Case Report

Non-Syndromic Keratocystic Odontogenic Tumor: A Rare Case Report

Sarfaraz Padda, Bhawandeep Kaur, Preeti Dhir and Gaurav Goyal

ABSTRACT

The keratocystic odontogenic tumor is relatively a rare benign neoplasm arising from dental lamina, or basal cells of oral epithelium can manifest in either of the jaw i.e. maxilla or mandible. Most commonly, it is asymptomatic and discovered as an incidental finding on radiographs. Because of the high rate of recurrence, patients should be kept under long-term follow-ups. This paper illustrates follow up the case of a nonsyndromic familial KCOT in a 30-year-old female patient.

Keywords: Keratocystic odontogenic tumor, Non-Syndromic, Enucleation, Recurrence

INTRODUCTION

Keratocystic odontogenic tumor (KCOT) is a developmental odontogenic cyst of epithelial origin arises from cell rests of dental lamina and is well known for its potentially aggressive behavior and significant high rates of recurrences.

The term odontogenic keratocyst was first used by a European Philipsen in 1956.[1] In 2005, World health organization renamed odontogenic keratocyst as Keratocystic odontogenic tumor as this entity exhibits the features of both cyst as well as the neoplasm. According to WHO, KCOT has been defined as "A benign uni- or multicystic intraosseous tumor of odontogenic origin, with a characteristic lining of para-keratinized stratified squamous epithelium and potentially aggressive, infiltrative behavior.[2] It may be solitary or multiple. The latter is usually one of the stigmata of the inherited naevoid basal cell carcinoma syndrome (NBCCS)."[3]

In mandible, keratocystic odontogenic tumors constitute between 2% and 11% of all mandibular neoplasms. They are more frequent in males than females. Many cases are asymptomatic and are discovered accidentally on radiographs.[4] The most important radiological finding of KCOT is its anteroposterior expansion as compared to buccolingual expansion having multilocular appearance, scalloped margins and can cause resorption of the teeth.[5]

It has been suggested that high rate of recurrence in KCOT might be because of incomplete removal of the original cyst lining; retention of micro cysts or epithelial islands in the wall of the original cyst; and development of new keratocysts from epithelial offshoots of the basal layer of the oral epithelium.[6,7]

The keratocystic odontogenic tumor may be solitary or multiple in occurrence. Multiple KCOTS are associated with some syndromes like nevoid basal cell, or Gorlin-Goltz syndrome,[8] EhlerDanlos Syndrome,[9] Noonan Syndrome,[10] Orofacial digital syndrome,[11] Simpson- Golabi- Behmel syndrome[12] or can be nonsyndromic.[13]

CASE REPORT

A 30 years old female reported to the Department of Oral Medicine & Radiology, Genesis Institute of Dental Sciences & Research with a chief complaint of painless swelling in his left lower third of the face since 15 days. On intra oral examination, obliteration of the left buccal vestibule was noticed with sinus present irt attached gingiva of 36 regions and 35 was vital on vitality test (Fig. 1).

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Accepted for publication on : July 25, 2015

To cite: Padda S, Kaur B, Dhir P, Goyal G. Non-syndromic keratocystic odontogenic tumor: A rare case report Asian J Oral Health Allied Sci 2015; 5:18-22.

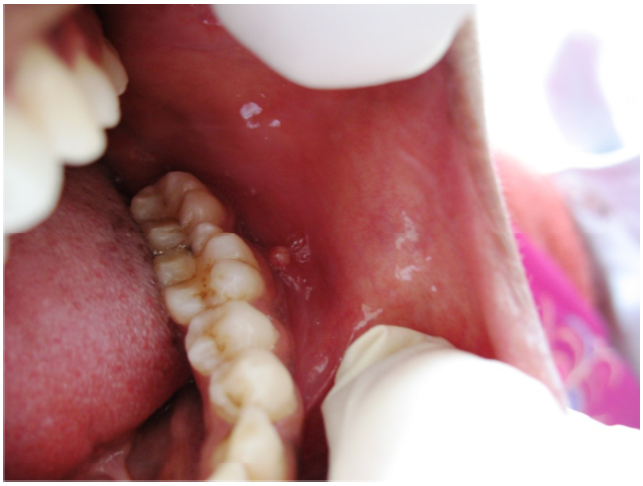

Figure 1: Intraoral picture showing obliteration of vestibule and sinus with respect to 36

From history and clinical examination, the provisional diagnosis of the chronic periapical abscess was given, keeping in mind the radicular cyst, dentigerous cyst, odontogenic keratocyst and ameloblastoma as the differential diagnosis.

Then the patient was subjected to the routine radiographic examinations including intraoral periapical (IOPA), occlusal and panoramic radiography. The IOPA radiograph reveals radiolucency in the periapical region and occlusal radiograph reveals the expansion of the buccal cortical plate. The panoramic radiograph revealed well corticated unilocular radiolucency in the left mandibular body and ramus area extending from 36 region till the 38 region (Fig. 2). The radiographic diagnosis of the dentigerous cyst was postulated.

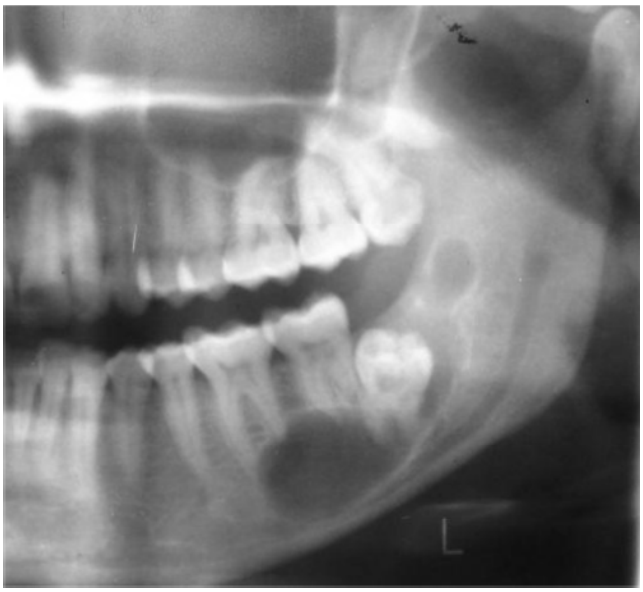

Figure 2: Panoramic radiograph showing well defined unilocular corticated radiolucency with respect to 36 to 38 areas.

Aspiration was attempted with a wide bore needle, under aseptic conditions from the anterior part of the lesion. Blood tinged fluid was aspirated which was subjected to protein estimation and protein content found to be 2.3 gm / 100 ml (Fig. 3). Peripheral smear of the aspirate also showed keratinized squamous cells on stained film. These features are highly suggestive of odontogenic keratocyst.

Figure 3: Blood tinged aspirate fluid

The swelling was treated by enucleation under local anesthesia along with the removal of 38 and tissue was sent for histopathological examination which confirmed the diagnosis of keratocystic odontogenic tumor (Fig. 4) depicting a uniform 6-8 cell thick para-keratinized stratified squamous epithelium lining a thin fibrous connective tissue wall (Fig. 5). The patient is under regular follow ups and healing is satisfactorily going on and even after 5 years no recurrence has been reported (Fig. 6).

DISCUSSION

It is a benign developmental odontogenic tumor with many distinguishing clinical and histologic features and having significant potential for local destructive behavior high recurrence rate. Brannon[14] in 1976 reported a wide age range distribution (7 to 93 years) with the peak frequency

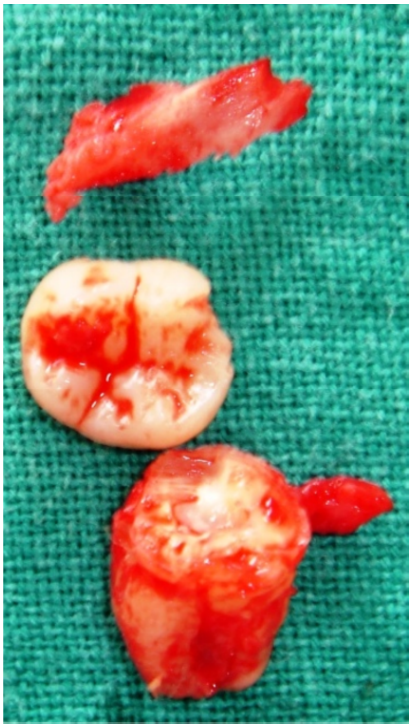

Figure 4: Figure showing surgically removed tissue.

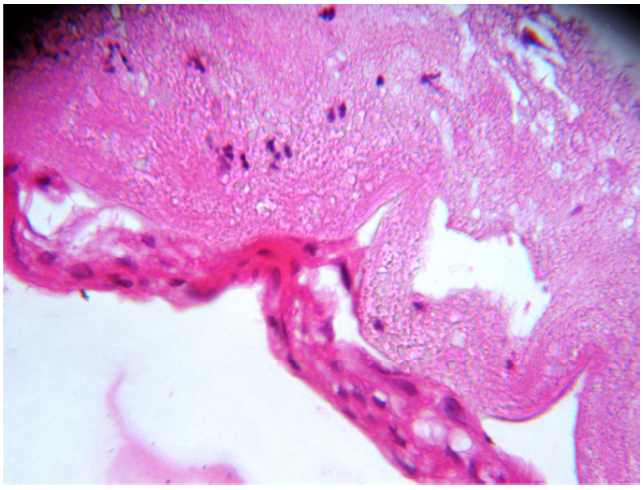

Figure 5: Haemtoxylin and eosin stained section showing 6- 8 cell layers thickness along with fibrous connective tissue wall.

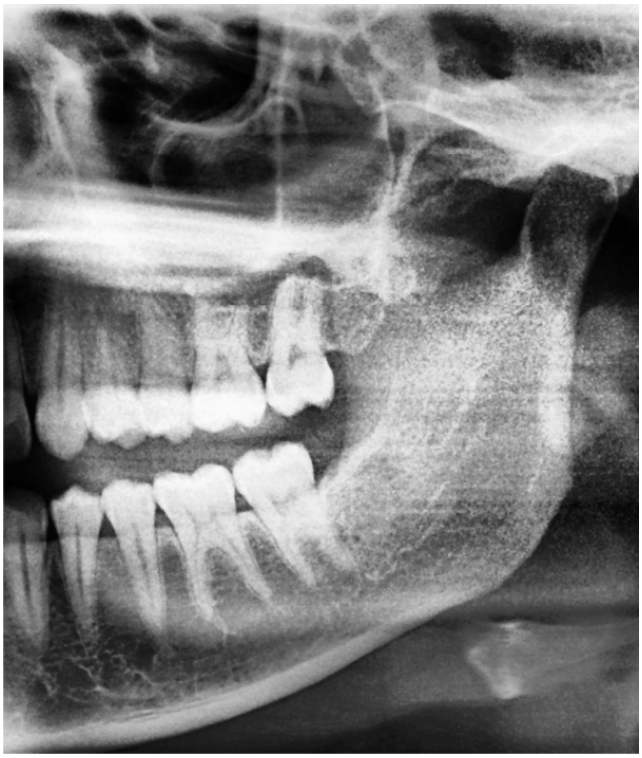

Figure 6: Panoramic radiograph of the patient after five years follow up.

in the second and third decade. Shear[4] in 2003 has reported a bimodal age distribution with a second peak in the fifth decade or later. Keratocyst are found more frequently in men than women, and this gender predilection is more pronounced in black than in white. Higher preponderance in females (17:1) has been observed when associated with Gorlin-Goltz Syndrome.[4,8]

OKC arises from the remnants of dental lamina or proliferation of basal cells of oral mucosa and tends to spread along the cancellous component of bone without producing much expansion of cortical plates. Various reasons have been postulated for enlargement of OKC including interluminal hypo-osmolality and epithelial proliferation. The epithelial expansion manifests itself by an eccentric and multilocular growth pattern possibly explained by an increase in mitotic activity within the cyst lining. Donoff *et al.*[15] in 1972 demonstrated the presence of collagenase within the cyst wall and believed that the enzymatic degradation may contribute to expansion. Kubota *et al.*[16] in 2005 have demonstrated an increase in levels of IL-1α which in turn enhances type 1 collagen- induced activation of matrix metalloproteinase-2, which stimulates enzymatic degradation of extracellular matrices of bone around OKCs, causing expansion. Li *et al.*[17] found levels of parathyroid hormone related proteins within the para-keratinized lining of OKC significantly high and thus speculated that these might modulate growth and bone resorption in OKC through its effect on osteoclasts and osteoblast activity. Toller[6] in 1976 showed that osmolality of the cyst was higher than that of serum which results in the growth of keratocysts. Mandible is more frequently involved than maxilla with figures ranging from 69% to 83%, with the mandibular molar-ramus area being the common affected site.[4,14] Few reports suggested maxillary canine being the favored site,[18] while Myoung *et al.*[19] reported posterior maxilla being the predominant site.

Radiographically, OKC is commonly seen as an unilocular radiolucency having well-corticated margins. Enlarging cyst tends to displace teeth. Displacement and destruction of the floor of orbit and proptosis of eyeballs have been reported in keratocyst involving maxillary sinus. Bony expansion has been seen in approximately 60% of cases.[20]

Sholapurkur *et al.*[21] in 2008 presented a 24 year-old case with multiple non-syndromic KCOTs in both jaws with the chief complaint of a slow growing swelling since 3 years and drainage since 15 days. The swelling was associated with pain with gradual onset radiating to head on the same side. Lesions were cyst-like radiolucencies associated with impacted teeth on the panoramic radiograph. Parikh[22] reported a 19- year-old case with two KCOTs in both jaws without any other concomitant syndromic features. The complaint was swelling for one year and pain for three months. A panoramic radiograph revealed two radiolucencies with corticated borders associated with impacted teeth. Kargahi[23] reported the 11-year-old case with two KCOTs in both jaws with a complaint of swelling in the left side of the upper jaw, and panoramic radiograph reveals two radiolucencies with the corticated border around the unerupted mandibular left canine and the unerupted maxillary left the second molar area. Maxillary second molar was displaced.[23]

Therapeutic interventions of KCOT include marsupialization followed by enucleation also combined with adjuvant cryotherapy with Carnoy's solution, marginal or radical resection in the adults. In our case enucleation was done along with the removal of the third molar to prevent reocurrence.

CONCLUSION

The keratocystic odontogenic tumor is a locally aggressive, cystic jaw lesion with a high growth potential and a propensity for recurrence. OKCs should be one of the differential diagnosis for the radiolucencies of the jaw. Hence careful clinical and radiographic examination has to be done to rule out any other existing lesion, followed by histopathologic correlation for proper treatment and follow up. The recommended follow-up for OKCs should be once in a year for at least five years as done in reported case.

Conflict of interest: No conflict of interest declared by authors.

Source of funding: Nil

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