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Original Research Paper

Atypical Presentation of A Keratocystic Odontogenic Tumor

Medical Science

KEYWORDS : Ameloblastoma, Odontogenic cyst, Odontogenic tumor

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ABSTRACT

Keratocystic odontogenic tumors[KCOT] are developmental in origin and comprise approximately 11% of all cyst form lesions of the jaw. KCOT are unique odontogenic lesions that have the potential to progress aggressively and have a higher recurrence rate. It usually occurs in the mandibular ramus region. Normally the lesion progresses through the medullary spaces with minimal bone expansion, which makes the lesion asymptomatic and often, an incidental finding. This paper presents an atypical case of KCOT with variant radiographic and clinical findings confirmed with histopathology.

INTRODUCTION

Atypical clinical presentations of odontogenic lesions often pose a diagnostic challenge with appropriate management protocols left in the lurch. In these instances, a multi-centric approach with an integration of astute clinical examination, imaging and histopathology will be the only recourse to manage the lesion and determine a prognosis. Odontogenic Keratocyst (OKC) was first described by Philipsen in 1956. In 1963, Pindborg and Hansen described the essential featuresof Odontogenic Keratocyst. Because of the unusual growth pattern and tendency to recur even after surgical removal, the lesion has raised considerable clinical interest.[1]

OKC is now designated by The World Health Organization (WHO) as Keratocystic Odontogenic Tumor (KCOT) and is defined as a" benign uni- or multicystic, intraosseous tumor of odontogenic origin, with a characteristic lining of parakeratinised stratified squamous epithelium and potential for aggressive infiltrative behavior". There by WHO recommends the term KCOT as it better reflects its neoplastic nature.[2,3]

OKC/KCOTs is the commonest developmental cyst which grows rapidly and has a tendency to invade the adjacent tissue including bone.[2,4] KCOTs are derived either from the epithelial remnants of the tooth or basal cell layer of the surface epithelium. Some authors suggest that as many as half of KCOTs are related to Nevoid Basal Cell Carcinoma Syndrome(NBCCS) or Gorlin syndrome. The most common site is mandibular posterior region and in maxilla is the canine region followed by the anterior maxilla.[5]The lesion generally progress in anterior to posterior direction rather than expanding and growing. Despite this aggressive growth, they often remain asymptomatic.[6] The recommended surgical management varies from marsupialization and enucleation to enbloc resection.[2] We hereby report a case of KCOT with atypical clinical features and there by elaborating on the myriad clinical presentation of this dichotomous entity.

Case Report

A 28 years old male reported to the Department of Oral Medicine and Radiology with a complaint of swelling in the lower left side of the face since four months[Fig 1]. The swelling was initially small and rapidly increased to its present size. The swelling was otherwise asymptomatic. On extra oral examination, a solitary well defined swelling measuring around 2×2 cm was noted over the left angle of the mandible. The skin over the swelling was smooth and appeared normal. On palpation a bony hard swelling with well-defined borders was noted with no tenderness. Intra oral examination revealed a well-defined bony hard swelling on the buccal vestibule in the region of 37, the swelling was smooth, mesio-distally the lesion extending from mesial aspect of 36 to the distal side of 38 and superior-inferiorly extend from the external oblique ridge to the lower border of mandible. No lingual cortical expansion was noted. Clinicopathologic correlation lead to a provisional diagnosis of Ameloblastoma.

Orthopantamogram revealed a unilocular well defined radiolucency with corticated borders of size about 5× 5 cm in the left angle of the mandible, displacing the roots of 38 and the mandibular canal inferiorly[Fig 2]. Mandibular occlusal radiograph of third quadrant reveals a well-defined radiolucent lesion with sclerotic borders noted in relation to 36,37,38 region with buccal cortical plate expansion with no lingual cortical plate expansion[Fig 3]. Pulp vitality test of the third quadrant revealed all the teeth to be vital. Aspiration of the swelling was carried out which resulted in a milky fluid aspirate[Fig 4]. Protein estimation of the fluid reported a value of 4.2 gm/dl.

An incisional biopsy was performed under local anesthesia from which soft and hard tissue samples were collected. The histopathology of the sample revealed parakeratinized corrugated stratified squamous epithelium which is 6 to 8 cell layers thick with hyperchromatic palisaded basal cell layer. The underlying connective tissue shows delicate fibrillar collagen with plump fibroblasts. The epithelium and connective tissue interface shows split. The section also shows a hyperplastic stratified squamous epithelium with dense collagenous connective tissue stroma with dense chronic inflammatory cell infiltrate and marked vascularity is seen. Section also shows deep haematoxyphilic structures resembling bone[Fig 5]. The histopathological finding's lead to an impression of keratocytic odontogenic tumor.

A final diagnosis of KCOT was arrived from the clinical and histopathological features. A treatment of enucleation was planned and performed under local anesthesia. The histopathology of the excised tissue also confirmed the diagnosis of KCOT.

Discussion

Early diagnosis of KCOT is difficult because the lesion normally involve the cancellous bone and tends to enlarge it

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to considerable extent before any significant buccal or lingual expansion appears. KCOT are locally destructive jaw lesions and must be distinguished from the other jaw cysts. The diagnosis of the lesion can be confirmed only by histologic examination. This is the reason why appearances of the lesion on roentgenograms and clinical presentation usually do not reveal the true nature of the lesion.[7]

One of the characteristic features of KCOT is recurrence. Donoff *et al.* associated recurrence of OKC with increased activity of epithelial lining and observed separation of the epithelium from the underlying connective tissue. They hypothesized that the enzymatic activity within the cyst might be responsible for the observed epithelial separation.[4] Brannon, , in his review of 312 OKCs, suggested three mechanisms responsible for recurrence: remnants of dental lamina within the jaws not associated with the original OKC, incomplete removal of the original cystic lining and cortical perforation with adherence to adjacent soft tissue, and cell rests of dental lamina and satellite cysts that remain behind after enucleation[8,9].

Radiographically the lesion presents as a small ovoid or round radiolucent areas on conventional plain radiographs. It may present as a unilocular radiolucency with smooth well defined margins, bilocular or multilocular with scalloped margins, sometimes with ill-defined periphery surrounded by thick sclerotic margin. Minimal medullary expansion in the mesio-distal aspect cannot be seen in normal radiographs but perforation can be seen as focal discontinuity of the cortical border with the absence of sclerosis at the peripheral margin. The desquamated keratin contents may appear cloudy or milky-way.[10] The KCOT seen bilaterally or in both the jaws in patients younger than 10 years of age should raise suspicion of Gorlin-Goltz syndrome.

The treatment strategies vary with the clinical presentation of the lesion, biologic nature and recurrence potential. The conventional options include enucleation and curettage, enucleation and peripheral ostectomy, osseous resection without (rim ostectomy/ marginal resection) or with (segmental resection) continuity defect. Aggressive treatment modality like enucleation with application of carnoy's solution can be done.[11]

CONCLUSION

Much controversy exists regarding the origin, growth and treatment of KCOT. Due its enigmatic clinical presentation and propensity for recurrence, rigid clinical follow up is mandatory and the minimum recommended is annual for atleast five years. Clinical, radiological, histopathological diagnosis and treatment plan plays a pivotal role in the management of KCOT and a multidisciplinary approach will ensure enhanced prognosis.

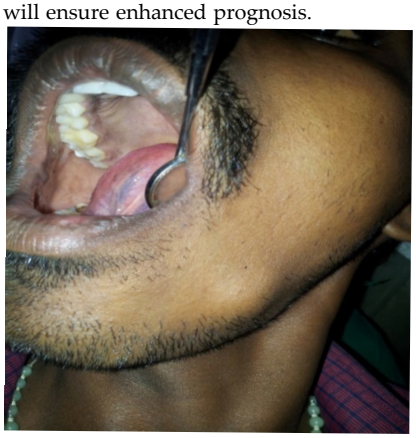

Figure 1- Swelling in the lower left side of the face

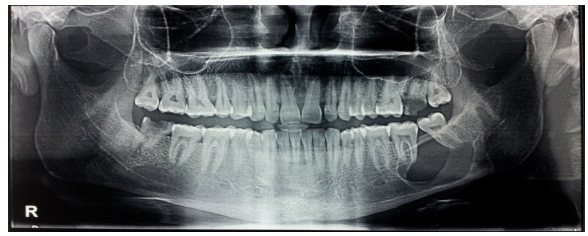

Figure 2- Orthopantamogram showing a unilocular well defined radiolucency with corticated borders of size about 5× 5 cm in the left angle of the mandible

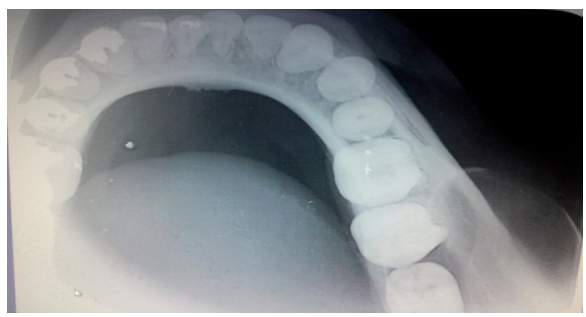

Figure 3-Mandibular Occlusal radiograph showing a well-defined radiolucent lesion with sclerotic borders noted in relation to 36,37,38 region with buccal cortical plate expansion with no lingual cortical plate expansion

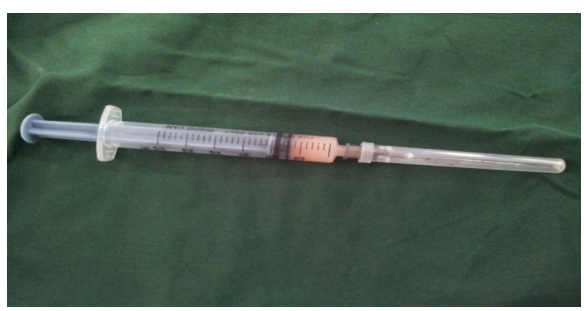

Figure 4- Milky fluid aspirate from the swelling

Figure 5- Histopathological view of the sample

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