



Calcifying Odontogenic Keratocyst: An Encounter and Recollection

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Abstract

Since the first description of the odontogenic keratocyst the twentieth century, it has been subjected to many controversies both in its behaviour and treatment. Occurrence of dystrophic calcification in the cystic wall of the odontogenic keratocyst is a very rare entity with only few of significance, quoted in the literature. This article describes a case report of a 20 year old male patient treated for odontogenic keratocyst in the right mandible. It also provides an overview of the occurrence of dystrophic calcification in the cystic wall of odontogenic keratocyst and the various theories of etiopathogenesis of these dystrophic calcifications.

Keywords: Cyst; Odontogenic keratocyst; Calcifying odontogenic keratocyst; Dystrophic calcifications

Introduction

The term 'Odontogenic Keratocyst' (OKC) perplexes clinicians and pathologists alike consequent of its changing conduct with regards to its aggressiveness. Evolution of this enigmatic entity has been affiliated with many terminologies over the ages. In literature, the keratocyst was earlier described as a cholesteatoma (Hauer, 1926; Kostecka, 1929) [1]. Robinson referred to this entity as a primordial cyst owing to its pathogenesis which is developmental in origin, arising from odontogenic epithelium. The term OKC was introduced by Philipsen in 1956. World Health Organization (WHO) accepted this terminology in 1992. In 2005, WHO christened OKC as Keratocystic Odontogenic Tumor (KCOT) owing to its aggressive behaviour, high recurrence rates and specific histological characteristics [2]. In 2017, WHO rechristened KCOT back into the cystic category citing that true neoplasm does not regress spontaneously on decompression [3].

OKC is described as "a benign uni or multi cystic intraosseous tumor of odontogenic origin, with a potentially aggressive infiltrative behaviour. It may present as a single/multiple lesion(s). The latter is usually associated with an inherited nevoid basal cell carcinoma syndrome [2].

OKC generally has a docile clinical presentation, often asymptomatic, unless secondarily infected or, if the affected teeth turn symptomatic. Large lesions can cause paraesthesia owing to pressure effects.

Histological diversity is a known feature of OKC which includes variations in the epithelial lining, basal layer and malignant transformation to name a few.

Hard tissue deposits, namely dystrophic calcifications, cartilage, dentinoid are uncommon in the connective tissue wall of the primary OKC [4]. Browne [5] reported a prevalence of 16.9% dystrophic calcifications, in primary OKC and 33.3% in syndromic OKC.

The incidence and implications of these calcifications on recurrence are unclear, either due to its rarity or due to lack of reporting. Hence, it is of paramount importance to report and follow these entities long term, to evaluate their true nature. This could aid us in formulating an ideal treatment protocol.

The following is a case report describing a 20 year old male patient treated for odontogenic keratocyst of right mandible demonstrating calcifications in the epithelial lining which is a rare histopathological appearance.

Case Presentation

A 20 year old male patient presented with a chief complaint of swelling on the right lower side

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of his jaw present since 6 months. The swelling was initially small in size which gradually increased. No complaints of pain discharge or paresthesia of inferior alveolar nerve was elicited.

On examination, a diffuse swelling was seen on the right lower side of the face approximately 2 cm in greatest dimension extending anteroposteriorly about 3 cm posterior to the corner of the lip. Superoinferiorly extending about 2 cm below the line joining corner of the mouth to the inferior border of the mandible. On palpation a bony hard swelling, non-tender, non-compressible and non-reducible was evident. Skin over the swelling appeared uninvolved. Intra-oral examination revealed bony hard swelling indicative of buccal cortical expansion in relation to 45, 46 region, retained 85, 84 was present with missing 43, 44 (Figure 1).

The orthopantomography (OPG) revealed a well defined multilocular radiolucent lesion extending from distal of 42 to distal of 48 anteroposteriorly and superoinferiorly extending from the height of the alveolar bone on the right quadrant to the base of the mandible with the displacement of the inferior alveolar canal towards the lower border of the mandible. The border of the mandible was intact. Deep vertically impacted 43, 44 were present along with a supernumerary tooth (bucco-version) between the retained deciduous molars and impacted permanent teeth. No root resorption was evident. The OPG also confirmed the presence of a supernumerary tooth between the upper first and second premolars bilaterally (Figure 2).

Cone beam computed tomography of right mandible confirmed the extensions of the radiolucent lesion along with the presence of vertically impacted 43, 44 and supernumerary tooth. Thinned out buccal and lingual cortex with associated expansion of the buccal cortical plate was present. With a well-defined breach in the buccal cortex noted at level distal to the tooth 45. This breach could be correlated to the site of biopsy which was done prior to the patient's consultation with us. A breach was also evident on the lingual cortex of 47 (Figure 3).

The lesion was provisionally diagnosed as odontogenic keratocyst of the right mandible; with the differential diagnosis of ameloblastoma, aneurysmal bone cyst and central giant cell granuloma. Aspiration cytology and incisional biopsy of the cystic lesion was performed at the thinned out buccal cortical region between 45 and 46.

Aspiration fluid was smeared and revealed the presence of scattered abundant hematoxyphilic calcifications (Figure 4).

Incisional biopsy revealed cystic lining comprising of 5 layers to 7 layers thick parakeratinized stratified squamous epithelium with surface corrugations. Basal cells are prominent and show palisading arrangement of nuclei. The unique finding in the cystic wall was the presence of hematoxyphilic; dystrophic calcifications resembling calcospherites (Figure 5 and 6). The histopathological features confirmed the diagnosis of OKC.

Enucleation of the cyst along with extraction of impacted 43, 44 and the supernumerary tooth was done followed with treatment of bony cavity with carnoy's solution and packing the same with BIPP (Bismuth Iodoform Paraffin Paste) soaked gauze.

Microscopic examination of the excisional specimen revealed similar features as described in incisional biopsy and the histopathological diagnosis of OKC was consistent with the incisional biopsy diagnosis. On follow up of 2 years, there was no recurrence.



Figure 1: Intraoperative view of the 4th quadrant.



Figure 2: Pre-operative orthopantomogram.

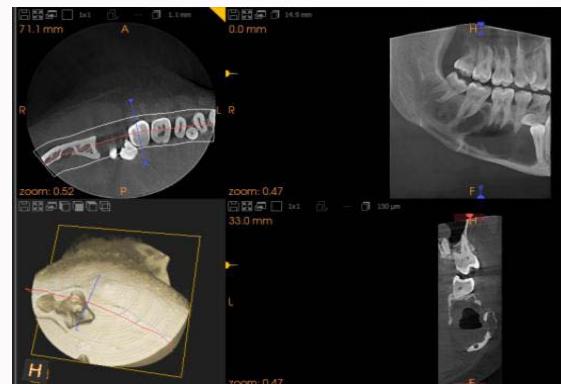


Figure 3: Cone beam computed tomography; Evidence of buccal and lingual cortical resorption.

Discussion

OKC has been recognized as a separate entity of benign cystic lesion of the jaws. This entity was also referred to as cholesteatoma, epidermoid cyst, sebaceous cyst, or primordial cyst of the jaw. In 1971, the WHO simplified the classification of jaw cysts and made the terms "primordial cyst" and "keratocyst" synonymous [6]. OKC was categorized by the latest WHO classification as a developmental, non-inflammatory odontogenic cyst that arises from cell rests of dental lamina [7,8].

The KCOTs (now renamed as OKC) are lined by a regular parakeratinized stratified squamous epithelium, usually about 5-8 cell layers thick and without rete ridges. There is a well-defined, often palisaded, basal layer of columnar or cuboidal cells. The nuclei of the columnar basal cells tend to be oriented away from the basement membrane and are often intensely basophilic. This is an important feature in distinguishing KCOT from jaw cysts with

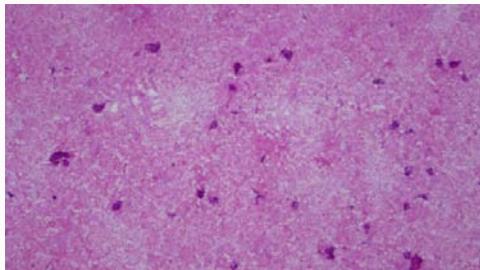


Figure 4: Aspiration smear with multiple areas of calcification.

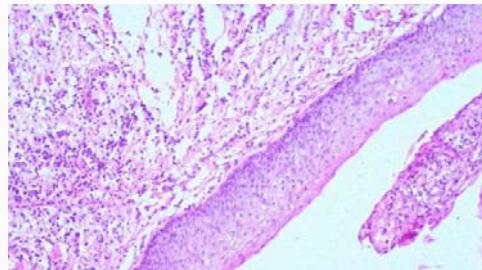


Figure 6: Lining of the cyst.

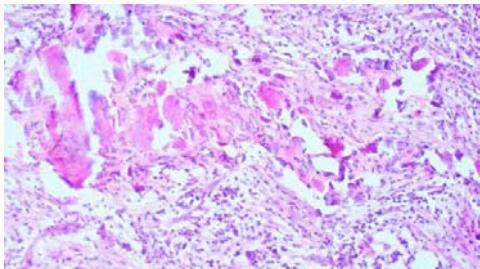


Figure 5: Calciospherulites in the cystic wall lining.

keratinization. The parakeratotic layers often have a corrugated surface. Desquamated keratin is present in many of the cavities. Mitotic figures are found frequently [2]. Separation of the epithelium from the supporting connective tissue of the cyst is common and is caused by metalloproteinases-mediated degradation of collagen in the juxta-epithelial regions [9,10].

Browne et al. [11] found a high incidence of crystalline calcium phosphates, hydroxyapatite and whitlockite, and inorganic phosphates in the aspirated fluid of the odontogenic keratocysts. This may be responsible for the increased frequency of calcific deposits in the walls of these cysts.

Primary non-recurrent odontogenic keratocyst (which is a primary keratocyst without recurrence within 5 years) shows a slightly higher prevalence for dystrophic calcifications than primary odontogenic keratocyst that recurred (which is a primary keratocyst with recurrence within 5 years). However, in recurrent odontogenic keratocyst (a keratocyst that occurs in the exact area of a previously removed keratocyst or where there is a reliable history of cyst removal in the same location), dystrophic calcification is comparatively uncommon [12].

The most common calcification in solitary KCOT is dystrophic calcifications, reported to be 4.5% to 16.8% [4,13,14]. These are usually caused by degeneration and degeneration can be the result of necrobiosis or a foreign-body reaction. Additionally, injured tissue of any kind is predisposed to dystrophic calcification. High incidence of crystalline calcium phosphates, hydroxyapatite, and whitlockite, and inorganic phosphates were found in the aspirated fluid of the KCOT. This may be responsible for the higher frequency calcium deposits in the walls of these lesions [15].

There are limited reports of presence of chondroid material in capsules of odontogenic keratocyst. There are several possible explanations for the presence of cartilage in the cyst wall which included the presence of a chondroma; persistence and displacement of vestigial remains of the Meckel's cartilage and nasal septal cartilage;

a possible metaplastic change of the fibrous connective tissue in response to chronic irritation; a possible induction of the cyst wall by the epithelial lining; and/or increased presence of "trapped" glycosaminoglycans [16-20].

Exceedingly rare is the presence of dentinoid which can occur as irregular eosinophilic masses with tubule formation or calciospherite-like mineralization. The possible pathogenesis could be the inductive changes which mesenchymal cells, can undergo, leading to calcium deposits. Bone Sialoprotein (BSP) is synthesized and secreted by bone, dentine and cementum-forming cells and has been implicated in de novo bone formation and mineralization [20]. The other alternative explanation is that the formation of dentin or dentinoid may represent a metaplastic change in the connective tissue [4].

Andrade Santos et al. [21] conducted a comparative study evaluating the immune-histochemical expression of nuclear factor Kappa B, matrix metalloproteinase 9, and endoglin (CD105) in OKC, Dentigerous cysts (DC) and Radicular cysts (RC). The results suggest that the more aggressive biologic behaviour of OKCs compared with RCs and DCs is related to the higher expression of MMP-9 and NF-Kappa B in these lesions [21].

Few of the histopathological criteria predicting recurrence of an OKC includes, subepithelial hyalinisation, basal layer budding (increased basal layer mitotic figures), separation of the epithelial lining from the capsule and the presence of odontogenic epithelial rests and/or satellite cysts within the capsule [22].

Therapeutic approaches vary in different studies from marsupialization and enucleation, which may be combined with adjuvant therapy such as cryotherapy or Carnoy's solution, to marginal or radical resection. The recurrent rate varies from approximately 20% to 62% [23-26]. The recurrence of OKC is affected by 1) enucleation with or without rupture of the cystic capsule; 2) the surgical approach used (conservative approaches such as enucleation and marsupialization vs. aggressive approaches such as total resection); and 3) the use of adjunct therapy (e.g., Carnoy's solution, cryotherapy, peripheral ostectomy) [27,28].

A review study conducted by Chirapathomsakul and colleagues [29] revealed that all types of treatment, except marginal resection, gave rise to recurrence. The recurrent lesions occurred more frequently in parakeratinized OKCs, symphysis-body region, and patients who had lesions associated with the remaining teeth and were treated by enucleation and enucleation with curettage.

Long term studies on the significance of the presence of dystrophic calcifications in the cystic walls of OKC have yet to be evaluated. Prompt reporting of these rare entities will enrich our understanding in terms of their pathogenesis, behaviour of growth

and their aggressiveness.

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Conflict of Interests

All authors declare that there is no conflict of interests.

Patient Consent

Written patient consent has been obtained from the patient to use clinical photographs and radiographs for publication purposes.

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