the left lower ramus of the mandible extending from distal root of 37 to posterior border of the mandible antero-posteriorly and from the lower border of the mandible to more than half of the ramus of mandible supero-inferiorly with impacted 38 pushed towards the left coronoid process of mandible. A well-defined unicystic radiolucent lesion with surrounding sclerotic border seen in relation to distoangularly placed 37.

On evaluation of CT of paranasal sinuses impression was given as large soft tissue density lesion with cortical thinning, breach and impacted tooth in maxillary sinuses and rami of mandible [Figure 3].

Blood aspirate was obtained when the patient was subjected to aspiration as chair side procedure from 38 region. Based on these findings, a provisional diagnosis of Odontogenic keratocysts was given. Ameloblastoma and dentigerous cysts were considered in the differentials. The patient was referred to a physician, dermatologist, and neurologist for systemic evaluation of syndromes associated with multiple OKCs. The review of systems did not show any abnormalities and Gorlin Goltz syndrome possibility was ruled out. Hemogram was also within normal limits. Under general anesthesia, surgical enucleation of cyst and chemical cauterization was done. Specimens were subjected to histopathological evaluation and odontogenic keratocyst were confirmed as diagnosis. Patient is under follow-up.

Discussion:

In 1956, Philipsen first described Odontogenic keratocyst (5), are developmental benign intraosseous lesions of odontogenic origin with distinct clinical and histopathological features. They are characterized by its aggressive behavior and very high recurrence rate of 30% (6)for the reason being the interface of the epithelium with the adjacent connective tissue is flat with a high potential for budding of basal layer causing the formation of satellite cysts along with high mitotic activity (7). OKC's constitute about 3-21.5% of odontogenic cysts (8). They are commonly seen in the second to fourth decade of life with male predilection.

The present case was a male patient in his second decade of life. Majority of the OKC lesions occur in mandible twice to that of maxilla and commonly in posterior mandible. In our case, two lesions were seen affecting the posterior mandibular region and one in anterior maxilla. Studies done by Narsapur et al (3) state that 20-40% of OKC's are associated with unerupted or impacted teeth like our case. MacDonald-Jankowski published a systematic review in 2011 which stated that patients presenting with early symptoms characterized by swelling and pain in East Asians in concordance with our lesion.

Radiologically, imaging is more useful in evaluating the extent and the effects on adjacent structures, rather than in characterizing a lesion. Characteristic imaging features in diagnosis of odontogenic keratocyst include unilocular osteolytic lesion around the crown of unerupted/impacted teeth with no septa or loculation within the cyst with more buccolingual expansion in the mandible.

Histopathologically, the present case showed cystic lining in non-keratinized stratified squamous epithelium with basal cell hyperplasia with abundant fibro cellular areas with epithelial rests and few areas of satellite cysts [Figure 4].

Multiple OKC's are considered as one of the main diagnostic criteria for NBCCS and their occurrence may be the first sign of the disease. NBCCS are also known as Gorlin – Goltz syndrome is an multisystemic autosomal dominant disease involving craniofacial, neurological, sexual, ophthalmic, and cardiac anomalies. Literature has reported that it is associated with mutations within the PTCH gene and are also observed in non-syndromic OKC's. Our case presented with no adjoining characteristic features supporting the diagnosis of Gorlin – Goltz syndrome. However, it should be noted that occurrence of multiple OKC's should indicate, until proven syndromic, the early stage of presence of syndrome and be followed regularly to assess the possibilities of other systemic manifestations.

Treatment modalities include conservative and aggressive modalities. Conservative management includes enucleation with or without curettage, cryotherapy or marsupialization thereby preserving the teeth. Aggressive treatment includes peripheral ostectomy, chemical curettage or en bloc resection, commonly recommended for NBCCS cases, large OKC's, and recurrent lesions. In the present case, surgical enucleation of the cyst along with chemical cauterization was done under general anesthesia[Figure 5].

Conclusion:

Multiple OKC's in the jaw as an isolated entity is rare. This case presents OKC in three locations. At the time of clinical presentation, the patient did not have other features of Gorlin Goltz syndrome. An early presentation of exclusive multiple OKC's could be an initial manifestation with other accompanying features possibly developing later. Constant periodic review is required to evaluate possible recurrence of OKC or other systemic accompanying changes.

Declaration of patient consent:

The authors declare that they have obtained consent forms from all the patients which provides their consent in usage of his/her/their images and other clinical information to be reported in a journal. The patients are conveyed that their names and initials will not be published, and efforts will be made in concealing their identities, but anonymity cannot be guaranteed.

Authors contribution:
Abhinaya LM has made substantial contributions towards study design, acquiring case data, drafting the final paper, and revising it critically.
M. Arvind has made substantial contributions towards study design, analysis of case data, drafting the final paper and revising it critically.

Conflict of interest: There are no conflicts of interest.

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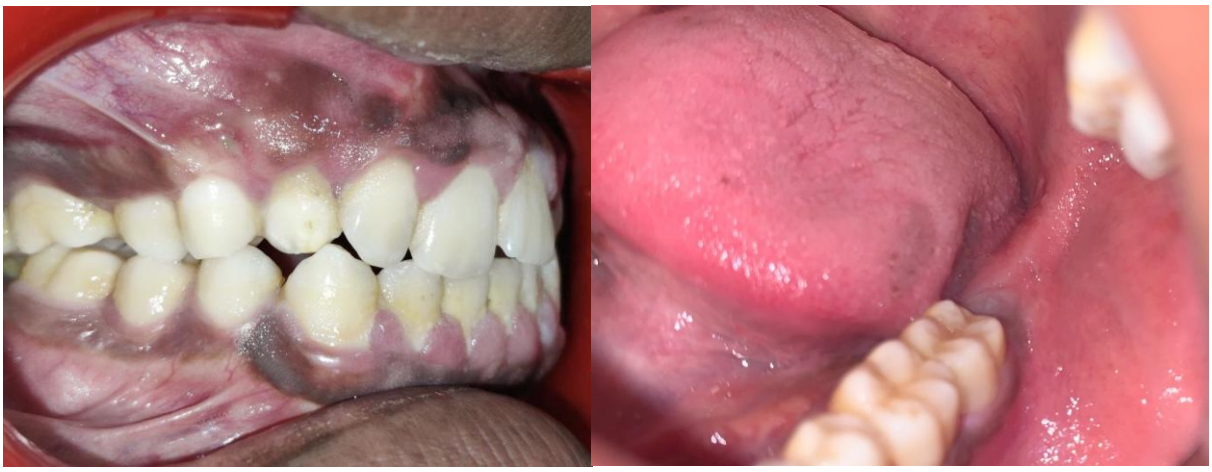

Figure 1: Intra oral images depicting sinus drainage in relation to retained deciduous 53 and diffuse swelling seen in relation to 38

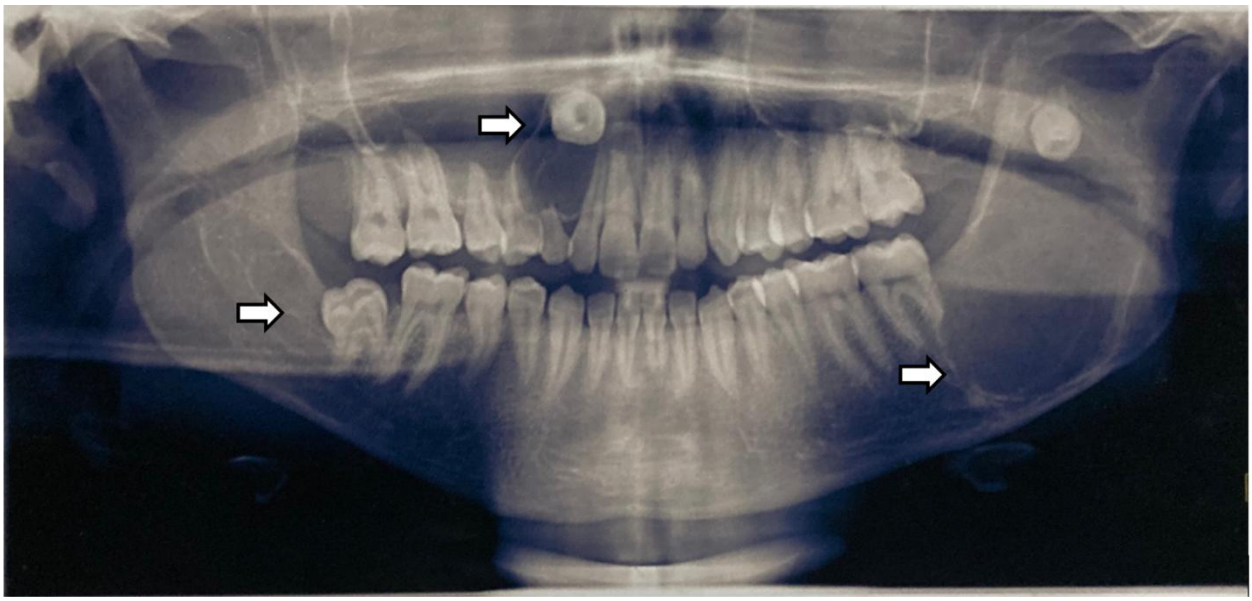

Figure 2: Orthopantomogram depicting multiple OKC's in relation to impacted 13,38 and distal to 37.

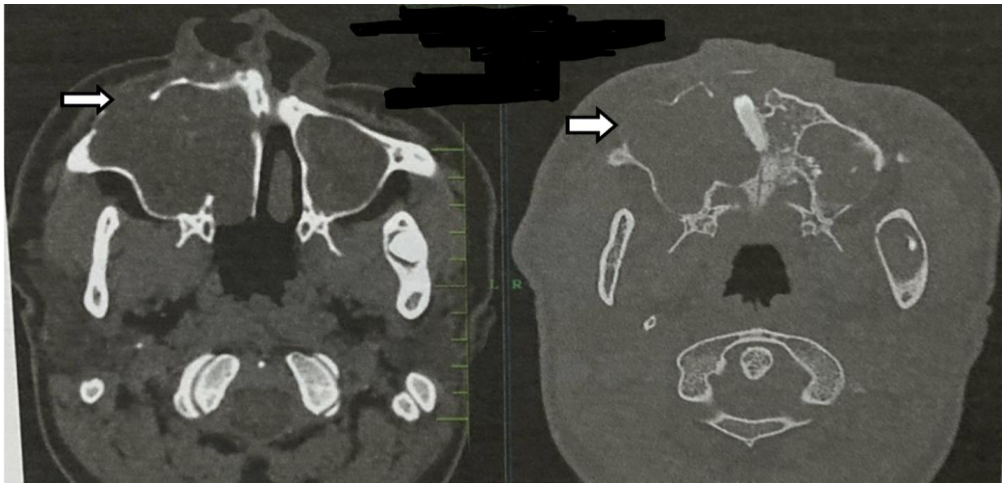

Figure 3: CT image of paranasal sinuses

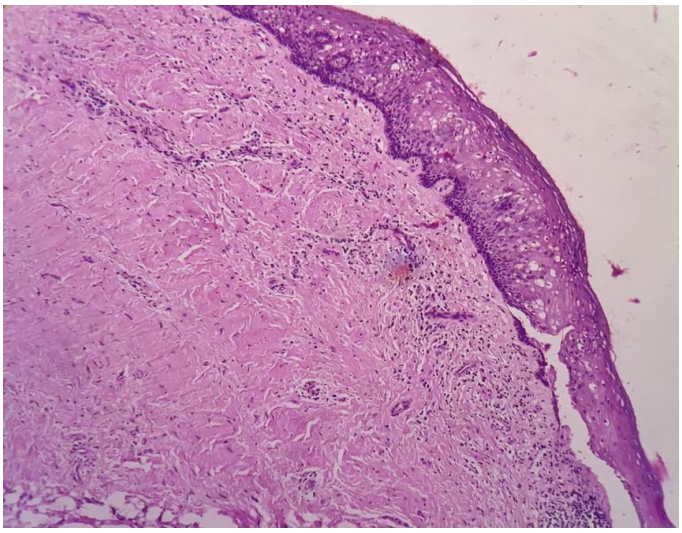

Figure 4: Histopathological image in 10X magnification

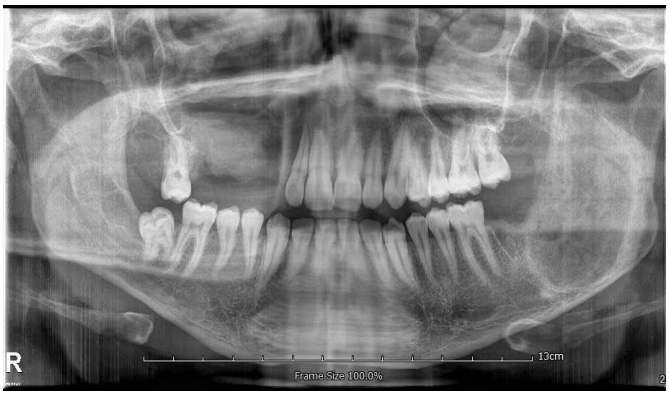

Figure 5: Post-op orthopantomogram