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Case Report

Keratocystic Odontogenic Tumor: A Case Report

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Abstract: Keratocystic odontogenic tumor (KCOT) comprises a unique pathological entity characterized by aggressive or destructive behavior and propensity to recurrence. Usage of the term odontogenic keratocyst (OKC) had been under a lot of dispute from the time it was introduced in 1956. The World Health Organization (WHO) reclassified this lesion in 2005 as a KCOT and defined it as "a benign uni or multicystic intraosseous tumor of odontogenic origin, with a characteristic lining of parakeratinized stratified squamous epithelium and potentially aggressive, infiltrative behavior." It may be solitary or multiple. Long standing KCOTs have been reported to transform into primary intraosseous carcinoma (PIOSCC) or an ameloblastoma.

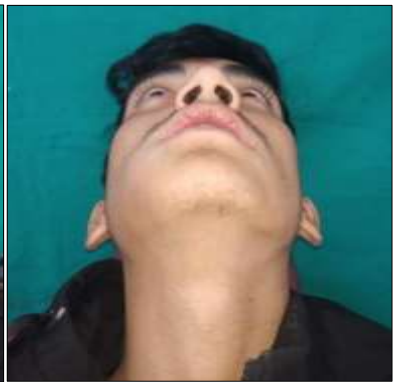
Keywords: Keratocystic odontogenic tumor

Case Report

A 22-year-old male complained of moderate to severe throbbing pain in his right lower back jaw region since 15 days. There was mild associated history of swelling. Patient could not recollect any past history of trauma. His past dental history revealed that he had not undergone extraction of any tooth in past. Patient had swelling around a tooth i.e 46 associated with severe swelling and mild pus discharge Antibiotics and pain killer were taken, following which the swelling subsided to some extent but the pain became continuous. Patient's past medical history was non-contributory. There was no significant personal, dietary and family history.

General Physical Examination was normal and vital signs were in normal limits. On extra oral examination, there was gross facial asymmetry [Figure 1a and b] Diffused swelling over right mid mandibular body region, extending anteriorly to right angle of mouth, posteriorly to 3cm anterior to angle of mandible, superiorly to occlusal plane and inferiorly to the lower border of the mandible. Swelling was hard, tender and with diffused border on palpation. Right and left submandibular lymph nodes were not palpable. Mouth opening was adequate.

(Fig.1a)



(Fig1.b)

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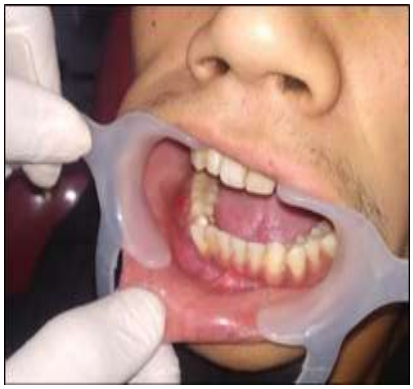
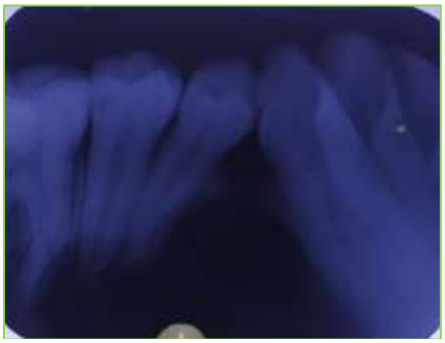
Gingiva was coral pink in colour, contour was scalloped, consistency was firm, stippling was present on surface but there was no bleeding on probing. White lesion was present on bilateral Buccal Mucosa. 26^{th} tooth was missing. Moderate calculus/stain was present on teeth. (Fig 2)

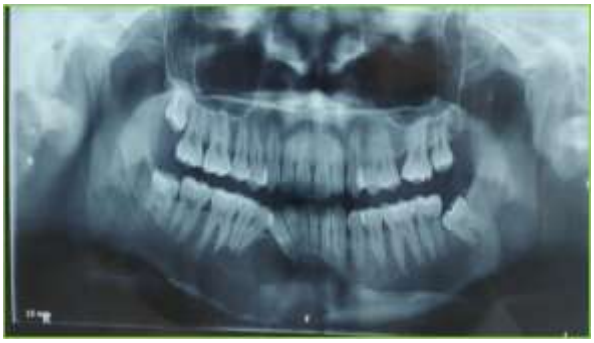


(Fig.2)

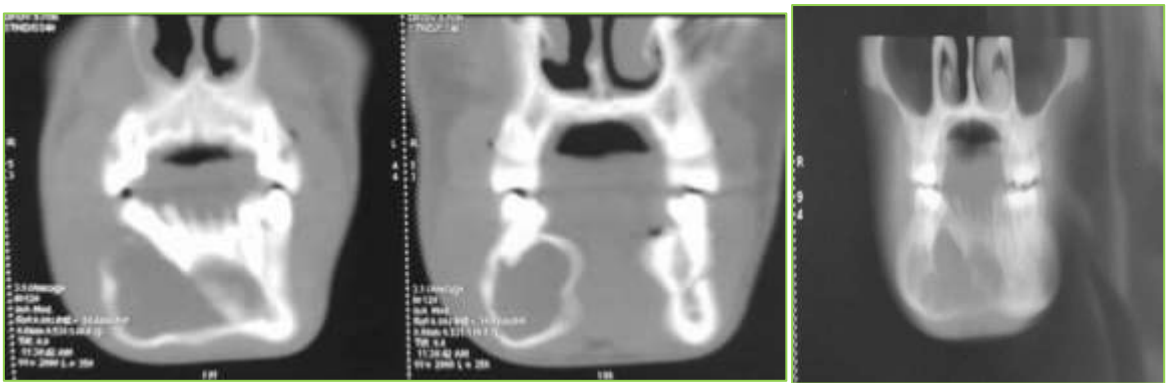
There was Localized erythematous gingival swelling on buccal aspect of tooth 46, Obliteration of buccal vestibule extending from mesial aspect of tooth 31 to distal aspect of 46, Distoocclusal tilting of crown of tooth 43 and mesioocclusal crown tilting of tooth 45. On palpation, mild expansion of the buccal and lingual cortical plates were evident immediately distal to 42, 43, 44 and 45 and the area was tender. There was no mobility of the adjacent teeth.



(Fig.3)

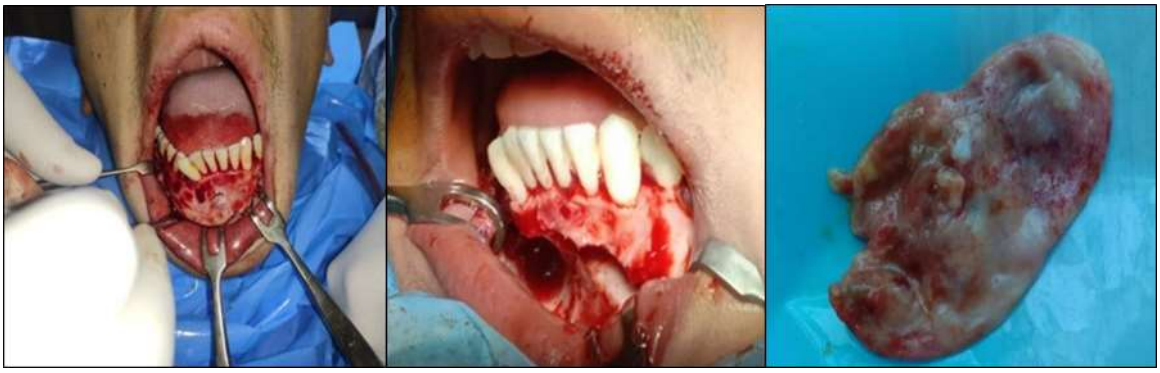


(Fig.4)

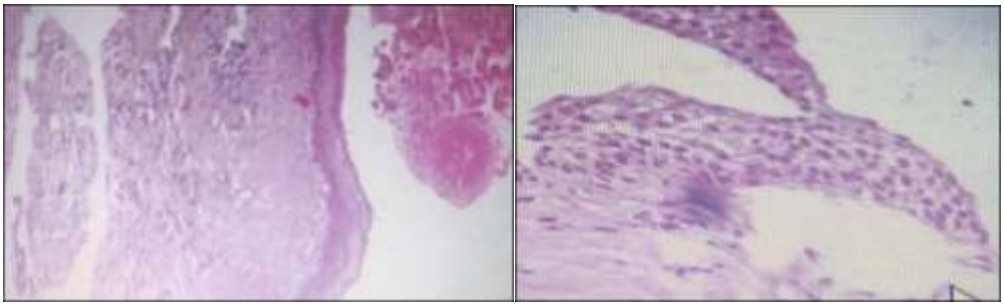


(Fig.5 A, B, C)

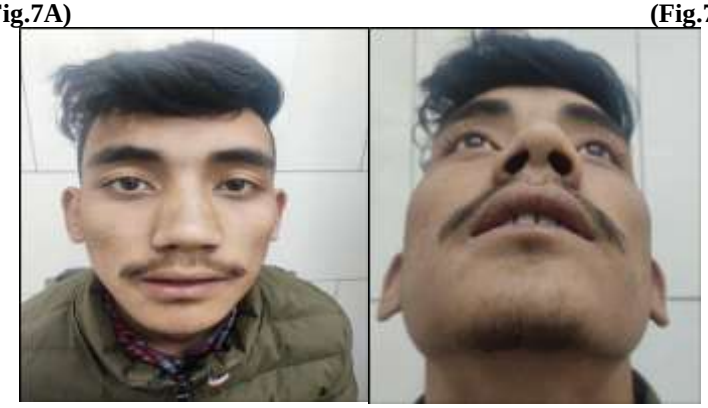
Panoramic radiograph (OPG) revealed a well-defined multilocular radiolucency involving the right body region of the mandible. (Fig 4) The radiolucency measured approximately 6 cm × 3 cm in its largest dimensions and was irregular in shape with well corticated scalloped borders. Antero-posteriorly, it extended from the distal aspect of 34 to distal aspect of 46. External root resorption i.r.t 33,32,31,41,42,43. Within the radiolucency, there was no evidence of internal septa. Inferior alveolar nerve canal was not traceable. Intraoral periapical (IOPA) radiograph revealed evidence of loss of lamina dura in relation to the mesial and distal root of 43,44 and (Figure 3). Computed tomography (CT) demonstrated expansion of the buccal and lingual cortical plates. The inferior alveolar nerve was intact. (Figure 5 A, B, C).



(Fig.6A) **(Fig.6B)** **(Fig.6C)**



(Fig.7A) **(Fig.7B)**



(Fig.8A) **(Fig.8B)**

The lesion was completely excised with marginal resection of the. (Figure 6 A, B, C). The resected specimen histopathologically revealed uniform epithelial lining 6-8 cells thick lacking rete ridges between epithelium and underlying fibroconnective tissue Epithelium characterized by palisaded hyperchromatic basal cell layer comprised of cuboidal to columnar cells with areas of budding growth from the basal cells. Luminal surface has wavy Lumen contain keratinaceous debris orthokeratosis, granular layer and poorly organized basal layer suggestive of features of KCOT (Figure 7 A, B). Currently, he is kept under regular follow-up (Figure 8 A, B).

DISCUSSION

The first description of OKC was published by Philipsen in 1956 for all the cysts that showed keratinization histologically (Philipsen, H. P. 1956). Pindborg and Hansen (1963) suggested the histological criteria, which were confirmed by Browne in 1970 and 1971. In 1992, WHO reported that OKC was the preferred terminology for cysts with keratinized lining. In 2005, WHO reclassified intraosseous parakeratinized variant as a tumor. In clinical practice and require special attention due to their specific histopathologic features, aggressive and infiltrative behavior, and tendency to recur. There are few factors which led to recharacterization of the keratocyst as KCOT (Toller, P. 1967; & Li, X. *et al.*, 2010).

- The KCOT exhibits locally destructive and highly recurrent behavior.
- KCOTs are characterized by parakeratinized epithelium, in contrast to the orthokeratinized variant seen in OKC. KCOT reveals budding of the basal layer into the connective tissue and frequent mitotic figures.
- KCOTs are associated with inactivation of PTCH, the tumor suppressor gene.
- Multiple KCOTs may present as one of the stigmata of the inherited NBCCS. It is also known as Gorlin syndrome.

The KCOT is believed to arise from cell rests of the dental lamina. The human homolog of the Drosophila segment polarity gene PTCH1 has been identified as the gene responsible for NBCCS as well as in sporadic KCOTs. PTCH1 has been mapped to 9q22.3-31. Markers of proliferation found to be associated with KCOT are proliferating cell nuclear antigen (PCNA), Ki67, p53, Bcl2 sequencing of the enzyme dihydrolipoyl acetyl transferase, matrix metalloproteinase 2 and 9. Higher proliferative activity in NBCCS-associated lesions reflects the underlying genetic abnormalities. Human papilloma virus (HPV) has been suggested as a possible etiology due to the presence of koilocytosis in the tumor. A male predominance has been reported in KCOT. On the contrary, few studies have reported a predilection for females. KCOT has been reported in a wide age range, with a peak incidence found in the second and third decades. The mandible is involved more frequently than the maxilla: About 65-83% of KCOTs occur in the mandible. Both in maxilla and mandible, it has a predilection for the posterior part of the jaw. A noticeable number of cases are diagnosed incidentally during routine dental examination. Most frequently occurring signs and symptoms include swelling, pain, and paresthesia. If secondarily infected, discharge, abscess and trismus, and cellulitis can be present. It almost always occurs within bone, although a small number of cases of peripheral KCOT have been reported. FNAC can be non-productive in case of KCOT or may produce a yellowish white keratin material (Philipsen, H. P., & González-Alva, P.,).

Characteristic radiographic features of KCOT are listed below:

- Unilocular or multilocular radiolucency with distinctly corticated, often scalloped, borders.
- Minimal expansion, especially toward the lingual (medial) side and growth along the length of the mandible.
- A radiolucent lumen is seen which can have a hazy appearance in conventional radiography. This hazy appearance or high attenuation is suggestive of a dense proteinaceous material such as keratin.
- Displacement of developing teeth and/or separation or rarely resorption of the roots of erupted teeth and extrusion of erupted teeth.

Morgan and colleagues categorize the surgical treatment methods for KCOT as conservative and aggressive. Conservative treatment includes enucleation, with or without curettage, or marsupialization. Conservative treatments preserve the anatomical structures, but have a risk of higher recurrence rate. Aggressive treatment is usually recommended which includes peripheral ostectomy, chemical curettage with Carnoy's solution, or en bloc resection. Treatment of cysts with Carnoy's solution before enucleation tends to damage the epithelial lining to such an extent that proper histological diagnosis may be difficult. Recent studies have shown possible treatment modalities for KCOT, such as cyclopamine, a plant-based steroidal alkaloid, which inhibits the cellular response to the sonic hedgehog (SHH) signal activity. Zhang *et al.*, postulate that antagonists of SHH signaling factors could effectively treat KCOTs. They suggest that intracystic injection of a smoothened (SMO) protein antagonist has the greatest potential as a future treatment option. The average reported recurrence rate ranges from 30 to 62%. The presence of residual epithelium or an epithelial remnant after the treatment is one of the suspected contributing factors for the high recurrence rate or presence of satellite cysts in the cyst wall. A higher recurrence rate in young patients and maxillary lesions (Bhargava, D. *et al.*, 2012; &Kramer, I. R. *et al.*, 1992).

CONCLUSION

Definite diagnosis of KCOT on a clinical and radiographic basis is not possible. But with appropriate and advanced imaging modalities, we can strongly suspect this entity, and they help us in selecting the necessary treatment protocol as it is an aggressive tumor. Very rarely, KCOT may show features which could possess the potential to evolve into PIOSCC or ameloblastoma, similar to the present case.

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