Non-Syndromic Multiple Keratocystic Odontogenic Tumor: A Rare Case Report and Review of Literature

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ABSTRACT

KeratocysticOdontogenic tumor (KCOT) has been the topic of various researchers and is distinctive for its potentially aggressive behavior and tendency for multiplicity. It is anodontogenic tumorthat exhibits a hyperkeratinizedepithelial lining. The lesion originates from the cell-rests of the dental lamina rather than from the reduced enamel epithelium or the cell rests of Malassez. It constitutes for 3.11% of all odontogenic cysts. It often occurs as a solitary lesion at the angle of mandible and multiple lesions may occur in association with systemic changes. Here is a rare case report ofmultiple KCOTsassociated with an ovarian cyst in 30 years old female.

Key words: *GorlinGoltz syndrome, KeratocysticOdontogenic tumor, Odontogenic*

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CASE REPORT

A 30years old female patient reported to the Department of Oral Medicine &Radiology with the chief complaint ofpain in left lower back teeth region since 1 year and swelling on right side of lower 3rd of face since 6 months.History of presenting illness revealed pain in the same teeth region since 1½ years which was gradual in onset, dull, aching, and non-radiating for which patient consulted the local dentist and the pain relieved on takingmedication and it again arose after 6 months.Patient noticed a swelling on the right side of lower 3rd of face that increased in size gradually with time to attain the present size with no history of trauma, fever, weight loss, numbness or paresthesia& similar swelling elsewhere in the body.Medical history and family history was non-contributory.

Extraoralexamination revealedgross facial asymmetry with unilateral, solitary, well defined oval shaped swelling in the right parasymphyseal regionwhich was 2.5cms in maximum diameter. On palpation, it was non tender, firm in consistency & non-fluctuant(**Fig. 1**). No abnormality of skin & eyes was detected. Intraoral examination showed a solitary, diffuse sub mucosal swelling in mandibular right labial & buccal vestibular region with respect to 43,44,45 region which was 2cmx1cm in maximum dimensions with overlying erythematous mucosa causing the obliteration of labial &buccal vestibule.On palpation, swelling was soft in consistency, mild tender & compressible. Hard tissue examination revealed retained tooth fragment irt 73 and distolabially tilted 33.(**Fig. 2**)

Based upon the history and clinical examination, provisional diagnosis of Benignodontogeniccystic lesion of right parasymphysis & retained 73 was made. The differential diagnosis of Central giant cell granuloma, Keratocystic Odontogenic tumor, dentigerous cyst & Unicysticameloblastoma were considered. The electric & thermal pulp testing of 43,44 & 45 showed normal response.The 2 ml of cystic fluid was aspirated which was blood-tinged.(**Fig. 3**) Hematological investigations revealed raised ESR & decreased neutrophil count. The periapical radiographs of 43, 44 and 73 showed well defined, homogenous radiolucencies in respect to periapicalregion of 43, 44, 45and 73, 33, 34 causing diversion of roots of 33 & 34 withretained tooth fragment in respect to 73 respectively.(**Fig. 4**)

OPG showedsix cysts-like, homogenousradiolucencies in the mandible extending from mid-body region of right side of mandible to the left side of ramus of mandible having corticated borders with Impacted 38(**Fig. 5**). No abnormality detected in the paranasal sinus view (**Fig. 6**) & chest radiograph **(Fig. 7**). Ultrasound scan of whole abdomen revealed left ovarian cyst measuring 36x26mm filled with organized course echoes. Spine radiography did not show any abnormality.(**Fig. 8**)

Incisional Biopsy was done at the site of swelling under local anesthesia and the specimen was sent for histopathological examination which showed 8-10 cells thick corrugated parakeratinized, stratified squamous epithelium with palisaded basal layer & absence of rete ridges. The underlying connective tissue is loose &fibrous suggestive of Odontogenickeratocyst.(**Fig. 9**)

Based upon the history, clinical examination & investigations, the Final Diagnosis of Non-Syndromic Multiple Odontogenickeratocysts of Mandible was made.The patient was then referred to the department of Oral & Maxillofacial Surgery where the lesions were enucleated under general anaesthesia followed by intracavitaryCornoy's solution application and Iodoform dressing.**(Fig. 10)** Patient is still under follow-up.(**Fig. 11**)

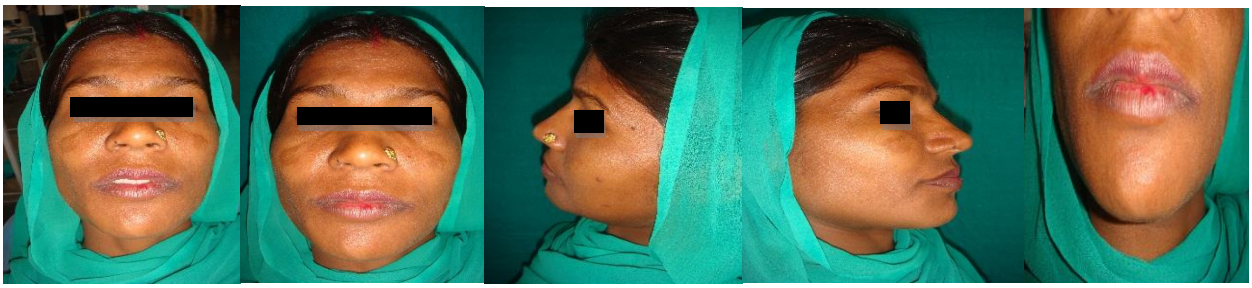

Fig. 1: Extraoral Photographs of a patient with swelling in right Parasymphysis

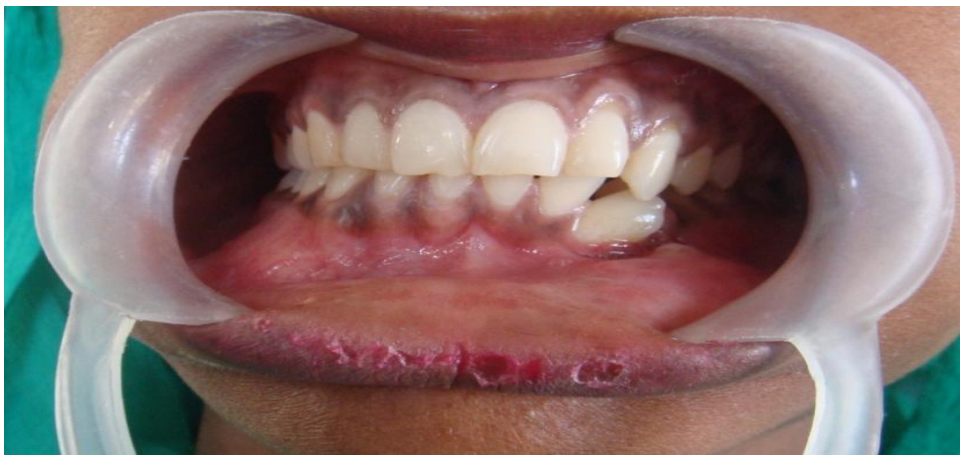

Fig. 2: Intraoral Photograph showing intraoral swelling in right mandibular Vestibular region

Fig. 3: **Aspirated fluid**

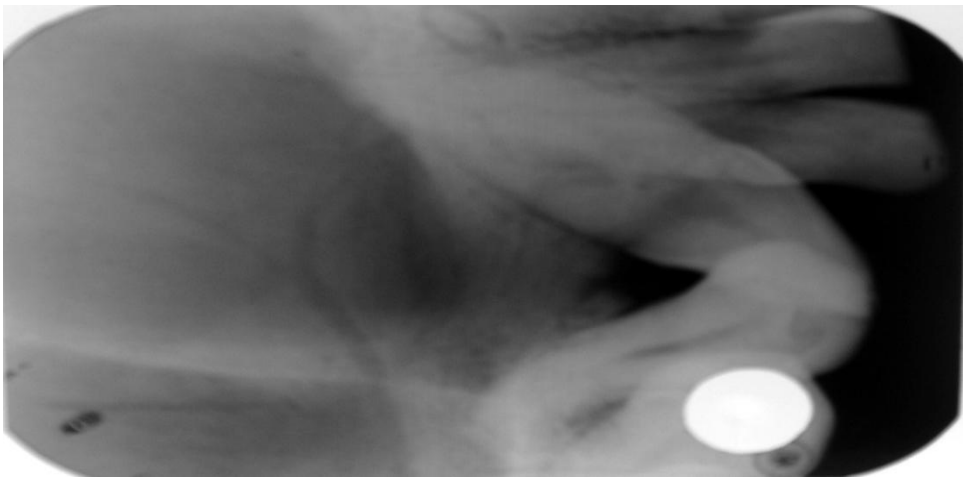

Fig. 4: Intraoral Periapical Radiograph showing well defined, homogenous radiolucency

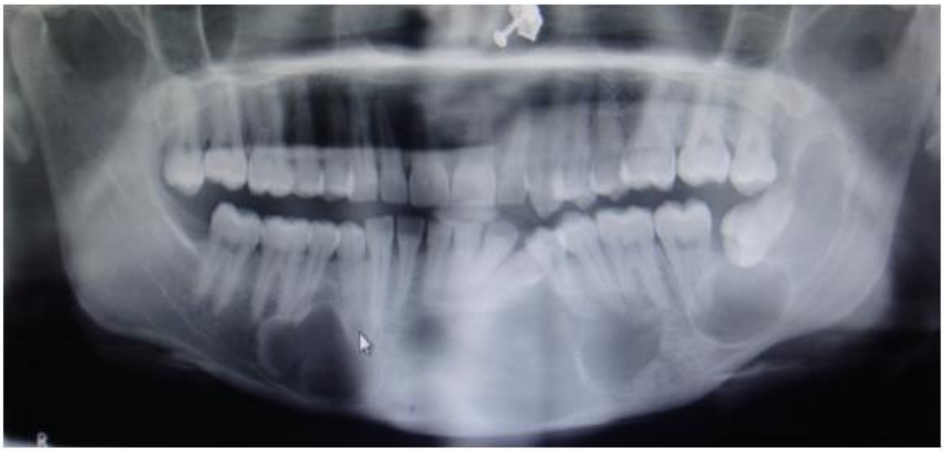

Fig. 5: OPG showing multiple radiolucencies in the mandible

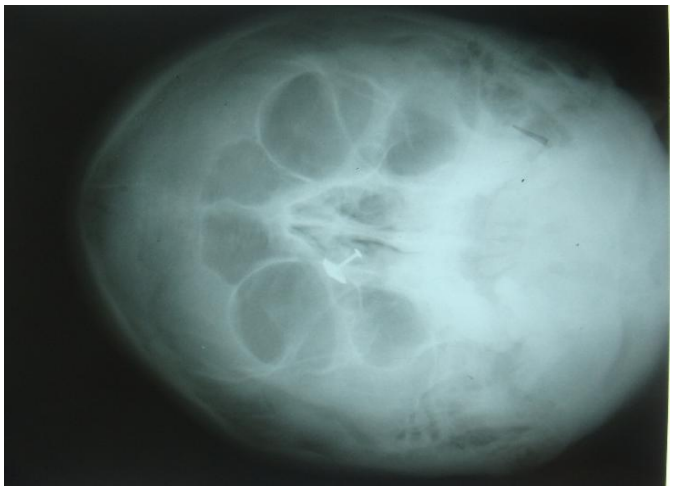

Fig. 6: Paranasal Sinus View showing absence of calcification of Falxcerebrii

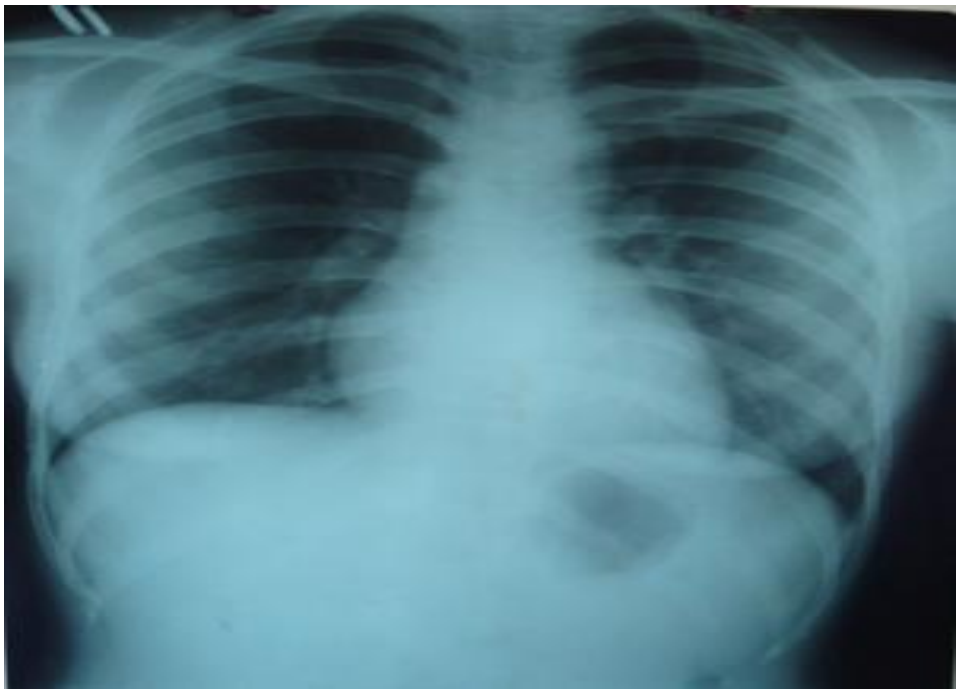

Fig. 7: No abnormality detected in Chest Radiograph

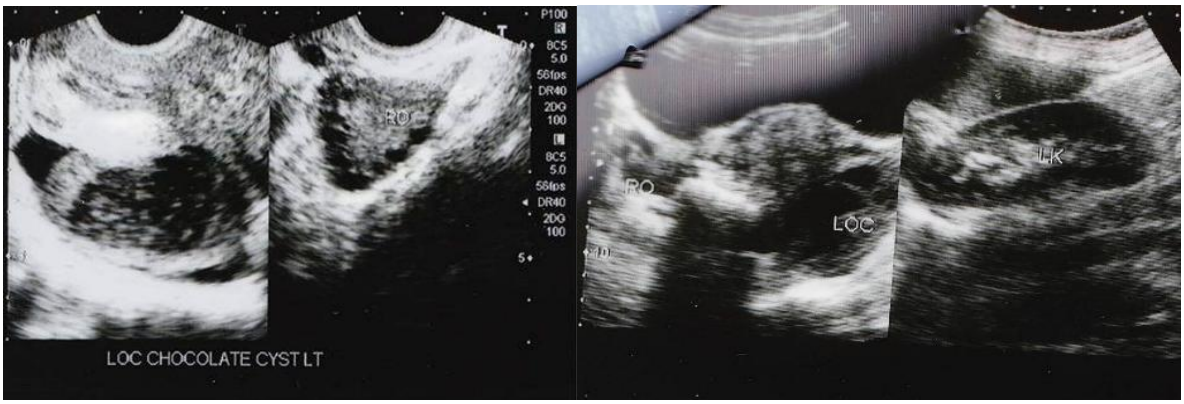

Fig. 8: Ultrasonography showing left ovarian cyst of about 36mmx26mms filled with organized coarse echoes

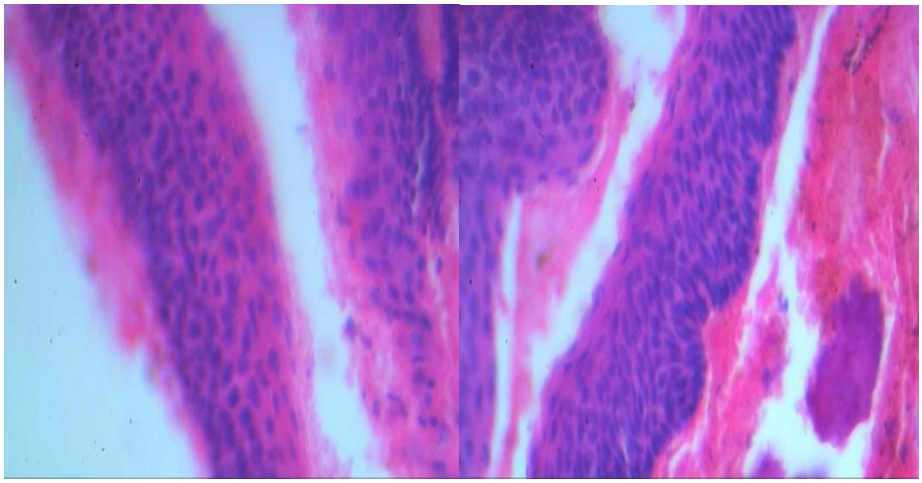

Fig. 9: Histopathological picture under10x resolution & 40x resolution

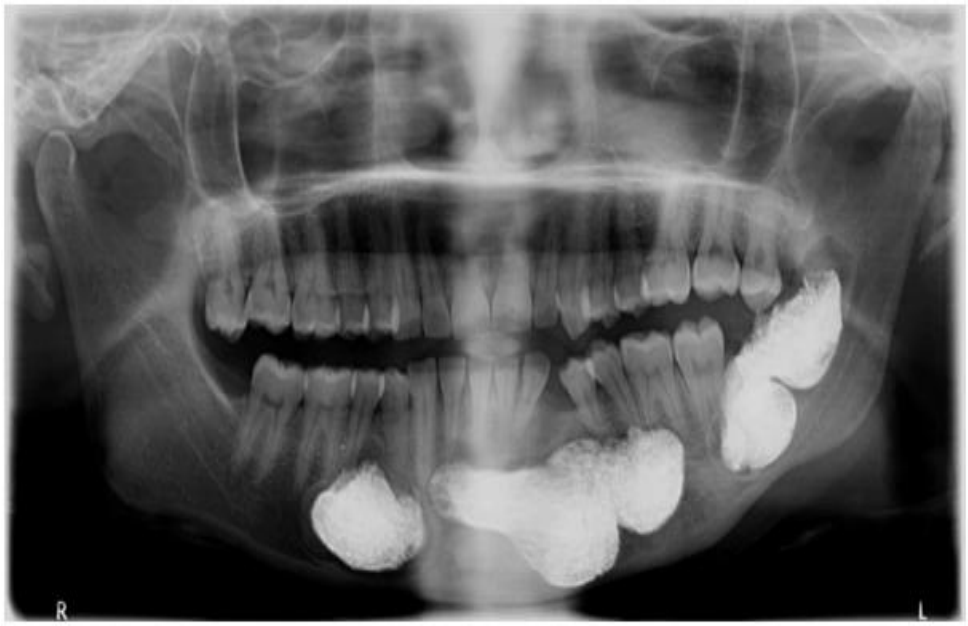

Fig. 10: Post-operative OPG after placement of Iodoform dressing

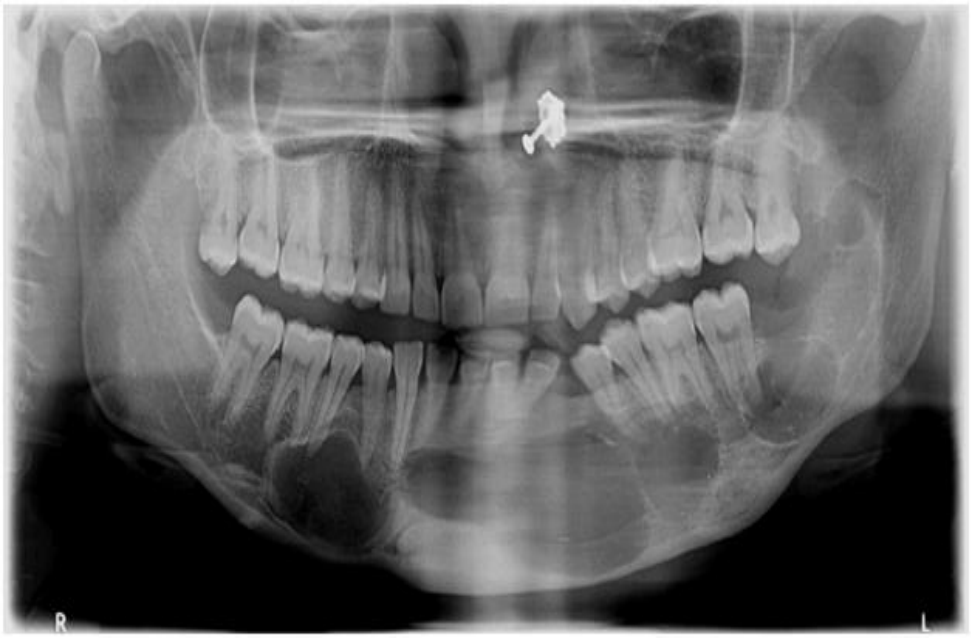

Fig. 11: Post-operative OPG in follow-up visit (1month)

DISCUSSION

KCOTs are among the common form of cystic lesions affecting the maxillofacial region.[1]The history goes back to mid 1950's when a group of oral pathologists in Europe gave the term odontogenickeratocyst, to denote a cyst with specific histological features and clinical behavior.[2,3]WHO(2008), based upon the clinical and histopathological features reclassified it as a tumor and included them in group of benign odontogenic tumors with mature, fibrous stroma as they are aggressive lesions clinically, having pathognomichistopathological features and lost PTCH gene functioning. According to WHO, these are defined as a benign, unicystic or multicystic intra-osseous tumors of odontogenic origin, with a characteristic lining of parakeratinised stratified squamous epithelium & potential for aggressive, infiltrative behaviour.[4] They are thought to originate from the dental lamina and its remnants.[5]

KCOTs account for 11% of cysts of jaws.There is slight predilection for males (1.3:1). The peak incidence is in 2nd and 3rddecades (60%).Mandible ramus-third molar area is most commonly involvedregion of jaw (80%); followed by anterior mandible.More oftenly they reach a large size before they are diagnosed as they tend to extend in the medullary cavity same as in our case.[4]

Multiple KCOTs commonly occur in association with Gorlin-Goltzsyndrome.[5]Diagnostic criteria of **Gorlin-Goltzsyndrome** was given by Evans *et al* and modified by Komones*et al* in 1997. It can be established by the presence of two major or onemajor andtwo minor criterias.[6]

MAJOR CRITERIA

1. More than 2 basal cell carcinomas (BCC)or 1 BCC underthe age of 20 years
2. Histologically proven odontogenickeratocyst of thejaws
3. 3 or more cutaneous palmar or plantar pits
4. Bifid fused or markedly splayed ribs
5. First degree relative with NBCCS

MINOR CRITERIA

Any one of the following features:

1. Proven macrocephaly after adjustment for height
2. One of several orofacial congenital malformation, cleft lipor palate, frontal bossing, coarse face, moderate tosevere hypertelorism
3. After skeletal abnormalities, marked pectus deformity,marked syndactyly of the digits
4. Radiological abnormalities, bridging of the sellaturcica vertebral anomalies such as hemivertebra, fusion or elongation of the vertebral bodies modeling defects of hands and feet
5. Ovarian fibroma
6. Medullobalstoma

In our case histologically proven multiple keratocysts of jaws (major criteria) and ovarian cyst (minor criteria) was present. Hence criterias for Gorlin-Goltz syndrome were not fulfilled. So this is the rare case of non – syndromic multiple KCOTs.

Most frequent clinical manifestations at first admission are reported to be swelling, pain or both[7,8] same as in our case.

Radiographically, OKCs present as a well defined radiolucent lesions with smooth, usually corticated margins and may be either unilocular or multilocular. Unerupted tooth is involved in 25% to 40% of cases.[9]Our case complied with these findings, with all the detected radiolucencies being unilocular, having

well corticated margins. There was radiolucency in the left ramus region associated with unerupted 38.

Histologically, KCOTs reveal the presence of 8-10 cells thickparakeratinized or orthokeratinized stratified squamous epithelium, with a prominent basal layer of columnar or cuboidal cells, more mitotic figures in suprabasal layers[10, 11] same as in our case.

Treatment modalities include enucleation which may be combined with cryotherapy or Carnoy's solution, and marginal or radical resection.KCOTs patients are treated to "prevent" recurrence at the margins of the initial lesion[5,12]. In our case, enucleation was done combined with Cornoy's solution.

In conclusion, due to their aggressive nature and higher rate of recurrence of KCOTs in NBCCs syndrome and the probable development of other associated problems in future, it is the responsibilityof the dentist to correctly diagnose the lesion, ruling out the presence of this syndrome andprovide a careful long follow-up to avoid recurrences.

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