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Extraneous keratocystic odontgenic tumor: An unusual presentation

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Abstract

Odontogenic Keratocyst is classified as a developmental cyst derived from the enamel organ or from the dental lamina. The 2005 WHO classification uses the term 'Keratocystic Odontogenic Tumor'. Odontogenic Keratocysts manifests as radiolucencies that may appear anywhere in the maxilla or mandible including mandibular ramus (extraneous variant). The treatment of Odontogenic Keratocyst of the jaw remains controversial but the conservative treatment modality based on enucleation with or without decompression offers an effective option in inaccessible areas like ramus thereby facilitating low patient morbidity and uneventful post operative period. We report a case of 40 year old male patient which presented with swelling in the parotid area. The radiographic and CT evaluation revealed an oval radiolucent defect in left mandibular ramus with sclerotic borders. The case was treated conservatively and showed no recurrence over a two year follow up.

Keywords

odontogenic keratocyst; enucleation, mandibular ramus

Introduction

The odontogenic keratocyst (OKC) is an epithelial developmental cyst. The first case was presented by MIKULICZ as "dermoid cyst" but 'Odontogenic Keratocyst' was introduced by Pindborg (1956) to designate any jaw cyst in which keratin was formed to a large extent [1,2]. The cyst was termed "odontogenic keratocyst" by Philipsen (1956) [3] and has been one of the most controversial pathological entities of the maxillofacial region. Due to its clinicopathological features, the revised classification of Head and Neck Tumors (2005) by WHO, reclassified the odontogenic keratocyst as a benign intraosseous neoplasm, recommending the term keratocystic odontogenic tumor (KCOT) [4].

The odontogenic keratocyst (keratocystic odontogenic tumour) is an aggressive cystic lesion most frequently present in second, third and fourth decades of life at the posterior mandible [5]; in (65% to 83%) [6] of male patients [5]. The frequency of OKC has been reported to vary from 3% to 11% of odontogenic cysts [6].

From a clinical point of view, OKC is one of the most aggressive odontogenic cysts due to its high

recurrence rate, its fast growth, and its predisposition to invade adjacent tissues [3]. Malignant transformation into squamous cell carcinoma, though rare, has been reported [4]. These striking inconsistencies are thought to be related to different lengths of postoperative follow-up periods, operative techniques employed or inclusion of cases with nevoid basal cell carcinoma syndrome (NBCCS) [4].

There is a wide variety of surgical approaches depending on the size and extent of the pathology, including decompression, curettage, marsupialization, enucleation or resection, with more scrupulous surgical approaches linking to a better prognosis [4]. The present case, however highlights Odontogenic keratocyst present in the left ramus of mandible and treated with a conservative approach.

Case Presentation

A 40 year old male patient visited the outpatient department with chief complaint of enlargement in left mid facial region since three months. The clinical examination, extra orally, revealed left facial asymmetry with swelling in parotid region also extending towards the infra orbital margin (Figure 1). The swelling represented diffuse borders and only mild stretching of skin. The surface temperature of the swelling was not elevated. Intraorally there was a diffuse soft tissue edema that represented in left retro molar area. The hard tissue examination revealed missing teeth in left posterior mandibular segment and generalized loss of attachment of teeth. The Orthopantomograph revealed an oval radiolucency in left ramus area with well defined sclerotic borders measuring roughly 1.5cm in greatest dimensions (Fig 2). The coronal CT scan of maxilla however, revealed a pear shaped radiolucent defect in left ramus measuring 3x1.5 cm in dimensions (Figure 3). Also, the defect revealed focal radio opaque foci.

The biopsy was then planned with complete enucleation of the pathology. The specimen was submitted for histopathological evaluation and the sections revealed the presence of parakeratinized stratified squamous epithelium overlying fibrocellular connective tissue stroma. The parakeratinized epithelium appeared 5-6 layered with focal ameloblastomatoid proliferations of upto 10-15 layers at places. Well defined palisaded basal layer containing columnar cells with intensely basophilic nuclei and increased mitotic figures both basally and supra basally were evident. The superficial layer appeared corrugated. The connective tissue stroma revealed satellite cyst suspended in edematous stroma with collagen fibres and plump fibroblasts. A diffuse infiltration of chronic inflammatory cells along with endothelium lined blood vessels and extravasated RBCs were also revealed (Figure 4).

Based on the clinical, radiographic and histologic features a diagnosis of "extraneous odontogenic keratocyst" was made. The patient was kept under survilllience to check the recurrences, if any.

Discussion

Cysts are more common in the jaws than in any other bone because of the ubiquitous presence of epithelial rests after odontogenesis [7]. In the past, odontogenic keratocysts (OKCs) were considered to initiate from the primordium (organ at its earlier stage of development) of a tooth before mineralization had taken place and hence called primordial cysts. As the years passed, the thought gained ground that remnants of the dental lamina played a role, particularly because many OKCs seemed to have an atypical relation to teeth when presenting in the dentate area. Their presentation in the ascending ramus of the mandible was explained by the hypothesis that offshoots of the dental lamina were probably responsible

for the development of keratocysts in this region and for this reason the term ‘‘laminal cysts’’ was suggested by Toller [8]. The dental lamina arises as an invagination of the basal layer of the epithelium overlying the future mandibular and maxillary alveolar process after approximately 6 weeks of gestation. Thereafter, it loosens this connection by disintegration. Epithelial remnants, however, may persist and are most likely to be found in the gingiva or even the periodontium, because when the teeth erupt they pass the area where the dental lamina was located [8].

KCOTs are more often located in the mandible, mainly in the posterior body, the angle region and the ascending ramus [9]. Downward displacement of the inferior alveolar nerve has also been reported [9]. Smaller odontogenic keratocysts usually appear as asymptomatic; larger cysts may cause bony expansion, and be accompanied by pain [10]. These lesions are more common in males than females, occur over a wide age range, and are typically diagnosed during the 2nd, 3rd, or 4th decade [10] as also seen in our case.

Radiographically, odontogenic keratocyst can be of diverse varieties- follicular, envelopmental, replacemental, extraneous and collateral [11]. Main [2] suggested that those cysts which occur in the ascending ramus away from the teeth are referred to as ‘extraneous’. The main radiographic characteristics of OKC are unilocular radiolucent area, with scalloped borders, surrounded by a fine sclerotic line and with little or no expansion of cortical bones simulating the findings of our case. The lining is typical and the cystic cavity always contains fluid or semifluid material that may absorb X-rays to differing degrees. The luminal content can have different consistencies described as a ‘‘straw-colored fluid’’; ‘‘thick pus like’’ material; or a caseous, thick, cheesy, milk white mass. The varying consistencies replicate various densities of keratinacious debris [12]. The effort to aspirate the cystic contents in our case could not reveal any such material.

OKC, even though they grow mainly through cancellous bone, have a capsule and an epithelial lining. Thus, the content definitely generates a higher hydrostatic pressure distributed along the entire surface due to wall dialysis. Since OKC growth is slow, the pressure would be sufficient to provoke a reaction of adjacent bone, with deposition of bone matrix and minerals (sclerotic border). In the panoramic technique, the incidence of the X-ray beams is not always parallel to a wall of the lesion but at times is oblique, blurring the borders and impairing clarity. A combination of these factors may be responsible for the presence of a radiopaque halo in a segment and its absence in another, thus preventing the distinction of the lesions [13].

CT provides additional information about the contents of the lesion. The high attenuation is thought to be the result of a high protein concentration in the condensed keratin filling the lumen. Other possibilities could include hemorrhage or calcification. If the high attenuation represented calcification rather than simply a high protein content, the differential diagnosis would include a Gorlin cyst (calcifying odontogenic cyst), Pindborg tumor (calcifying odontogenic tumor), and adenomatoid odontogenic tumor. High attenuation on CT scans also could have been caused by blood. A hemorrhagic bone cyst (simple bone cyst), vascular lesion or malformation can also be considered in the differential diagnosis. However, with a vascular lesion, a change in attenuation should occur when a contrast-enhanced CT scan is compared with a non enhanced CT scan [12]. These lesions are often difficult to evaluate on the basis of their radiographic features alone. The final diagnosis must be done based on

macroscopic and microscopic examination because several other lesions (including ameloblastoma, adenomatoid odontogenic tumor, calcifiying odontogenic cyst, etc.) show similar radiographic findings [7]. Other differential diagnosis may include ameloblastoma, simple bone cyst and arteriovenous malformations [9].

Recent genetic and molecular research has lead to important breakthroughs as to the physiopathology of KCOTs. Some proliferation markers (PCNA, p53 and Ki-67) are already known to be correlated with this pathology. Other markers known to be rapidly induced in response to growth factors, tumor promoters, cytokines, bacterial endotoxins, oncogenes, hormones and shear stress, such as COX-2, may also shed some light over the biological mechanisms involved in the development of this aggressive neoplasm of the jaws [9]. PTCH ("patched"), a tumour suppressor gene involved in both NBCCS and sporadic KCOTs, occurs on chromosome 9q22.3-q31. Normally, PTCH forms a receptor complex with the oncogene SMO ("smoothened") for the SHH ("sonic hedgehog") ligand. PTCH binding to SMO inhibits growth-signal transduction. SHH binding to PTCH releases this inhibition. If normal functioning of PTCH is lost, the proliferation-stimulating effects of SMO are permitted to predominate [11].

According to Main (WHO, 2005), KCOTs have following pathognomonic histopathological features:

–Well defined, often palisaded, basal layer of columnar or cuboidal cells.

–Intense basophilic nuclei of the columnar basal cells oriented away from the basement membrane.

–Parakeratotic layers with an often corrugated surface.

– Mitotic figures frequently present in the suprabasal layers [9]

Keratinization can occur in the lining of many different types of dental cysts, but there is a specific type in which the keratin is predominantly of the parakeratinizing variety [13].

The clinically aggressive behavior of OKC is a result of the properties of the lining epithelial cells and the connective tissue capsule. Raised osmolality of cystic fluid also plays an important role in expansile growth, while an alternative view to this is that, the mural proliferations contributes to the enlargement of these lesions. This latter view is supported by authors who believed that the multilocular outline exhibited by OKC suggested a multicentric pattern of cyst growth brought about by the proliferation of local group of epithelial cells against a semisolid cystic content. The aggressive behavior of OKC was further attributed to the infolding of the epithelial lining into the capsule which suggested that this was the result of the active epithelial proliferation [1]. Through projections described as "glove fingers" and through the production of osteolytic enzymes, OKCs grow through medullary spaces, rarely deforming cortical plates [12].

In 1984, Ahlfors and others suggested that "if the OKC were recognized as a true, benign cystic epithelial neoplasm, the question of modified treatment schedules would be raised"[11]. The thin friable epithelial lining, partial surgical access, skill and understanding of the surgeon, cortical perforation, and the desire to preserve adjacent vital structures may lead to incomplete removal of the OKC [9]. There is a wide variety of surgical approaches depending on the size and extent of the lesions, including decompression, curettage, marsupialization, enucleation or resection [4]. Among the adjunctive

therapies that have been proposed, use of Carnoy's solution, peripheral osteotomy, cryotherapy and electrocautery are the most common ones [9]. Carnoy's solution, which has a mean bone penetration depth of 1.54 mm after 5 minutes, is considered enough to eliminate any epithelial islands which are bound to be located rather superficially in the defect [9]. Similar treatment modality was executed in our case and the patient showed no recurrence over a two year follow up period. Also, the main advantage of the conservative treatment is the preservation of bone structure and this proves to be less traumatic for the patient, eliminating medication and hospitalization expenses, and in most cases, avoid the need of extensive reconstructions [14].

The most important feature of the OKC is its unusually high recurrence rate that ranges from 5% to 62.5% [15]. Brannon stated that the recurrence rate of keratocyst, which was treated with enucleation alone, was 12% [15]. Woolgar et al. listed 3 different hypotheses that might explain the high recurrence rate in KCOT:

(a) Incomplete removal of the original cyst lining.

(b) Growth of a new KCOT from small satellite cysts or odontogenic epithelial rests left behind by the surgical treatment.

(c) Development of an unrelated KCOT in an adjacent region of the jaws that is interpreted as a recurrence [9].

Most recurrences are thought to present within the first 5–7 years, although recurrences have been reported to occur 9 or more years after the initial treatment. Gosau et al. reported that recurrences occurred more often in larger lesions than in smaller ones, although Kuroyanagi et al. has reported that size did not have any influence in the recurrence rates portrayed in their study. Nakamura et al. and Myoung et al. found that OKCs in the angle-ramus region of the mandible had a higher tendency to recur than those in the mandibular body. They explained this difference because of the difficulty in removing OKCs from the ramus [7].

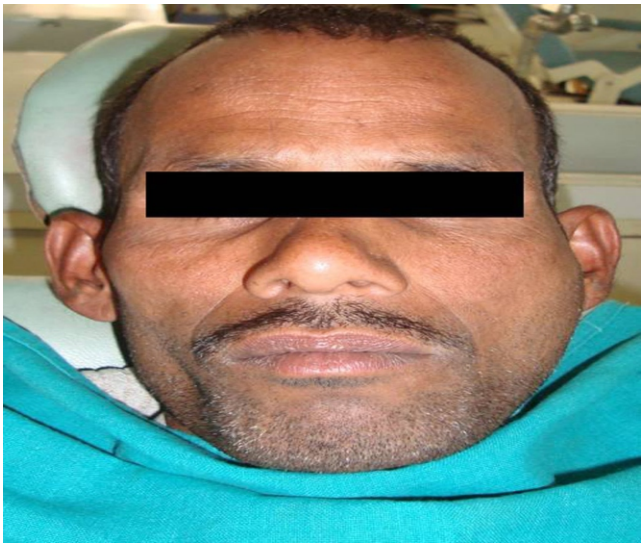


Figure 1: Extra oral photograph revealing left facial asymmetry with swelling in parotid region

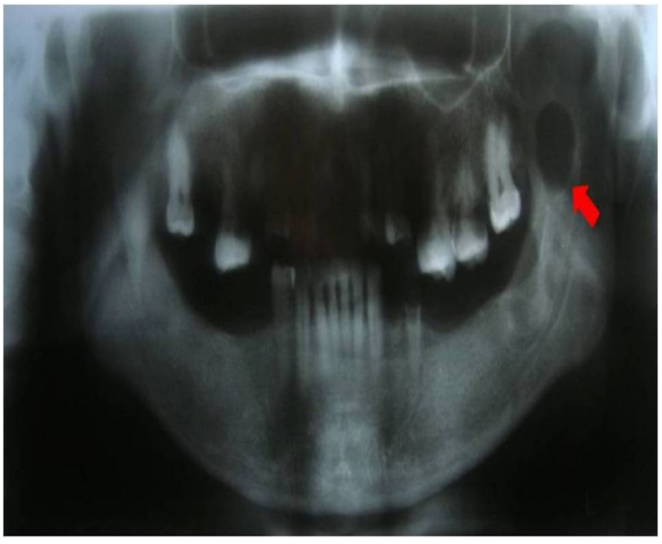
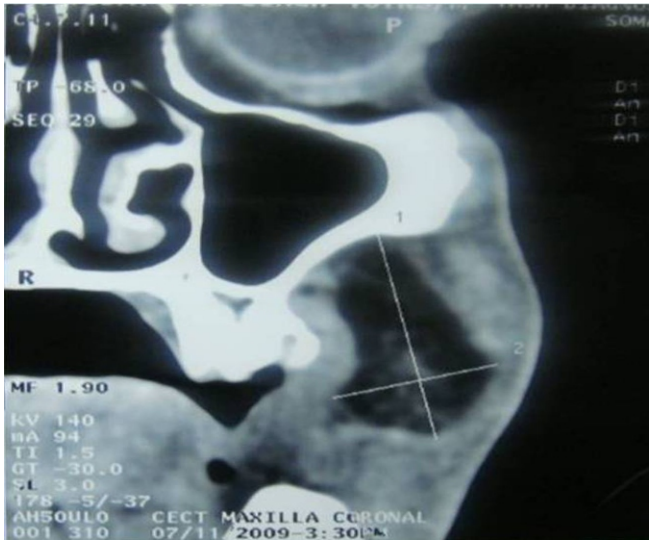


Figure 2: Orthopantomograph showing an oval radiolucency with well defined sclerotic borders in left mandibular ramus



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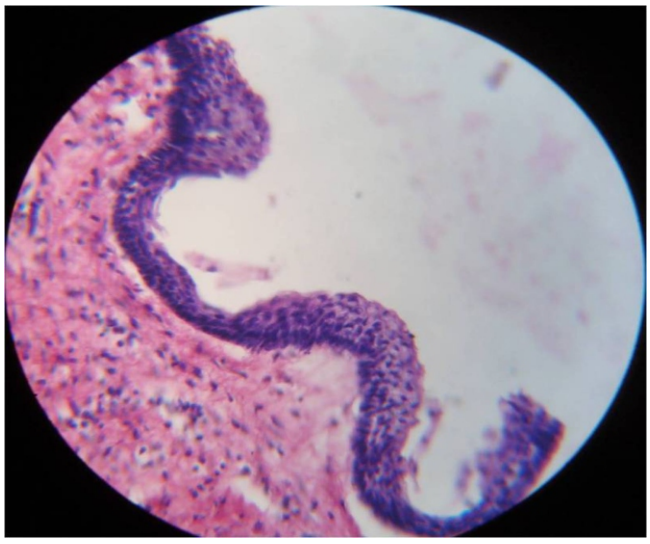

Figure 4: Photomicrograph showing cystic lining with pathognomonic features (Trinocular research microscope (Kyowa), Digital camera (Sony cyber-shot; 7.2 megapixels, Carl Zeiss lens, H& E stain, 400 X)

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