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CASE REPORT

Keratocystic Odontogenic Tumor: Case Reports and Review of Literature

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ABSTRACT

The lesion traditionally known as odontogenic keratocyst has been renamed by WHO in 2005, as "keratocystic odontogenic tumor" as it is more appropriate and reflects its potential for local, destructive behavior. It is a benign intraosseous neoplasm of jaw, which is unusual due to its characteristic histopathological and clinical features, including potentially aggressive behavior, high recurrence rate and association with the nevoid basal cell carcinoma syndrome. The purpose of this review is to highlight the importance of proper diagnosis of keratocystic odontogenic tumor in order to prevent the recurrence due to improper surgical excision of the lesion.

Keywords: KCOT, Odontogenic keratocyst.

INTRODUCTION

The term 'odontogenic keratocyst' was first used by Philipsen in 1956, while Pindborg and Hansen in 1963, described the essential features of this type of cyst.[1] The diagnostic metamorphosis of odontogenic keratocyst into a recognized cystic neoplasm, keratocystic odontogenic tumor (KCOT), occurred after observation of its biological behavior and the association of chromosomal and genetic abnormalities consistent with neoplastic progression. The KCOT is unique among odontogenic cysts because of its pathognomonic microscopic features, aggressive behavior and high recurrence rate.[1,2] It is estimated that the KCOT makes up 10 to 12% of all developmental odontogenic cysts.[3]

KCOT may occur as solitary lesions, as multiple cysts or as a component of the basal cell nevus syndrome.[4] It is most frequently found in mandible than in the maxilla. In mandible, majority of cases are seen in ramus-third molar area, followed by first and second molar and then anterior region. In maxilla, commonly occurring site is third molar area followed by cuspid area.[1] Although due to its distinct microscopic features, diagnosis depends entirely on its microscopic features and is independent of its location.[2,5]

The most frequently encountered problem with KCOTs is high frequency of recurrence related to residues of cyst epithelium following excision and an intrinsic growth potential causing cortical expansion and erosion.[1,2]

CASE REPORTS

Case 1

A 25-year-old female had visited the Department of Oral Medicine and Radiology, Sharad Pawar Dental College, with a gradually increasing swelling in the palatal region since 10 years and pain with the same since 1 month.

Intraoral examination (Fig. 1) revealed a large smooth surfaced, soft, non-tender swelling of size 3 × 2 cm in the left palatal region. The deciduous maxillary lateral incisors and canines were over retained, and the permanent maxillary laterals and canines were clinically missing. The panoramic radiograph showed missing permanent lateral incisors and impacted canines bilaterally (Fig. 2). There was a large well-defined unilocular radiolucency seen in maxillary anterior region extending from mesial of impacted right canine to the periapical region of left first premolar with scalloped inferior border.

The occlusal radiograph (Fig. 3) showed the multilocular radiolucency extending up to the left maxillary canine. The differential diagnosis of dentigerous cyst, KCOT, unicystic ameloblastoma and AOT was enlisted. Aspiration revealed

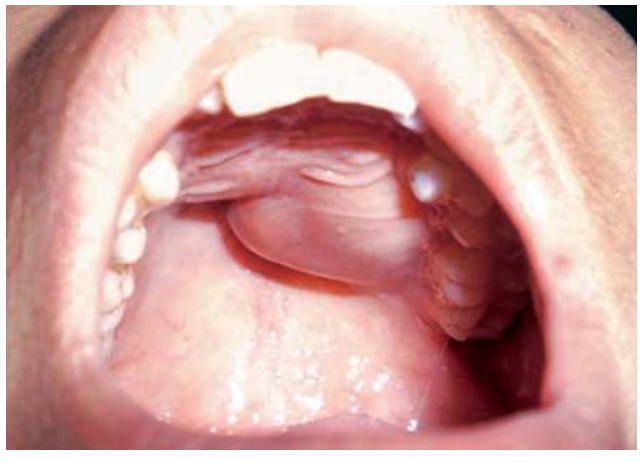

Fig. 1: Intraoral swelling with left maxillary palatal region since 10 years

straw colored fluid. With a provisional diagnosis of dentigerous cyst, surgical enucleation along with extraction of impacted canines was done, but histopathologic examination (Fig. 4) revealed cystic cavity along with epithelium and connective tissue, which was suggestive of infected KCOT.

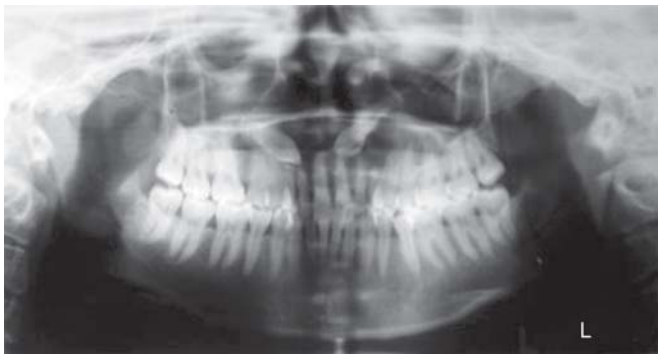

Fig. 2: Panoramic radiograph showing a well-defined radiolucency associated with impacted left maxillary canine

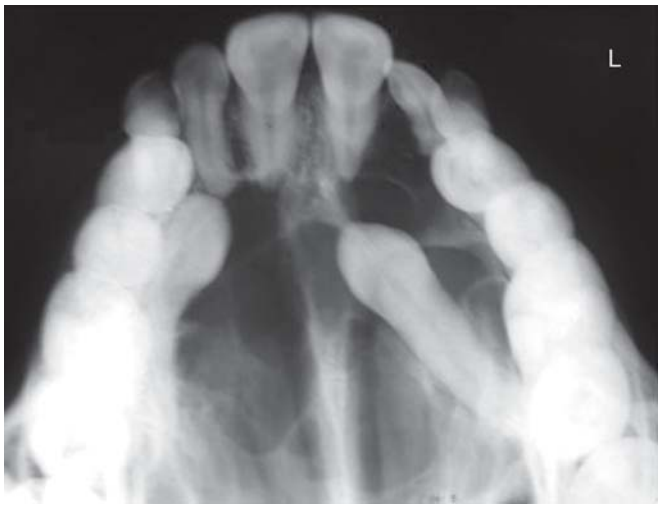

Fig. 3: Occlusal radiograph showing large multilocular radiolucency with scalloped borders associated with both maxillary impacted canines

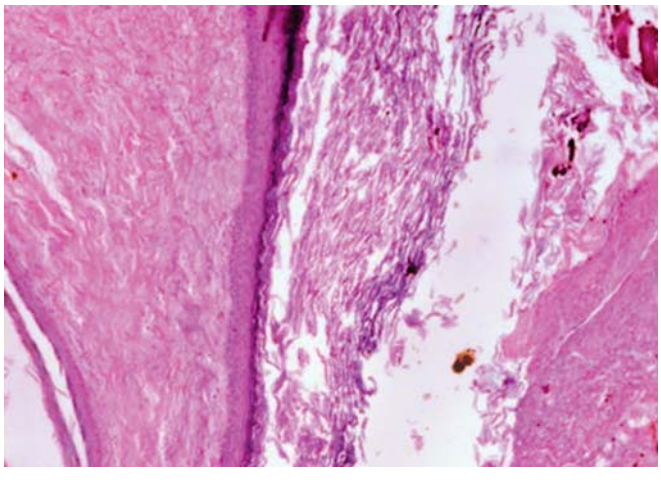

Fig. 4: Histopathologic slide (10 xs) of the lesion diagnosed as infected keratocystic odontogenic tumor

Case 2

A 45-year-old male had visited the Department of Oral Medicine and Radiology, Sharad Pawar Dental College, with the complaint of pain in lower right mandibular region since 10 days and associated swelling since 2 days. An extraoral diffuse facial swelling on right side extending from the corner of mouth to angle of mandible was seen. Intraoral examination revealed unerupted mandibular third molars with partial trismus. Intraoral swelling was seen in the right retromolar region with pus discharge on manipulation distal to the second molar.

Panoramic radiograph (Fig. 5) showed a large well-defined radiolucency with corticated borders associated with right impacted mandibular third molar extending vertically upward into the ramus area. An incidental finding of similar large well-defined radiolucency with corticated borders was seen associated with left impacted mandibular third molar extending to involve the complete ramus area. Thinning of the inferior cortex of mandible was seen on both sides in the third molar region. A provisional diagnosis of bilateral dentigerous cyst was given, and excisional biopsy and extraction of impacted molars was performed. Histological examination (Fig. 6) revealed cystic cavity lined by corrugated parakeratinized stratified squamous epithelium, 8-10 layer thick with fibrous and hypercellular connective tissue stroma which was suggestive of keratocystic odontogenic tumor.

DISCUSSION

Keratocystic odontogenic tumor is believed to be a developmental cyst which originates from the dental lamina or its remnants (glands of Serres).[6]

Ostrofsky found 'epithelial residues' in the retromolar regions and discussed their possible relationship to the formation of KCOTs. The fact that they are consistently found in recurrent keratocyst extending in the ascending ramus and in a considerable number of primary KCOTs in the same region, warrants consideration of an origin different from the dental lamina. The reason why one or two of these microcysts or epithelial islands begin to grow is not clear. In some patients they probably remain dormant for a long period (peak incidence of KCOTs in 5th to 6th decade) or will never produce clinically significant cysts. [7]

Mervyn Shear in 2001[8] observed that KCOT, dentigerous and radicular cyst epithelium reacted positively for epithelial growth factor receptor (EGFr), but with a trend indicating the most intense staining in the KCOTs, followed by the dentigerous and then the radicular cyst linings; these findings led to the conclusion that the KCOTs have an intrinsic growth potential not present in other odontogenic cysts.

Keratocystic odontogenic tumor is more common in males than females and occurs over a wide age range and is typically diagnosed during the second to fourth decade.[1,2,5]

In contrast to this, in the first case reported here, the age of the female patient with the palatal cyst was 25 years.

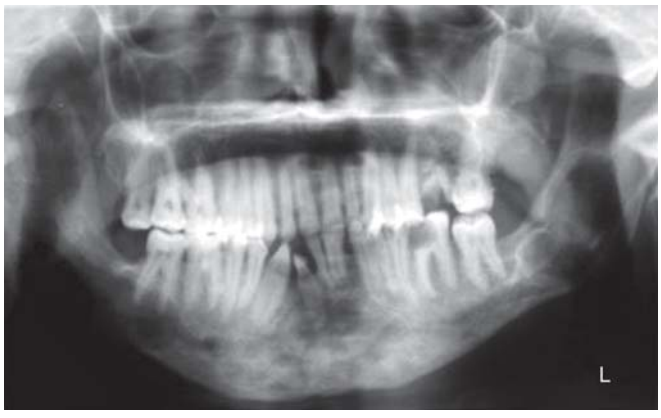

Fig. 5: Panoramic radiograph showing bilateral unilocular radiolucency associated with impacted mandibular third molars. Thinning of the inferior cortex on both sides can be seen

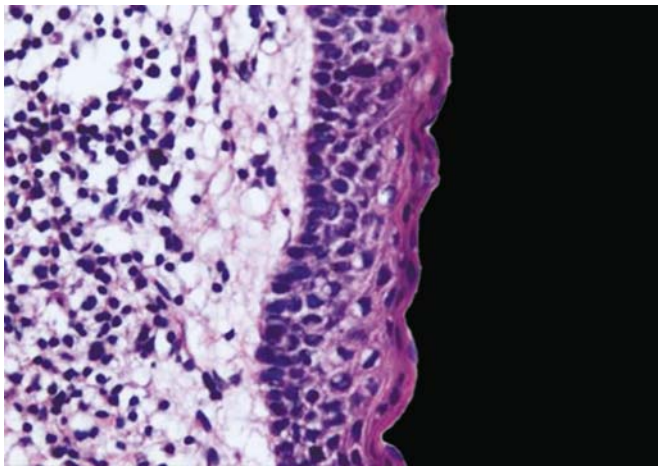

Fig. 6: Histopathologic slides (40 xs) diagnosed as parakeratinized keratocystic odontogenic tumor

KCOT has a predilection for occurring in the mandible (75.58%) as compared to maxilla as reported by many studies carried out in the past.[1,7,9] In mandible, majority of cysts occur in ramus-third molar area, followed by the first and second molar and then the anterior mandible. In maxilla, the most common site is third molar area followed by cuspid region.[1,2,5]

Although keratocystic odontogenic tumor occurs in many jaw locations, it also occurs periradicularly associated with teeth having necrotic pulp and are frequently misdiagnosed as radicular cyst or periapical granulomas. Therefore, it has to be emphasized that the differential diagnosis of periapical radiolucency associated with a tooth and pulp necrosis or an endodontically treated tooth should include KCOT.[11]

In the first case reported here, the lesion was located in the palatal region and in the second case, although located in mandibular third molar region, it was bilateral, both of which are considered to be unusual.

There are no characteristic clinical manifestations of the keratocyst, although about 50% of the patients in Brannon's series were symptomatic prior to seeking treatment. Among the more common features are pain, soft tissue swelling and expansion of bone, drainage and various neurologic manifestations, such as paresthesia of the lip or teeth. The maxillary KCOT tends to be secondarily infected with greater frequency than the mandibular ones, due to its vicinity to the maxillary sinus.[1]

In a review of 256 patients by Myoung et al in 2001, 118 of 256 patients had swelling (46.1%) at first admission, while 51 patients reported with pain (19.9%), and 42 patients (16.4%) had swelling and pain simultaneously. Purulent discharge was evident in 17 patients (6.6 %), while discomfort was evident in 12 patients (4.7%) and paresthesia in two patients (0.8%). About 14 patients had no symptoms (5.5%), i.e. lesions were found accidentally during radiographs.[9]

A similar finding was seen in both the cases reported here. In the first case report, the patient gave a presenting complain of palatal swelling since 10 years and pain since 1 month suggesting the possibility of secondary infection. While in the second case report, patient had a complaint of swelling and pain only on the right side, but the lesion on the left side was asymptomatic and detected only after it was seen on the radiograph.

On radiographic examination, the KCOT cannot be distinguished from other intrabony cysts. In the mandible, the epicenter is commonly located superior to the inferior alveolar nerve canal. It usually shows evidence of a cortical border with a scalloped outline which represents variations in the growth pattern of the cyst.[2] An important characteristic of KCOT is its propensity to grow along the internal aspect of jaws, causing minimal expansion.

Radiographically, KCOTs may present as unilocular or multilocular radiolucencies with a well-defined peripheral rim.[1,2,5] Larger keratocystic odontogenic tumor frequently mimic other pathologic entities, such as the dentigerous cyst, lateral periodontal cyst and the ameloblastoma (multilocular). Smaller KCOTs usually appear as asymptomatic unilocular radiolucencies with corticated borders.[11]

In a prospective study of 82 KCOT cases carried out by Stoelinga et al[7], only 42/82 cases (51.21%) presented radiographically with typical feature of keratocyst. About 40/82 cases (48.78%) showed unilocular appearance, while 17 (20.73%) showed scalloping features and 18 cases (21.95%) showed multilocular presentation out of which only seven cases (38.88%) were seen in the ascending ramus. Studies by Nakamura[12] and Tsukamoto[13] also found a similar distribution.

Tsukamoto et al[14] in 2002 reported differences between KCOTs associated with mandibular third molar and those not associated with teeth. In associated group, 93% had a scalloped periphery while in nonassociated group only 48% gave a scalloping appearance. Mean area of the lesions in associated group was 16 cm^2 and those in nonassociated group was 9.6 cm^2.

The keratocystic odontogenic tumor wall is usually rather thin unless there has been superimposed inflammation.[1,2,5] The lining epithelium is highly characteristic and is composed of:

- A parakeratinized surface which is typically corrugated, rippled or wrinkled

- A remarkable uniformity of thickness of epithelium, usually ranging from 6 to 10 cells thick
- A prominent palisaded, polarized basal cell layer of cells often described as having 'picket fence' or 'tomb-stone' appearance.

Numerous surgical modalities have been suggested for the treatment of KCOTs, including enucleation with primary closure, enucleation with open packing and resection with or without loss of jaw continuity. The treatment depends on several factors, such as age, location and size of lesion, and whether the lesion is primary or recurrent. Total enucleation with or without "peripheral ostectomy" is treatment of choice for most KCOTs unless lesion is recurrent or has significantly invaded soft tissue.[4]

Myoung et al and Brannon, suggested that epithelial remnants or residual tissues are ostensibly prime potentiators of recurrence and for this reason, chemical cautery after enucleation, aggressive curettage of bony walls, cryotherapy modalities, peripheral ostectomy with a bone bur or even radical resection of involved jaw have been advocated as means of treatment for lowering the recurrence by removing the epithelium.[9,15]

Stoelinga et al in 2001 suggested that excision of the overlying mucosa, attached mucosa and treatment of the bony defect with Carnoy's solution aim at the elimination of two possible causes for recurrences: epithelial rests from the cyst wall; secondly, clusters of epithelial islands and microcysts (consistently found in large group of cysts).[7]

In his comprehensive review of 312 KCOTs, Brannon proposed that reasons for recurrence include: technical and/or surgical difficulties resulting in incomplete cyst removal; the thin and friable nature of the capsule; bony perforation; adherence to adjacent soft tissue structures; and remnants of dental lamina epithelium not associated with the original KCOT, which may be activated in susceptible patient to form keratocysts.[15]

Ahlfors et al in 1984, reported the mean time elapsing from treatment of the first keratocyst to the first recurrence for the 69 patients were 5 years, with the mean time for males being 4 years (median 2 years) and for females being 7 years (median 4 years).[16]

Brondum and Jenson in 1991, correlated rate of recurrence with histopathological findings using the Forssell histopathological classification of KCOT as mentioned previously. They found recurrence in 18.0% of KCOTs. Interestingly, all these recurrences were found in cysts with thin, parakeratotic, band-like epithelium with palisade-like basal cells (Forssell group Ia). They also suggested the importance of decompression technique in reducing the tendency for recurrence. In all the 12 large cyst treated by decompression, no recurrence was seen after a period of 7 to 17 years of follow-up.[10]

Nakamura et al in 2002 evaluated the effects of marsupialization on 28 lesions of KCOTs and developed a formula to measure the reduction rate (RR) on the basis of pixel count of the lesions before and after marsupialization {RR(%) = (A [pixel count premarsupilization] – B [pixel count postmarsupialization])/A × 100}. They found that greater the reduction rate, higher the success rate. In some of the lesions, cysts completely disappeared after marsupialization and no further surgery was done. Lesions of the body of mandible showed extremely effective results, whereas half of the lesions in angle and ramus region showed moderately and poorly effective results.[12]

Both our cases were treated by enucleation along with extraction of associated teeth. The patients have been under follow-up since last one year without any complications.

CONCLUSION

Any unilocular or multilocular lesions of either jaw irrespective of the internal structure and type of borders should include a differential diagnosis of KCOT as the typical features of KCOT may not always be present. Postoperative follow-up of minimal 5 years is essential following surgical management, considering the high recurrence rate of these lesions.

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