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MULTIPLE ORTHOKERATINIZED ODONTOGENIC CYST : A RARE CASE REPORT

Oral Pathology

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ABSTRACT

Orthokeratinized odontogenic cyst is a rare developmental cyst of the jaw region.OOC is now considered as an individual entity due to its different biologic behaviour. The purpose of this article is to present a rare case of OOC arising from three different areas ie. Right mandible , Left mandible and Left Maxillary Sinus in the same 15 year old female patient. The most intriguing part of this case is that all the three lesions displayed different clinical and radiological presentations.

KEYWORDS

Orthokeratinized odontogenic cyst , KCOT , Mandible , maxilla , Maxillary sinus

INTRODUCTION:

The orthokeratinized odontogenic cyst is a developmental odontogenic cyst . It arises from the cell rests of Serre. It was first described by Schultz in 1927 as an orthokeratinized variant of the previously called odontogenic keratocyst ,today known as the keratocystic odontogenic tumour. It is in 1981 that Wright defines this as an independent entity.[1] The OOC occurs predominantly in males during the third and fourth decades , with a mean age of 33.5 years.[2] The lesion is located mainly in the posterior mandible , chiefly in the molar region.[2] Clinically they presents as a swelling along with pain and can reach a large size with cortical expansion.[2] Radiographically the cyst appears as a well-circumscribed ,unilocular or multilocular radiolucency that occasionally can beassociated with an unerupted tooth or with the root causing its resorption. [3] The histopathological analysis confirms the diagnosis which shows a cystic cavity lining composed of a thin and uniform stratified squamous epithelium with a thick granular layer and orthokeratin.[4] Surgical enucleation is the treatment of choice for the OOC . The low recurrence rate has been described between 0 and 2% of the cases.[2]

Case report:

A 15 year old female patient reported to the Department of Dentistry IGIMS , Patna with the chief complaint of swelling in the right lower back region of the jaw since 6 months. The medical and dental history were non-contributory. On extra-oral examination , a diffuse swelling was seen on the right side of the mandible extending anteroposteriorly from the right corner of the mouth upto 2 cm away from the ear lobe, superoinferiorly at the level of the corner of the mouth upto 2 cm beyond the inferior border of the mandible. (Fig-1)On palpation , swelling was non-tender , firm in consistency ,with well-defined margins. On intra-oral examination , a small swelling with buccal cortical plate expansion was seen in relation to 45 ,46 measuring approximately 2*3 cms. (Fig-2) 48 was missing clinically. Based on the history and clinical examination , a provisional diagnosis of Dentigerous cyst was suggested. Patient was then sent for further radiological investigations. The orthopantomography revealed a unilocular radiolucency on the right mandible with well defined corticated margins measuring approximately 3.5*4.5 cms extending from root apex of 45 upto mesial root of 47. Distal Root displacement of 46 with resorption was also seen. The entity is displacing the inferior alveolar nerve canal inferiorly leading to its indistinct appearance near the entity. (Fig-3)

Incidental findings-

1) Massive multilocular radiolucent lesion on the left mandible extending from 37 upto approaching ascending ramus and sigmoid notch partially involving the coronoid area measuring approximately 8*4.5cms. Tooth like radiopaque structure is seen within the entity at the angle of the mandible. The margins are thin and well definedcausing displacement of inferior alveolar nerve canal inferiorly. (Fig-3)
2) Small areas of opacification are seen in right and left maxillary sinus.

In order to determine the extension of the lesion , patient was then advised for CT Scan of face. The CT findings showed involvement of entire left maxillary sinus .(Fig-4) A fine needle aspiration cytology was performed on the right side of the mandibular swelling and a pearly -whitish creamy liquid material was obtained. The cytology of the material confirmed the presence of benign squamous epithelial cells with predominance of neutrophils. The complete enucleation of the lesion from three different sites ie. right mandible, left mandible andleft maxillary sinus were performed and the tissue retrieved were submitted for histopathological examination in three separate containers. Grossly the excised specimen revealed a cystic sac. The histopathology of the tissue from all the different sites revealed a cystic lining comprising of 4-7 layers of stratified squamous epithelium with prominent granular cell layer and orthokeratosis. (Fig-5) Basal cells were cuboidal with palisading and nuclear hyperchromatism.Few areas showed surface corrugation. (Fig-6) The epithelial connective tissue interface was flat. The cystic lumen was filled with dense flakes of orthokeratin. (Fig-7) The cystic wall was fibrous with mild to moderate inflammatory infiltrate predominantly neutrophils.Based on the above features, a confirmatory diagnosis of Orthokeratinized Odontogenic Cyst was rendered.

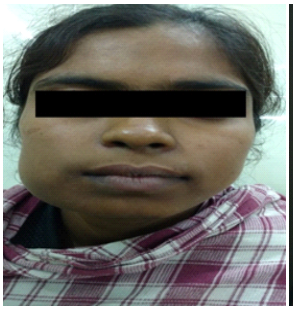

Fig 1

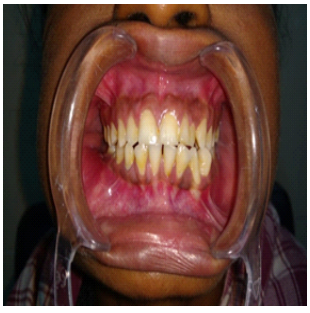

Fig 2

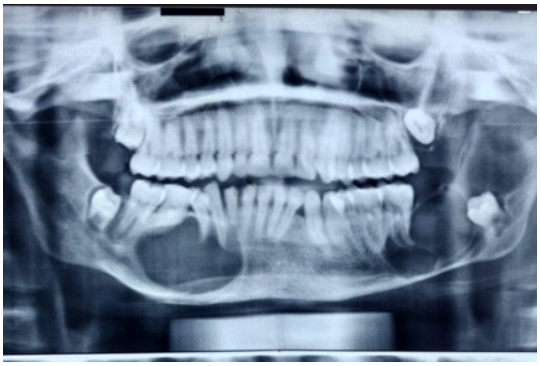

Fig -3

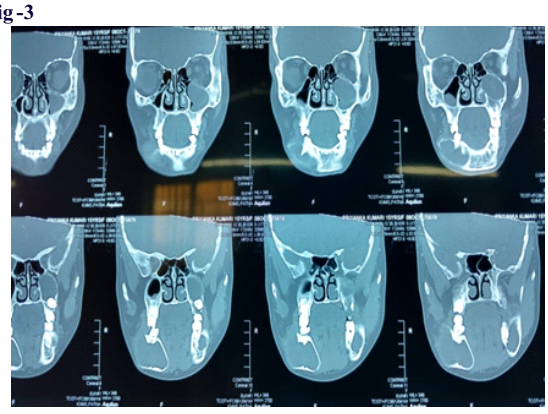

Fig - 4

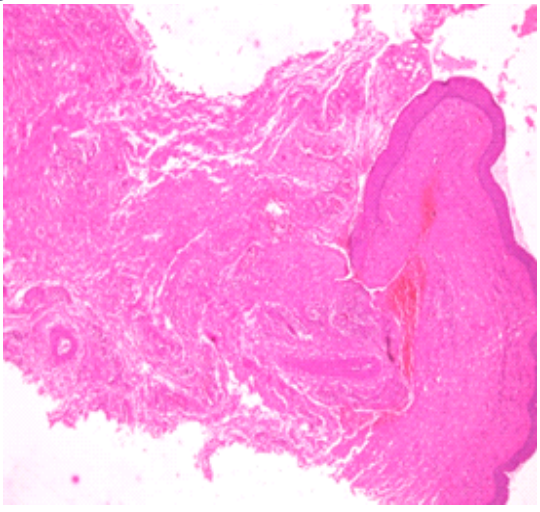

Fig -5

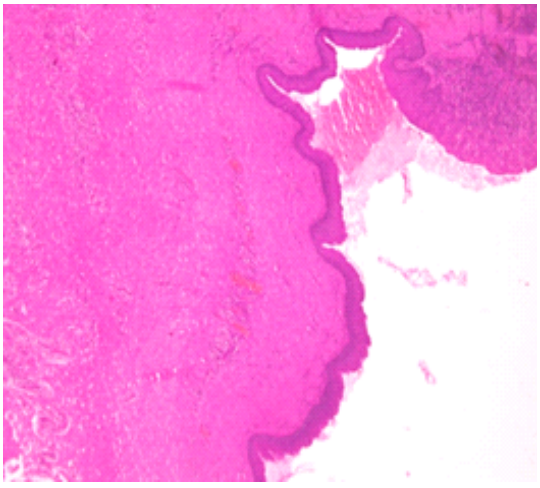

Fig -6

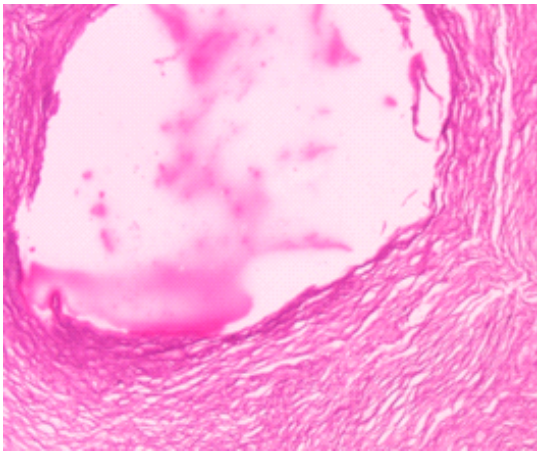

Fig - 7

DISCUSSION

The presence of dentition in the jaws and the development due to fusion of various embryonic process increases the incidence of cysts in the jaw bones than the other bones in the body. With rare exceptions, epithelium-lined cysts in the bone are seen only in the jaws. Other than a few cysts that may result from the inclusion of epithelium along embryonic lines of fusion, most jaw cysts are lined by epithelium that is derived from odontogenic epithelium.[12]The 2005 edition reclassified the parakeratotic type as KCOT and stated "Cystic jaw lesions that are lined by orthokeratinizing epithelium do not form a part of spectrum of a KCOT".The less aggressive clinical behaviour and recurrence pattern of the orthokeratinized variant ultimately warranted the designation of the orthokeratinized variant as a separate entity , "Orthokeratinized Odontogenic Cyst).[8]

OOC represents 7-17% of all keratinizing jaw cysts.[9] Vuhahula et al , in their study stated that reduced enamel epithelium involved in formation of dentigerous cyst that had completed its tooth-forming function had the capability to keratinize under appropriate stimuli , thus forming a true dentigerous cyst with keratinization.[8] The size can vary from less than 1 cm to large lesions greater than 7 cm in diameter.[5] Some rare forms of OOC have been described in literature like peripheral orthokeratinized odontogenic cyst, orthokeratinized odontogenic cyst histopathologically associated with calcifying odontogenic cyst or ameloblastoma or heterotrophic cartilage or squamous cell carcinoma.[7]

Both the OOC and KCOT show similar findings clinically regarding age , sex and site of occurrence but the OOC are generally solitary asymptomatic lesions whereas KCOT associated with Nevoid basal cell carcinoma syndrome can exhibits multiple lesions.[9] Radiographically they more frequently present as unilocular radiolucencies (87%) in comparison with KCOTs (69.4 to 73.3%). OOC is more often associated with an impacted tooth (60.8%) than the OKC (7-48%).[5] Wysocki and Sapp showed that there are distinct ultrastructural differences between OKC and OOC. The surface morphology of OOC is more uniform and entirely covered with keratin squames. As cells mature , there is increase in tonofilaments and granular cell layer shows compact layer of degenerated cells that contain large amounts of keratohyaline granules.5In relation to the immunohistochemical pattern, the OOC does not show activity of the epithelial membrane antigen (EMA) and of the carcinoembrionary antigen (CEA)[11]. Recent immunohistochemical studies that compared the OOCs with KCOTs have shown distinct differences in the expression of Ki-67 proliferative index, p53, p63 and bcl-2. Reduced expression of all these markers in OOC reflect that they have a different cell differentiation and exhibit a lower cellular activity than the keratocystic odontogenic tumor. [9]The reactivity to cytokeratins has showed differences, as OOC stains to cytokeratins which would suggest a normal differentiation of the epidermis whilst the KCOT reacts to cytokeratins 4, 13, 17, and 19, demonstrating that these are different entities.[10] The expression of cell surface glycoprotein marker gp38 is negative in OOC whereas depicts neoplastic potential of KCOT ,similarly cell migration and tumor invasion marker podoplanin has low expression in OOC whereas depicts neoplastic potential of KCOT.[7]

CONCLUSION-

OOC should always be considered in the differential diagnosis of all

the radiolucent lesions of the jaw. It should be differentiated from other cysts as they exhibit different behaviour pattern which in turn can affect the treatment plan and prognosis. Histopathology remains the gold standard for diagnosing these cysts.

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