Case Report

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EXPANSILE KERATOCYSTIC ODONTOGENICTUMOR OF MANDIBLE TREATED SURGICALLY: A CASE REPORT

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ABSTRACT:

The term OKC was first described by Philipsen in 1956. This cyst arises from the cell rests of dental lamina. It can occur both in maxilla and mandible but most commonly seen in posterior part of mandible. The lesion was traditionally known as odontogenic keratocyst it is renamed by WHO in 2005 as "Keratocystic Odontogenic Tumor" because of its local destructive behavior. It is the benign intraosseous lesion of the jaw because of potentially aggressive behaviour and high recurrence rate.The diagnostic approach is based on medical history, the clinical appearance and the radiographic appearance. The diagnosis may be confirmed by the histopathology report. The purpose of this case report is to discuss the prevalence, clinical features, differential diagnosis, radiological findings and treatment of Keratocystic Odontogenic Tumor.

KEY WORDS: Keratocystic Odontogenic Tumor, Odontogenic Keratocyst, nevoid basal cell carcinoma, Gorlin-Goltz syndrome

INTRODUCTION

The keratocystic odontogenic tumor (KOT) is defined as a benign unicystic or multicystic, intraosseous tumor of odontogenic origin with a characteristic lining of parakeratinized stratified squamous epithelium and potential for aggressive, infiltrative behavior.[1] Odontogenic keratocyst (OKC) was a neoplasm described by Philipsen in 1956 later it was confirmed by Browne in 1970 and 1971 Initially it was believed to be a benign, but potentially aggressive and recurrent, odontogenic cyst.[2]Pindborg and Hansen in 1963 describe the essential features of this type of cyst. KCOT is a unique cyst because of its pathognomic microscopic features, aggressive behavior, and high recurrence rate. It constitutes 10-12% of all odontogenic cysts.[3] . Its origin is from remnants of dental lamina. It is most often seen in the mandibular ramus and angle region (69% - 83%). The most characteristic clinical feature of OKC is the high frequency of recurrence (0% - 60%; Nakamura et al, 2002).[4] It is most commonly seen in 3rd to 4th decade of life with male to female ratio of 2:1. It can occur as a single entity or in association with nevoid basal cell carcinoma (Gorlin-Goltz Syndrome). It has two histological varities parakeratinized and

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orthokeratinized OKC. Histologically, parakeratinized OKC's is having higher recurrence rate as compared to the other variety, orthokeratinized OKC's (Shear, 2003). The term keratocystic odontogenic tumor was given because of the aggressive behavior of the lesion and associated chromosomal and genetic abnormalities.

Thus, the aim of this case report is to discuss the aspects regarding the early diagnosis,and prompt treatment of KCOT.

CASE DETAIL:

A 26 year old male comes to the Oral Medicine and Radiology Department, Shree Guru Gobind Singh Tricentenary University with a chief complaint of swelling in lower left back region since 1-2 years and pain in same region since 3 months.

On extraoral examination a diffuse swelling of size 6 x 8 cm is seen with respect to lower left 1/3rd of face in region of mandible extending from 2cm behind angle of mouth to 1 cm behind posterior border of ramus of mandible and 2cm below inferior border of mandible. Skin over the swelling was normal with no localized increase in temperature. (Fig.1, Fig.2)

Intraoral examination revealed no obvious swelling on inspection but on palpation a diffuse swelling is seen in vestibular region extending from mesial aspect of 36 to distal aspect of 37 till left retromolar region. (Fig.3, Fig.4) 38 was clinically missing.

Panoramic radiograph revealed a well definedmultilocular radiolucency with corticated borders of size 7 X 4 cm is seen associated with mesioangularly impacted 38 extending vertically upward upto ramus of mandible on left side . It causes thinning of posterior inferior cortex of mandible.(Fig.5)

Mandibular cross sectional radiograph shows buccal cortical plate expansion extending from region of 33 to 37 with no cortical perforation and discontinuity of inferior border of mandible.(Fig.6)

CT (axial, coronal and saggital section) shows a lytic lesion with cortical thinning seen involving the left ramus and angle of mandible. There is cortical breach at multiple sites especially along medial cortex. Multiple complete or incomplete bony septations are also seen. The 38 appears to be impacted within the lesion.(Fig.7.a,7.b)

With a provisional diagnosis of odontogenic keratocyst, surgical enucleation along with extraction of impacted 37, 38 was done (Fig.8) , histopathological examination shows cyst wall lined by stratified squamous epithelium 8-10 cell layer thick with corrugated parakeratinized surface. Basal cell layer exhibits palisading appearance. The epithelial lining is supported by a connective tissue with dense collagen

bundles, blood vessels and inflammatory cells.(Fig.9) which was suggestive of keratosystic odontogenic tumor.

Patient returns every six months for follow-up.

DISCUSSION:

The OKC is histopathologically and behaviorally unique. It is the most aggressive and recurrent of all the odontogenic cysts and shows characteristics resembling both a cyst and a benign tumor. This term was replaced in the 2005 ,World Health Organization Classification of the Head and Neck Tumors, by the term "Keratocystic Odontogenic Tumor" (KCOT). The WHO believed that this new term truly reflects the neoplastic nature of the OKC. This reclassification was determined by the clinical features of this tumor, including its potential for locally destructive behavior, high recurrence rate, and tendency to multiplicity.[3]

There is two types of OKC on the basis of origin one that originate from dental lamina rests or from the basal cells of oral epithelium and are thus called as primordial-origin odontogenic keratocysts. (Fig.10 a) The remaining 40% arise from the reduced enamel epithelium of the dental follicle and are thus dentigerous-origin odontogenic keratocysts (Fig.10 b) This clinical identification is of some importance because recurrences are more frequently seen after treatment of the primordial-origin type.[3]

Clinical features: The majority of patients are in the age ranges of 20-29 and 40-59, but cases ranging from 5 to 80 years have been reported. The distribution between sexes varies from a male to female ratio of 1.6:1, except in children. Odontogenic keratocysts may occur in any part of the upper and lower jaw with the majority occurring in the mandible, most commonly in the angle of the mandible and ramus.[5]

It grows mostly in anteroposterior dimension and thus lesions may attain remarkable size without significantly deforming the jaw. The particular tendency to rapid growth is due to higher activity of the epithelial cells of the cyst lining stimulating osteolytic activity of prostaglandin substances in the cell population of the cyst lining and higher accumulation of hyperkeratotic scales in the lumen of the cyst with resulting greater difference in hydrostatic pressure.[6]

They almost occur within bone, although few cases of peripheral KCOT have been reported. Patients may have following signs and symptoms like swelling, pain and discharge or may be asymptomatic. Multiple KCOT is associated with nevoid basal cell carcinoma syndrome (NBCCS) or Gorlin-Goltz syndrome.[7]

Lindeboom et al. reported on the multiple occurrences of OKCs in

connection with the oral, facial and digital syndrome (OFDS) which was described first in 1954. This syndrome includes the typical combination of cleft lip and palate with malformations and anomalies of tongue and finger shape (syndactyly, polydactyly, brachydactyly, and clinodactyly).[6]

KCOTs have a high recurrence rate, reportedly between 25% and 60%.In 1976, Brannon proposed 3 mechanisms for KCOT recurrence: incomplete removal of the cyst lining, growth of a new KCOT from satellite cysts (or odontogenic rests left behind after surgery) and development of a new KCOT in an adjacent area that is interpreted as a recurrence.[7]

A thorough review of the biological behavior of this aggressive pathological entity of the jaws and a molecular (growth factors, p53, PCNA and Ki-67, bcl-2) and genetic (PTCH, SHH) alterations associated with this odontogenic neoplasm, provides a better understanding of the mechanisms involved in its development and also support the concept that the KCOT should, be regarded as a neoplasm.[8]

Histopathology: Pindborg, phillipsen and Henriksen (Pindborg*et al*., 1962) suggested series of histological features for the diagnosis of OKC which includes:

OKCs/KCOTs consist of thin epithelial layer, composed of 8 to 10 cell layers. The basal cell layer shows palisaded arrangement. Basal cells are columnar or cuboidal and have hyperchromatic nuclei with picket fence arrangement. In some cases there is invasion of the basal cell layer into the region of surrounding connective tissue is seen along with formation of satellite micro-cysts. The fibrous walls of the cyst may be relative thin and usually without inflammatory cell infiltrates. On the basis of histopathology there are two types of KCOT, parakeratinized and orthokeratinized. Orthokeratinized type consist of prominent granular layer lying immediately under the thin surface layer. Others findings are satellite cysts, daughter cysts (7-30%), solid epithelial proliferation, cholesterol crystals and Rushton bodies can be seen.

Radiographic features: On the radiography KCOTs appear as well-defined radiolucencies, which can be either unilocular or multilocular. Large unilocular KCOTs can be difficult to distinguish from cystic ameloblastoma.[9] The lesion's proximity to the tooth may cause displacement and/or root resorption, although displacement (28.3%) is more commonly seen than resorption (5%)[10]

Conventional radiographic imaging, such as panoramic views and intraoral periapical films, occlusal radiographs in most cases are adequate to determine the location and estimate the size of KCOT. Advanced imaging techniques like CT and MRI can be useful in large cases

involving the maxillary sinus and the rare cases that extend to the skull base[9]

Radiological Types of keratocyst

1. Replacement type: Cyst which replaces the normal teeth and forms in place of normal teeth.

2. Envelopmental type: Cyst which covers an adjacent unerupted tooth.

3. Extraneous type: Cyst which occur away from the teeth bearing region of jaw.

4. Collateral type: Cyst which occurs adjacent to the root of teeth.

CT can be used to determine the extent and site of any cortical perforations with involvement of soft tissue. In the present case, axial CT images shows a significant expansion as well as perforation of the buccal and lingual cortical plates of the mandible. Usually significant expansion may occur in the upper ramus and coronoid process, but the buccal and lingual cortical plates of the mandible revealed slight expansion only.

Ultrasound is an noninvasive and cost-effective modality, and is recommended as a complementary imaging method for intraosseous lesions of the jaw. However, it is not useful in case of intact and thick cortical bones, usually the growth of KCOT is more in mesial-distal direction than in the buccal-lingual, thus maintaining the vestibular and lingual/ palatine bones intact. Therefore, US examination could be inconclusive in some KCOT lesions.[11]

Differential diagnosis:

Radiographically:Dentigerous cyst (40%), Residual cysts, radicular cyst, Primordial cyst (25%), Unicystic ameloblastoma, A-V malformation, Fibroosseous lesion at initial stages.[12]

Treatment: Treatment of KCOTs remains a controversial subject .The choice of treatment approach should be based on the size of the cyst, recurrence status, and radiographic evidence of cortical perforation.[8]Radical surgery including complete resection with or without continuity defect, resection of complete lesion with less recurrence rate after surgery has been advocated.

More aggressive treatment - resection or enucleation supplemented with Carnoy's solution with or without peripheral ostectomy - results in a lower recurrence rate than enucleation alone or marsupialization.[8]

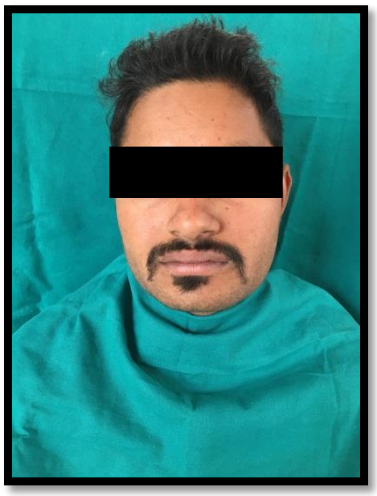
CONCLUSION:

In any unilocular or multilocular lesion of the jaw one should include KCOT in differential diagnosis. An important characteristics of the KCOT is its tendency to grow internally with minimum expansion and high recurrence rate hence a minimum of 5 yr follow up is necessary postoperatively.

REFERENCES:

1. Contar et al. Keratocystic odontogenic tumor: case report. Odonto 2011; 19 (37): 25-32
2. Motwani MB. Keratocystic odontogenic tumor: case report and review of literature.Journal of Indian Academy of Oral Medicine and Radiology.April-June 2011,23(2):150-154.
3. BelmehdiA ,Chbicheb S, Wady WE. Odontogenic Keratocyst Tumor: A Case Report and Literature Review. Open Journal of Stomatology, 2016, 6:171-178
4. Maria A, Sharma Y, Chabbria A. Marsupialization as a treatment option of a large Odontogenic keratocyst: A case report with the review of literature. People's Journal of Scientific Research, 2012;5(1):46-51
5. Valiente Rivera, N. Ghanee,E.A.Kenny,K.H.Dawson.OdontogenicKeratocyst: The Northwestern USA Experience.The Journal of Contemporary Dental Practice, Volume 1, No. 2, Winter Issue, 2000.
6. Pazdera J et al. Odontogenic keratocysts/keratocystic odontogenic tumours: biological characteristics, clinical manifestation and treatment. Biomed Pap Med FacUnivPalackyOlomouc Czech Repub. 2014 Jun;158(2):170-174.
7. Shah K, MistryJ, Koppikar R, KaragirA.Keratocystic Odontogenic Tumour: Current concepts, theory and presentation of 2 contrasting cases. IOSR Journal of Dental and Medical Sciences (IOSR-JDMS),2013;9(2):49-53.
8. Yildirim G et al.Conservative Treatment Protocol for Keratocystic Odontogenic Tumour: a Follow-up Study of 3 Cases. J Oral Maxillofac Res 2010 (Jul-Sep) | vol. 1 | No 3.
9. Ali M, Baughman RA. Maxillary odontogenic keratocyst: a common and serious clinical misdiagnosis. J Am Dent Assoc. 2003 Jul;134(7):877-83. [Medline: 12892445] [FREE Full Text]
10. Haring JI, Van Dis ML. Odontogenic keratocysts: a clinical, radiographic, and histopathologic study. Oral Surg Oral Med Oral Pathol. 1988 Jul;66(1):145-53. [Medline: 2457195]
11. Sumer AP.Keratocystic odontogenic tumor: case report with CT and ultrasonography findings. Imaging Science in Dentistry 2012; 42 : 61-4
12. Passi D.et al. Odontogenic keratocyst or keratocystic odontogenic tumor-journey of OKC from cyst to tumor to cyst again:comprehensive review with recent updateson WHO classification 2017. International journal of current research.2017 july;9(7):54080-54086

FIGURES:



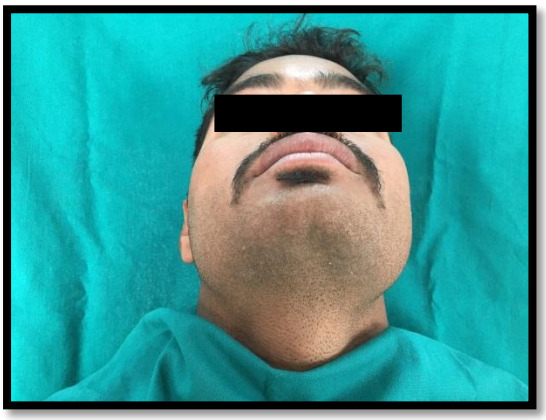
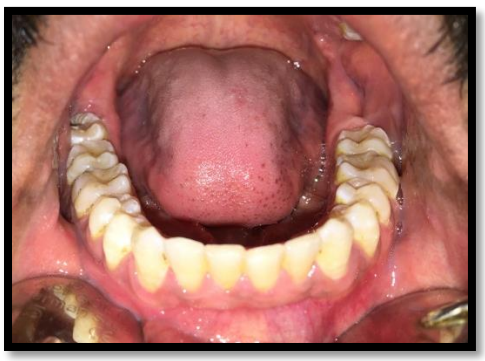


FIG.1, 2 :Extraoral pictures showing swelling on left side of lower 1/3rd of face



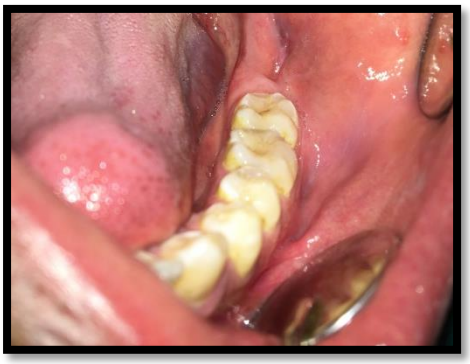


FIG.3,4 Intraoral examination revealed no obvious swelling on inspection

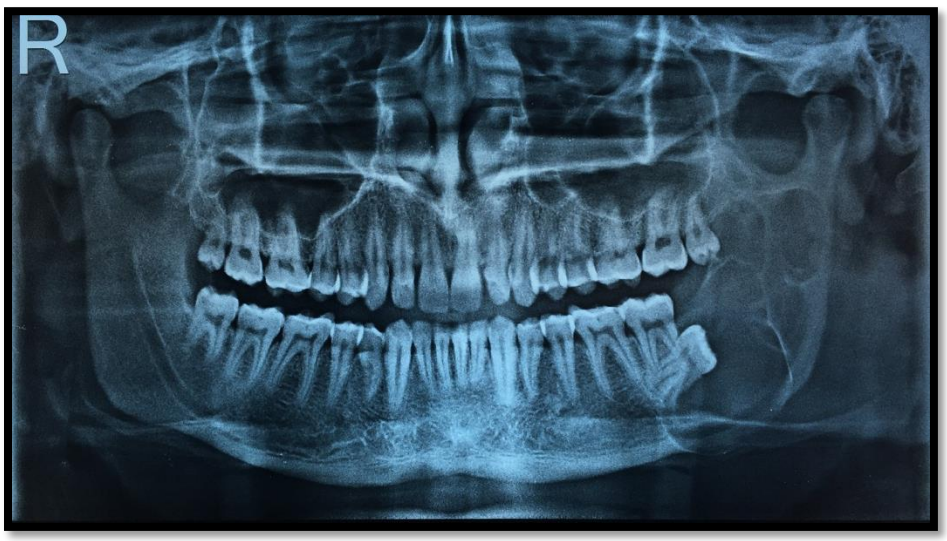


Fig.5: OPG (Multilocular Radiolucency In Left Angle And Ramus)

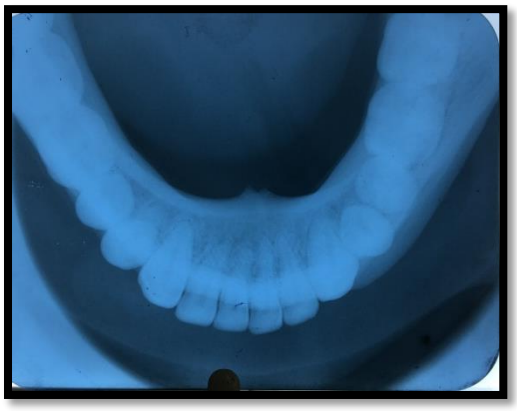


FIG .6: Mandibular Cross-SectionalOcclusal

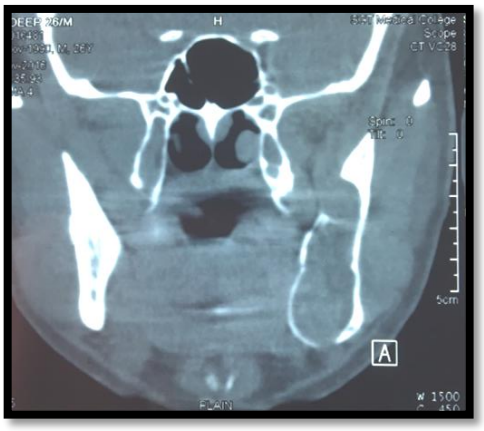


FIG.7a: Coronal CT Section

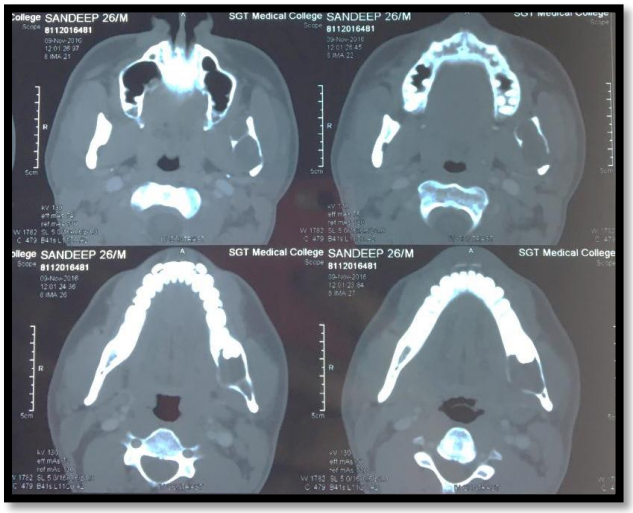


FIG .7b:Axial CT Scans multilocular radiolucency

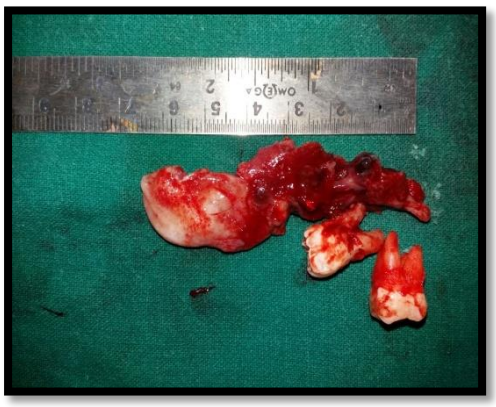


FIG. 8: Operatory biopsy with extracted 37,38 with impacted 38

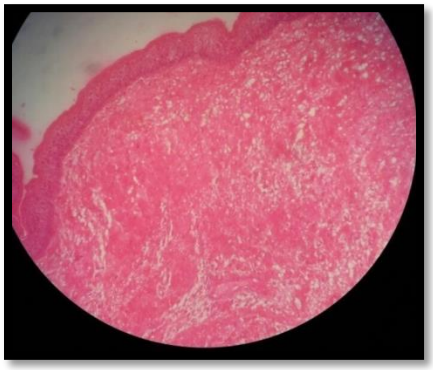


FIG.9: Histopathology (H&E staining)

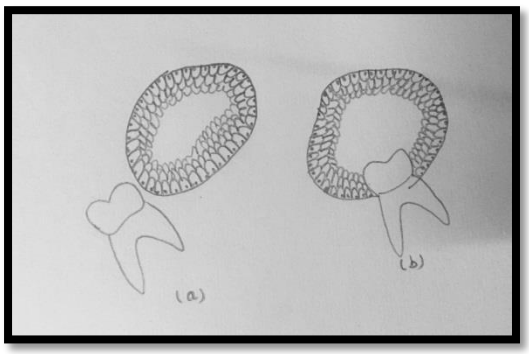


FIG.10 a)Primordial origin OKC ; b)Dentigerous origin OKC