Case Report

Keratocystic Odontogenic Tumor (KCOT) in Maxillary Sinus arising from an Infected Dentigerous Cyst

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ABSTRACT

Keratocystic odontogenic tumor (KCOT) is one of the most aggressive odontogenic pathology, which is now being considered more as a benign tumor rather than it's previously known name Odontogenic Keratocyst (OKC). It's aggressive nature is attributed to its high recurrence rate. Its typical feature shows a thin, friable wall, which is often difficult to enucleate from the bone in one piece as many times it has multiple adhesions known as small satellite cysts within the fibrous wall. At times, it is also associated with bifid-rib basal cell nevus syndrome (Gorlin syndrome). Multiple surgical approaches were introduced including decompression, marsupilization, enucleation with or without adjunct (Carnoy's solution, enucleation), wide local resection followed by reconstruction. Many treatment modalities have been advocated for its management, but still its specific management is debatable. Considering its unpredictable and higher recurrence rate, WHO in 2005 categorized it under benign tumor and hence now the terminology of this pathology is changed to 'keratocystic odontogenic tumor'. Herein a case of Keratocystic Odontogenic Tumor (KCOT) in Maxillary Sinus region extending up till infraorbital rim precipitated due to an unerupted/ infected maxillary third molar is being presented.

KEY WORDS: maxillary sinus; infected maxillary third molar; KCOT (Keratocystic Odontogenic Tumour)

INTRODUCTION:

Odontogenic keratocyst (OKC), developmental epithelial cyst of jaw[1], was first described by Phillipsen in 1956. Pindborg and Hansen presented the the histologic criteria necessary to diagnose OKC[2]. Later, WHO reclassified OKC as keratocystic odontogenic tumour (KCOT) and defined it as a benign uni or multicystic, intraosseous tumour of odontogenic origin, with a characteristic lining of parakeratinised stratified squamous epithelium with a potential for aggressive, infiltrative behavior[3]". Therefore, the term KCOT reflects the neoplastic nature of the keratocyst. Since the 1960's, the nature and possible histological types of keratocysts have been under discussion for finding the most appropriate treatment plan and clinical management of KCOT[3]. Many treatment modalities, ranging from the use of simple surgical techniques such as enucleation to advanced techniques such as cryotherapy, have been recommended for the treatment of KCOT, but inspite of this its high recurrence rate have always been the subject of debate.

Odontogenic keratocysts are generally thought to be derived from either the epithelial remnants of the tooth germ or the basal cell layer of the surface epithelium and represents between 4–12% of all odontogenic cysts[4,5]. KCOTs are most common in the third decade (average 30.8–32.8 years), in men and in the mandible (70.5–76.5 %) with the majority occurring in the angle of the mandible and ramus[6,7]. In 25–40% of cases, there is an unerupted tooth involved in the lesion. KCOT tend to grow in the anteroposterior direction within the medullary cavity of the bone without causing obvious bone expansion causing its delayed observation by the patients. These are regarded as aggressive because of their high rates of recurrence and potential to destroy bone and involve adjacent soft tissues. In two recently reported large series of 256 cases and 183 cases recurrence rates were

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58.3 and 13.1 % respectively with significantly higher rates of recurrence in the mandibular angle in the first study and the maxilla in the other study[8]. In addition to its inherent aggressive nature, KCOT may rarely undergo malignant change with an incidence of 0.13–2 %[9].

Typical radiographic features such as scalloped margins or a multilobular and multilocular appearance are certainly indicative but are not unequivocal proof of OKCs because other lesions may exhibit similar radiographical signs. These features are also difficult to interpret in the maxilla because of over-projection of natural spaces such as the maxillary sinuses and nasal aperture[10]. .

CASE REPORT:

A 30 year old female patient reported to Department of Oral and Maxillofacial Surgery, with chief complaint of pain & swelling on upper left side of the face since one and half month. Patient gave the history of mild to moderate, continuous pain in the upper anterior & posterior tooth region for past one year. Pain was aggravated by chewing and relieved by medications. The patient was asymptomatic before 1 month and the swelling was slow growing and increased to the present size. Patient visited a local doctor, a week before where she was given some medications but there was no relief from pain. On clinical examination a diffused swelling was evident extending from left ala of nose to left zygomatic prominence mediolaterally and from the upper border of ala of nose to upper lip. A thorough clinical examination revealed an intra-oral draining sinus in relation to maxillary left second molar region. On palpation, the swelling was soft in consistency indicating buccal cortical plate resorption.

Orthopantomogram (OPG) (Figure 2) revealed a well-defined, unilocular radiolucent lesion associated with an impacted third molar displaced to left maxillary sinus . Mucosal thickening was also seen in left maxillary sinus.

CT Scan (Figure 3) gave an impression of large, single destructive lesion including lateral and posterior wall of maxillary sinus. The lesion was extending from left first molar region to last molar area and the associated tooth was displaced to the left maxillary sinus.

Based on the clinical examination and radiographic findings, the provisional diagnosis was an infected dentigerous cyst . Aspiration of the cystic lesion revealed a straw colored fluid of about 1 ml and the protein analysis of the same was 5.2 mg/dl. The patient underwent surgical removal of impacted third molar,

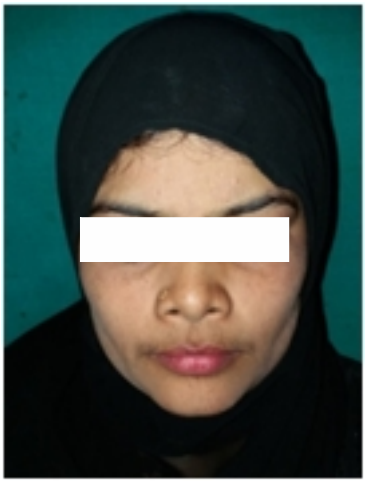

Figure 1: Profile view of Patient.

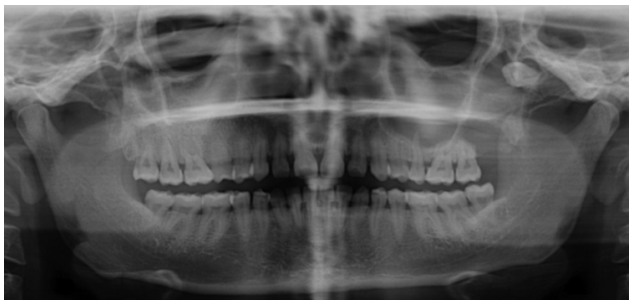

Figure 2: OPG shows a well-defined, unilocular radiolucent lesion with an impacted third molar.

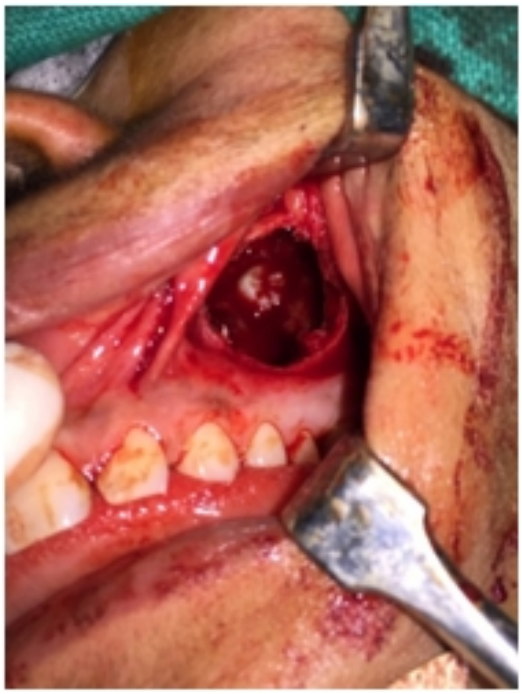

Figure 3: Intra-operative photograph showing the lesion & associated impacted 3rd molar after the incision and flap reflection.

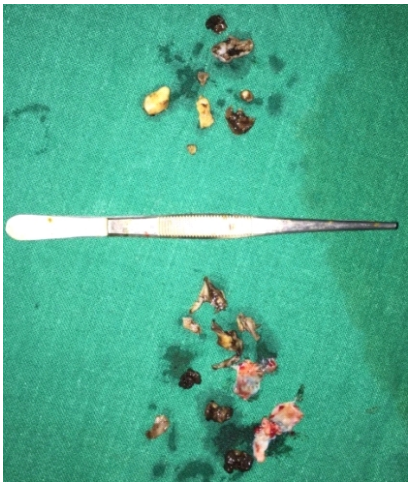

Figure 4: Surgically Resected Cystic Lining & impacted tooth.

curettage & enucleation of the lesion followed by application of Carnoy's solution used for reducing the recurrence rate and tanning of the epithelial lining of cyst {consists 3 ml of chloroform, 6 ml of absolute ethanol, 1 ml of glacial acetic acid and 1 g of ferric chloride} under general anesthesia (Figure 4 & 5). Excised tissue specimen along with the embedded third molar was submitted for the histopathological examination.

Microscopically, H & E stained sections reveals connective tissue capsule and parakeratotic stratified squamous epithelium lining. Lining of the epithelium was corrugated. About 4-6 cell layers of thick basal cells were seen having palisading arrangement. The lining of epithelium was lifted from the connective tissue capsule at some regions. Connective tissue capsule showed interposed collagen fibres with few inflammatory cell infiltrations. The diagnosis of KCOT in a pre-existing dentigerous cyst was made (Figure 6). The healing was satisfactory and the patient was followed up for two years with no evidence of recurrence. The latest radiographic examination revealed normal bony pattern.

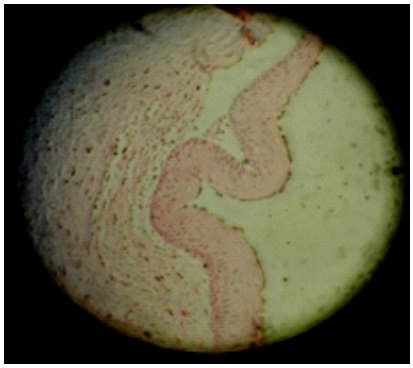

Figure 5: Histological appearance of the lesion.

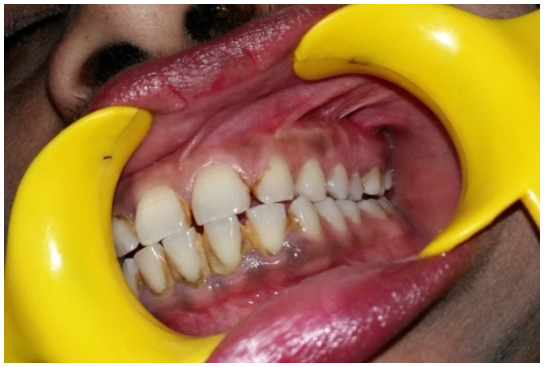

Figure 6: Post operative Excellent Healing.

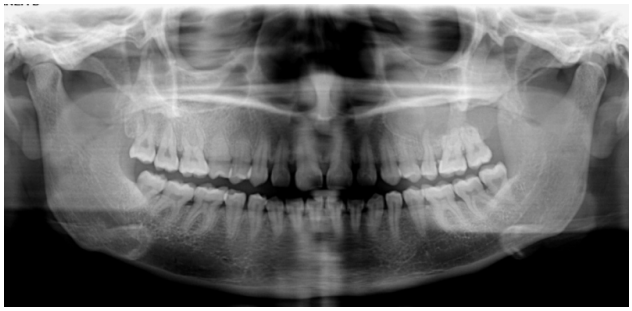

Figure 7: Postoperative Radiographs (6 month).

DISCUSSION:-

OKC are relatively common developmental odontogenic cysts and account for 10–12% of all jaw cysts[11]. An OKC usually occurs as a single lesion. Multiple lesions are associated with the nevoid basal cell syndrome (Gorlin– Goltz syndrome)[12]. The OKCs originate either from epithelial remnants of tooth germ in the mandible and maxilla or the basal cell layer of the overlying surface epithelium[13].

The peak incidence is in the second and third decades of life with a gradual decline thereafter. Peak incidence in both sexes occur in the third decade frequency of occurrence is higher in male than female[14]. Though it can occur in any part of mandible and maxilla, but in 70% of the cases, arise in the posterior body and 6.9% at the symphyseal region of the mandible . The mandible is the more favoured site than the maxilla. In approximately 50% of patients, lesion is asymptomatic. In other cases pain, swelling, expansion, drainage, and bone perforation are reported . These lesions grow to sizes larger than any other odontogenic cyst. They more often penetrate the bone rather than expand it and grow in an anterior to posterior direction . Despite this aggressive growth, they often remain asymptomatic[15].

An OKC has a fibrous wall lined by epithelium with a thin layer of stratified squamous epithelium on histopathological examination. This

epithelium has a basal cell layer of six to eight cells thick and a lining of flattened keratotic epithelial cells. The formed keratin lines the luminal surface of the epithelial cells in a slightly wavy of corrugated pattern . The luminal content can have different consistencies as a "straw-coloured fluid"; "thick pus like" material; or a caseous, thick, cheesy, milk white mass. The varying consistencies reflect various densities of keratinaceous debris. Histologically OKCs have been classified into parakeratotic and orthokeratotic subtypes. These types refer to the histological characteristics of the lining and the type of keratin produced. Compared with the parakeratotic subtype, the orthokeratotic subtype produces keratin closely resembling the normal keratin produced by the skin. The keratin (orthokeratin) does not contain nuclei. The parakeratotic subtype has a more disordered production of keratin. The keratin contains nuclei and is referred to a para keratin. The parakeratotic type is the most frequent (80%) and has a more aggressive clinical presentation than the orthokeratinized variant[16].

Immuno histochemical studies have shown higher levels of interleukin- 1a (an inflammatory multifunction cytokine) in OKC compared with dentigerous cyst . Interleukin-1a is thought to play a crucial role in expansion of OKCs by inducing the secretion of keratocyte growth factors from interactive fibroblasts. Nature of the cyst lining before and after decompression with cytokeratin stains have reported positive cytokeratin-10 staining in pre-decompression biopsy and negative cytokeratin-10 stain in post-decompression specimen indicating a return to more normal oral epithelium[17]. BCL-2, is an anti-apoptotic protein and has been shown to be strongly and consistently expressed by all basal cells of OKCs but not in other odontogenic cysts. Hence it was likely that BCL-2 could be used to differentiate keratocyst lining from normal epithelium[17].

Radiologically lesion characteristically presented as an extensive well defined area of bone destruction. The border of these lesions appears to be thinly sclerosed . Characteristic radiographic features of OKCs are an intinctly corticated, often scalloped, border, expansion toward the lingual side, displacement of developing teeth and/or separation of resorption of the roots of erupted teeth and extrusion of erupted teeth, a radiolucent lumen, and occasionally a cloudy or milky appearance of the lumen on the panoramic radiograph. Multiple or bilateral cysts are suggestive of basal cell nevus syndrome . CT provides additional information about the contents of the lesion. The high attenuation is thought to the result of a high protein concentration in the dense keratin filling the lumen[18].

Neville et al. after studying 18 cases of OKCs of maxillary midline concluded that it is important to include OKC in the differential diagnosis of anterior midline maxillary radiolucency, especially when they occur in older individuals. Since this case being an OKC occurring in the maxillary midline of the younger patient, the young age cannot be excluded from the same. Its presentation with impacted Third molar tooth, location and draining sinus made us provisionally diagnose the case as infected dentigerous cyst. Hence it is important for the clinician to consider OKC in the differential diagnosis of lesions crossing the maxillary midline, when they occur in a younger patient also. WHO's reclassification of this lesion from cyst to tumour underscores its aggressive nature and should motivate the clinician to manage the disease in a correspondingly aggressive manner[19].

There are different opinions regarding the management of OKC. These cysts are most aggressive forms of odontogenic keratocysts because of the high recurrence rates due to the presence of epithelial remnants of satellite cysts in the osseous margins. The parakeratinized variant has a higher recurrence rate than the orthokeratinized variant. For this reason a more aggressive treatment has been advocated[20]. The recommended treatment is; curettage with peripheral Ostectomy, cryosurgery (curettage with liquid nitrogen therapy, curettage plus application of Carnoy's solution, localized en bloc resection and occasionally segmental resection , enucleation with postoperative intra-oral suction and rinsing the bone defect with 3% hydrogen peroxide in order to detect and remove eventual remains of the capsule[21]. The goal of using Carnoy's solution and cryosurgery is to kill epithelial remnants and dental lamina in the osseous margins. Use of liquid nitrogen maintains the osseous structure and facilitate new bone formation. Carnoy's solution is a tissue fixative that penetrates bone to a depth of 1.54 mm and decreases risk of recurrence, by killing the epithelial remnant or satellite cysts[18].

Cyclopamine, a plant-based steroidal alkaloid, blocks the activation of sonic hedgehog (SHH) pathway, therefore makes it a potential "mechanism- based" therapeutic agent for those human tumours whose pathogenesis involves excess SHH pathway activity. Antagonists of SHH signalling factors could also be effectively used to treat KCOT .

The suggested strategies include re-introduction of a wild- type form of PTCH (a tumour suppressor gene), inhibiting SMO molecule (an oncogene) by synthetic antagonists and suppressing the downstream transcription factors of the SHH pathway. Intracystic injection of an SMO protein-antagonist has the greatest potential as a future treatment option[19].

The most important feature of OKC is its high recurrence rate ranging from 5 to 62.5% . Recurrence is documented even after 10 years of follow up . Recurrence is usually characterized radiologically by evidence of further bone destruction with or without clinical evidence of infection[25].

CONCLUSION:

To conclude, the present report describes a rare case of keratocystic odontogenic tumor (kcot) in maxillary sinus arising from a infected dentigerous cyst. We have done enucleation with application of carnoys solution followed by iodoform dressing for 6 months. There was no sign of recurrence on a follow up. Patient is asymptomatic and under regular follow-up.

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