Keratocystic odontogenic tumour - An unusual presentation

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Abstract

Keratocystic Odontogenic Tumour (KCOT) is one of the most controversial odontogenic tumor which remains an enigma to the world of dentistry, because of its varied biological behaviour. Initially, it was considered as a cyst but in 2005, WHO reclassified it as keratocystic odontogenic tumor because of its aggressive & recurrent behaviour & placed it in the categories of odontogenic tumors. Histopathologically it has pathognomonic appearance, but in some cases, it may show unusual histopathological presentation & can be confused with other cystic odontogenic tumors. Hard tissue formation in KCOT capsule & presence of Rushton bodies are an uncommon findings. Herewith, we report a case of 26 year old male patient with an unusual histopathologic presentation of KCOT mimicking unicystic ameloblastoma (UAB) at places & also showing Rushton bodies, areas of calcification & darkly stained cells within the capsule.

Keywords: Odontogenic Tumor, Rushton bodies, Unicystic ameloblastoma.

Introduction

Cholesteatoma was the first term ever used to describe keratocystic odontogenic tumour (KCOT). (Hauer, 1926; Kostecka, 1929). The term "odontogenic keratocyst" (OKC) was used for the first time by Philipsen in 1956, who defined it as an odontogenic cyst with parakeratinized epithelial lining.(1) This entity in the past had also been referred as epidermoid cyst, sebaceous cyst, or primordial cyst of the jaw.(1,2) Shear (2003) provocatively used the term 'keratocystoma', while Reichart & Philipsen in 2004 gave the term 'keratinising cystic odontogenic tumour'. In the revised World Health Organization classification 2005, OKC was recognized as a benign odontogenic tumor & the terminology "keratocystic odontogenic tumor" (KCOT) which was proposed by Philipsen in 2005 was adopted.(3)

Hard tissue deposits namely dystrophic calcifications, cartilage & dentinoid are uncommon in the connective tissue wall of the primary KCOT.(4) Brown reported a prevalence of 16.9% of dystrophic calcifications in primary KCOT & 33.3% in syndromic KCOT (multiple jaw cysts).(5)

Rushton bodies (RBs) are exclusively seen in odontogenic cysts & are most commonly observed in radicular cysts with a reported frequency of 10% followed by dentigerous cysts (4-10%) & odontogenic keratocyst (7%).(6)

Herewith, we report a case of KCOT with an unusual histopathologic picture showing calcifications & RBs along with a review on pathogenesis of different calcifications seen in KCOT.

Case Report

A 26 year old, male patient reported to the Department of Oral Pathology & Microbiology at our institute with the chief complaint of intermittent swelling on right lower back tooth region since eight months. For the past 15 days, he experienced pain & noted a swelling that gradually increased to the present size. Extraorally**,** slightly bony hard, tender, swelling was present in the lower right body of mandible (Fig. 1A). Intraorally, diffuse swelling was seen extending from 45 to 47, causing obliteration of mucobuccal fold (Fig. 1B). Root stumps of 46 was clinically evident. Medical history, family history & habit history were non-contributory.

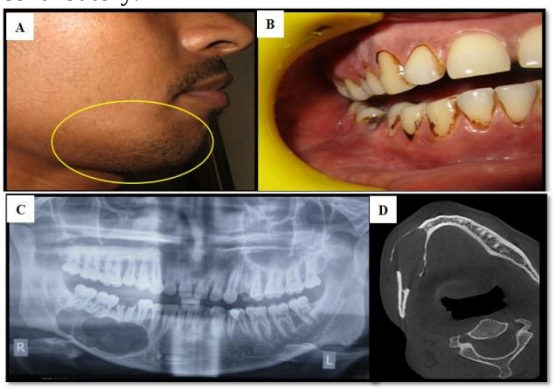

Fig. 1: A. Mild diffuse swelling in right mandibular body region; B. Diffuse swelling seen extending from 45 to 47, mucobuccal fold; C. OPG showed unilocular radiolucent lesion along with root resorption of 45,46,47,48; D. CBCT showed a hydodense lesion causing expansion, thinking and perforation of bucco-lingual cortical plates.

Orthopantmograph (OPG) & cone beam computed tomography (CBCT) was done. OPG showed a well-defined, well corticated, unilocular radiolucent lesion

seen extending from distal of 44 to distal of 48 antero-posteriorly & supero-inferiorly from apical third of 44 to lower border of mandible. Root resorption of 45,46,47,48 was evident. It also shows a well-defined radiolucency within the radiolucent interior of the lesion or 'Window formation' below the root apices of of 47 suggestive of perforation of the cortical plate (Fig. 1C).

On CBCT examination, a hypodense lesion was seen causing bucco-lingual expansion of cortical plates with perforation at certain locations (Fig. 1D).

Patient's routine blood investigations were within the normal limits. On aspiration of the lesion a dirty greenish black colour fluid was received which on microscopic wet mount examination showed RBCs, pus cells & cholesterol crystals. Considering it to be a cystic lesion it was completely enucleated.

On macroscopic examination 25x10 mm soft tissue specimen was received. The cut section revealed a cystic cavity with intraluminal proliferation.

The microscopic examination of Hematoxylin & eosin (H & E) stained slide showed cystic lumen, lining epithelium & connective tissue capsule. Cystic lumen was filled with degenerated material & necrotic debris. At one area within

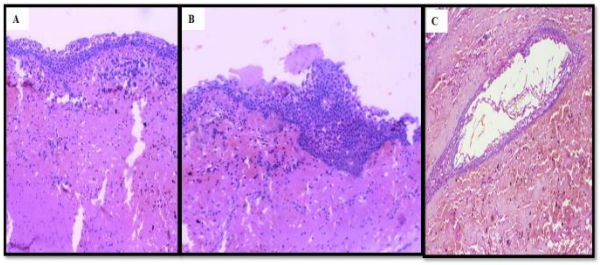

Fig. 2: A. Cystic lumen, odontogenic lining epithelium and connective tissue capsule. Flat epithelium connective Tissue interface (H & E 100X) B: Odontogenic lining epithelium showing proliferation (H & E 100x) C: Daughtercyst within the connective tissue wall showing keratinization (H & E 40X)

The cystic lumen curved eosinophilic structure were seen, suggestive of RBs. Lining epithelium varied from predominantly 4-6 cell layer thick to atrophic & proliferative in some areas. Stellate reticulum's like cells were seen at few places within the lining epithelium thus resembling unicystic ameloblastoma. The epithelium connective tissue interface was flat. At places epithelium also showed thin keratin layer & proliferation. Connective tissue capsule was loose in appearance & infiltrated with chronic inflammatory cells with areas of haemorrhage. Daughter cyst with keratinization was also seen within the capsule. (Fig.2A, 2B, 2C & 3A)The periphery of capsule had hyper cellular areas showing darkly stained cells with calcification. (Fig. 3 B)

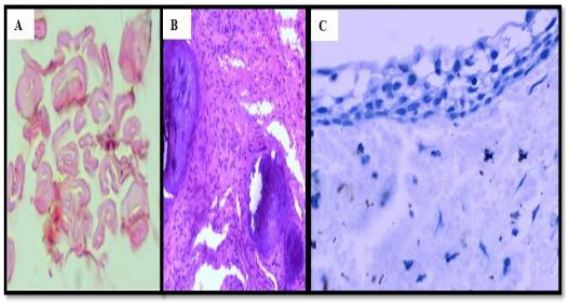

Fig. 3: A. Rushton bodies within the cystic lumen (H & E 100X); B. Peripheral area showing calcified masses with darkly staining cells (H & E 100X); C. Negative calretinin immunoexpression (IHC 400X)

Immunohistochemistry was done to rule out the possibility of unicystic ameloblastoma (UAB) or KCOT changing into UAB. However, the calretinin immunoexpression was negative (Fig. 3C).

On the basis of histopathologic & IHC findings a final diagnosis of KCOT was given & patient was kept under observation. No recurrence has been reported till date.

Discussion

KCOT was formerly considered to be an odontogenic cyst & was known as odontogenic keratocyst. The World Health Organization in 2005 reclassified & renamed the odontogenic keratocyst as keratocystic odontogenic tumor, an odontogenic tumor derived from the odontogenic epithelium.[3] KCOT is believed to originate from the dental lamina & its residues[1,7] or from the extensions of basal cells from the overlying oral epithelium.[1]KCOT occurs most commonly in the mandible than maxilla, especially involving the posterior body & ramus regions.[8,9]

In our case the lesion involved the mandibular premolar – molar area.

On radiograph KCOT usually shows a well-defined unilocular radiolucent lesion, but can also be multilocular with scalloping border.[8,9] Clinically & radiographically KCOT commonly mimics unicystic ameloblastoma. Similar findings were noted on OPG in our case, which showed unilocular radiolucency with scalloped margins.

Contrary to the usually encountered anteroposterior growth with minimal expansion in KCOT, the present case also shows buccolingual expansion & perforation, which makes it unique.

Histopathologically, KCOT usually shows classical picture of palisaded, basophilic columnar basal cell layer & a corrugated parakeratin surface layer. But in the presence of inflammation within the capsule, this classic picture is lost & the epithelium may show degeneration, lack of surface keratin, epithelial thickening, hyperplasia, spongiosis, resembling closely to stellate reticulum & the acanthomatous differentiation of ameloblastoma.[7,9,10] If the tissue sample is small & the neoplastic epithelium displays reactive changes induced by inflammation, it can

closely resemble unicystic ameloblastoma histologically.[10] The epithelial connective tissue interface may become irregular. Small satellite cysts, cords, or islands of odontogenic epithelium (7% to 26%) may be seen with in the fibrous wall.

In our case, the cyst was secondarily infected, leading to inflammatory oedematous changes within the cystic lining, giving stellate like appearance to the suprabasal cell layer. Also, the presence of proliferative epithelium & densely stained round cells within the cyst wall gave the impression of mural UAB at places, making histopathological diagnosis difficult.

The lining epithelium may also show uncommon findings like presence of Rushton bodies in 7% of cases. These measure up to about 0.1mm & are linear, straight or curved or of hairpin shape & sometimes they are concentrically laminated. In our case, RBs were also seen within the cystic lumen. Different theories have been postulated regarding their origin viz these are secretory products of odontogenic epithelial cells formed in the same way as the secondary enamel cuticle.[1] Rushton suggested, that RB represented a cuticular or keratin like product of odontogenic epithelium.[11] Others are of the opinion, of a haematogenous origin, being derived from thrombi in venules of the connective tissue that have become varicosed & strangled by epithelial cuffs which encircled them.[1]

Two different variants of RBs had been described i.e. Granular or Homogenous. It is proposed that granular appearance is because of disintegration of entrapped RBCs within the epithelium & then their calcification. RBC degeneration leads to granular RBs.[6]

The homogenous appearance of RBs is because of the exudate & transudate entrapped in the epithelium & which then undergoes calcification.[6]

Hard tissue deposits, namely dystrophic calcifications, cartilage & dentinoid are uncommon in the connective tissue wall of the primary KCOT. The most common calcification in solitary KCOT is dystrophic calcifications & is reported to be 4.5-16.8%.[2]

The various reasons of calcification as mentioned in the literature are

A. Degeneration, as the result of necrobiosis or a foreign body reaction.
B. Additionally, injured tissue of any kind is predisposed to dystrophic calcification.
C. High incidence of crystalline calcium phosphates, hydroxyapatite, whitlockite, & inorganic phosphates were found in the aspirated fluid of KCOT. This may be responsible for the higher frequency of calcium deposits in the walls of these lesions.

The presence of chondromatous tissues in the connective tissue wall of KCOT has also been reported in eight cases &may be due to metaplasia of fibrous connective tissue in response to chronic irritation.[2]

Presence of dentinoid is exceedingly rare, which can be seen as irregular eosinophilic masses with tubule formation or calcospherite like mineralization. The possible pathogenesis could be the inductive changes which mesenchymal cells can undergo leading to calcium deposits. Metaplasia has also been proposed as a reason for this phenomenon. The dentinoid formation or calcification if near the epithelium can be explained on the basis of inductive phenomenon. Thus different types of calcification can be seen in KCOT.

Conclusion

Here we conclude that histopathological picture of KCOT is very pathognomic but, because of secondary infection it may lose its classical picture & mimic other cystic tumors making diagnosis difficult. Also the presence of calcification is not a usual histopathologic finding. Whether hard tissue should be regarded as metaplastic process or represent a true inductive effect is still to be clarified. Ultrastructural studies on these calcified bodies may throw more light on the pathogenesis.

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