Non-syndromic multiple odontogenic keratocysts - a case report

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Abstract
Odontogenic keratocyst (OKC) is a developmental maxillofacial cyst. Incidence of multiple OKC without any associated syndrome is very exceptional. Multiple OKCs are features of nevoid basal cell carcinoma syndrome (Gorlin-Goltz syndrome). The intent of this paper is to report a non-syndromic multiple OKC, so as to add to the increasing number of such cases to the available literature.

Keywords: Gorlin-Goltz syndrome, multiple odontogenic keratocysts, nevoid basal cell carcinoma, radiographic diagnosis

Introduction
Odontogenic keratocyst (OKC), now reclassified by the World Health Organization as keratocystic odontogenic tumors (KCOT) is one of the most common form of maxillofacial cystic lesion.[1] It is a benign uni- or multi-cystic intraosseous tumor of odontogenic origin. It is locally aggressive and infiltrative and has a distinctive lining of para keratinized stratified squamous epithelium.[2] KCOT constitute about 3-21.5% of odontogenic cysts.[3]

The peak incidence is during the second to fourth decades of life.[4] It is reported that 5% of the patients with KCOTs tend to have multiple KCOTs.[4] Occurrence of multiple KCOTs is characteristically seen with cutaneous, skeletal, ocular, and neurologic abnormalities or syndrome associated. As KCOTs are known for its recurrence, patients with multiple OKCs have significantly high-recurrence rate of 30% compared to solitary keratocyst of 10%.[5]

However, multiple non-syndromic KCOTs are extremely rare entity and have barely been documented in the available literature. This article presents a brief review, and a rare case report of non-syndrome associated multiple KCOTs with a long-term follow-up.

Case Report
An 18-year-old male patient came to our department with the chief complaint of a painless swelling in the upper left front jaw region for 1 month.
Considering an asymptomatic swelling of 1-month duration in a young patient which is firm in consistency a provisional diagnosis of OKC was reached.

The differential diagnosis considered was dentigerous cyst, unicystic ameloblastoma, and adenomatoid odontogenic tumor.

Intraoral periapical radiograph in relation to 22, 23, 24, 25, and 26 shows a radiolucent lesion roughly oval approximately 3×4 cm with distal drifting of 25 and mesial drifting of 26 and no root resorption. OPG reveals multilocular radiolucencies in the left anterior maxilla and left posterior mandible which are approximately 3×2 cm and 2×1 cm, respectively, mesial drifting of 22 and distal drifting of 23 and 26 are seen [Figure 3]. Chest radiograph was advised to rule out the presence of bifid rib. Axial computed tomography images show a loculated expansile lesion on the left side extending into the maxillary antrum with cortical breach. The maxillary antrum is obliterated by the lesion there is an unerupted tooth within the lesion [Figure 4]. There is another expansile lesion of size 25×20×20 mm in the right maxillary region with an impacted tooth. Another loculated lesion of size 45×45×20 mm with floating teeth in the ramus of the mandible.

Incisional biopsy of the lesion under local anesthesia revealed cystic lining made up of para keratinized stratified squamous epithelium of fairly uniform thickness showing corrugation, hyperchromatic basal cells stromal tissue shows dense fibrous connective tissue suggestive of para keratinized OKC [Figure 5].

The lesions were surgically removed into and no recurrence is reported till date.

Discussion

The term OKC was first used by Philipsen in 1956. In 1967, Toller suggested that the OKC may best be regarded as a benign neoplasm rather than a conventional cyst based on its clinical behavior later in 2005 Philipsen in coined the term “KCOT” for OKC.

Multiple OKCs usually occur as a component of nevoid basal cell carcinoma syndrome (NBCCS) or Gorlin-Goltz syndrome, Noonan syndrome, Oro-facial-digital syndrome, Simpson-Golabi-Behmel syndrome, Ehler-Danlos syndrome.

Studies reveal that 5.8% of multiple OKCs presented without any features, Of a syndrome, 8.1% were associated with NBCCS, and 7.6% of them had recurrences.

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Figure 5: Pictomicrograph shows dense fibrous connective tissue suggestive of para keratinized odontogenic keratocyst

The cyst may occur at any age, from the very young to the very elderly. The peak incidence is in the second and third decades of life with slight male predilection (2:1). The mandible is invariably affected more frequently than the maxilla. In the mandible, the majority of the cysts occur in the ramus-third molar area, followed by the first and second molar area and then, the anterior mandible. In the maxilla, the most common site is the third molar area followed by the cuspid region.

The occurrence of multiple OKCs may be the first and only manifestation of NBCCS. Multiple OKCs can occur a decade before other symptoms associated with NBCCS and in patient younger than those with the single OKC. The possibility of our young patient developing other features of NBCCS in the future cannot be excluded.

Radiologically, KCOTs demonstrate a well-defined radiolucent area with smooth and often corticated margins and may be unilocular or multilocular. In 25-40% of cases, an unerupted tooth is seen in association with the lesion.

Auluck et al. discussed a 22-year-old patient with multiple recurrent KCOTs in all four quadrants with a complaint of pus drainage over the previous week without pain or facial swelling. The patient had no other features associated with NBCCS.

Guruprasad and Chauhan discussed a 16-year-old patient with multiple KCOTs and a complaint of slow progressing swelling in both jaws without any other features of syndrome which was similar to our case.

Sholapurkar et al. presented a 24-year-old patient with multiple non-syndromic KCOTs in both jaws with chief complaint of a slow growing swelling for 3 years and drainage for 15 days. The swelling was associated with pain with gradual onset radiating to head on same side. Lesions were cyst-like radiolucencies associated with impacted teeth on panoramic radiograph.

Parikh[11] reported a 19-year-old case with two KCOTs in both jaws without any other concomitant syndromic features. The complaint was swelling for 1 year and pain for 3 months. Panoramic radiograph revealed two radiolucencies with corticated borders associated with impacted teeth.

Bartake et al. reported a 20-year-old case with multiple recurrent KCOTs without any other noticeable features indicative of Gorlin syndrome. No recurrence occurred after 3-year follow-up. When cysts are associated with teeth, several entities might be considered, such as dentigerous cyst, ameloblastoma, odontogenic myxoma, adenomatoid odontogenic tumor, and ameloblastic fibroma. Non-odontogenic tumors such as central giant cell granuloma, traumatic bone cyst, and aneurysmal bone cyst, might be included in a differential diagnosis of this entity in young patients.

Therapeutic interventions of KCOT include marsupialization and enucleation, combined with adjuvant cryotherapy with Carnoy’s solution, and marginal or radical resection. Marsupialization followed by enucleation has the lowest recurrence rate among the conservative treatments.

Conclusion

Multiple KCOTs could occur in patient without any associated syndrome due to its multifocal nature. It is the responsibility of the dentist to evaluate thoroughly and exclude the presence of any associated NBCCS and start the necessary treatment immediately. Owing to the high rate of recurrences related to such cases, a keen follow-up is mandatory. The chance of other characteristics of NBCCS should be explained to the patient and his relatives, so as to allow apt genetic counseling and serial screening for the development of malignancies and other complications besides KCOTs.

References