Case report

Title: ODONTOGENIC KERATOCYST MIMICKING A DENTIGEROUS CYST: A CASE REPORT.

Abstract: Odontogenic keratocyst is a developmental, non – inflammatory chronic cyst that may be unilocular or multi locular. OKC is a cyst of tooth origin with an aggressive clinical behavior including a high recurrence rate. It is known for its tendency to invade the adjacent tissues including bone. Clinically OKC is manifested by an asymptomatic growth. Radiographically, it appears as a well-defined unilocular or multilocular osteolytic lesion. The diagnostic approach is based on a combined analysis of the medical history, the clinical appearance and the radiographic appearance. A case of odontogenic keratocyst involving the ramus of the mandible is presented in this article emphasizing on the characteristics and various features of OKC.

Key words: *odontogenic keratocyst, keratocyst odontogenic tumor.*

Introduction:

Odontogenic keratocyst is a distinctive form of developmental odontogenic cyst that deserves a special consideration because of its specific histopathologic features & clinical behavior. The term odontogenic keratocyst was first given by philipsen in 1956[(1)]. OKC's most commonly occur in the second & third decades of life and show a slight predilection for males (males to female ratio 1.3:1). The recent WHO classification categorizes OKC as a developmental non-inflammatory odontogenic cyst that arises from the cell rests of dental lamina [(2)]. Majority of the OKCS occur in the mandible, most commonly in the angle of mandible and ramus.

The clinical & radiographic features of OKC are indefinite; while some may be associated with pain, swelling or drainage, Most of them are asymptomatic. 82% of the OKC'S occur in the tooth bearing areas and 27% of the cases show an association with atleast one impacted tooth (mostly mandibular third molar)[(3)]. Here we report a case of odontogenic keratocyst associated with an unerupted third molar.

CASE REPORT:

A 50 year old female patient reported to the department with a chief complaint of swelling and pus discharge in the left lower back tooth region since 4 months. History revealed that the swelling was initially small in size but it gradually increased to the present size. There was also history of difficulty in swallowing and foul taste. Past medical history revealed that the patient was blind since 15 years.

On extra oral examination slight facial asymmetry was present due to the presence of diffuse swelling over the right mandibular angle, approximately 4 x 3.5 cms in its anteroposterior dimension & 3.5 x 3 cms supero inferiorly (Fig 1) Swelling was non tender and firm in consistency. Skin over the swelling was normal.

On intra oral examination there was obliteration of buccal vestibule irt 36, 37 teeth (Fig 2). Overlying surface of the swelling was of same colour as that of surrounding mucosa. On palpation it was firm in consistency, non tender, on application of pressure a white creamy exudate oozed out of the area distal to 37. On aspiration a cream coloured hazy fluid was obtained.

With the above clinical findings a provisional diagnosis of odontogenic keratocyst of the left mandible was given. Differential diagnosis of dentigerous cyst was included.

Panoramic radiograph showed a well defined radiolucency on the left ramus of the mandible which was approximately 4x2x1 cm in size, oval In shape extending anteriorly from distal aspect of 38 to posteriorly 0.5 cms below the condyle; radiopaque scalloping margins with uniform radiolucency, expansion of inferior border of the mandible at the left angle region, inferior alveolar nerve canal is pushed inferiorly (Fig 3). Computed tomography revealed a cystic lesion with scalloped and well corticated borders (Fig 4a & 4b). Based on the radiographic findings a provisional diagnosis of odontogenic keratocyst was given with a differential diagnosis of unicystic ameloblastoma, dentigerous cyst, and odontogenic myxoma.

An incisional biopsy was done under local anesthesia. Histopathologic specimen revealed stratified squamous cell epithelium with parakeratosis and prominent basal layer without rete ridges (Fig

5a). Sub-epithelium showed fibro collagenous stroma with islands of squamous epithelium (daughter cysts) (Fig 5b)The overall features were suggestive of odontogenic keratocyst and the final diagnosis of odontogenic keratocyst irt left ramus area was made.

Discussion:

The term odontogenic keratocyst was first coined by philipsen in1956 [1]and its characteristics were first described by pindborg and Hansen in 1963 [4] since then the cystic nature of OKC has been disputed and gained a special attention since last two decades. Some investigators classify classify the OKC as a benign tumor but the aggressive nature of OKC has put the dilemma that it is cyst or neoplasm. In 1967 TOLLER suggested that OKC is to be named as benign neoplasm [5].OKC was reclassified and renamed as keratocystic odontogenic tumor (KCOT) in the WHO classification of head and neck tumors in 2005 [6]. Redesignation of the OKC as the KCOT is based on the well known aggressive behavior of the lesion, histology and new information regarding its genetics.

The patched gene PCTH, a tumor suppressor gene involved in both nevoid basal cell carcinoma syndrome and sporadic KCOTS, commonly occur in chromosome 9q22.3-q31.36-40 [7]. PCTH forms a receptor complex with the oncogene SMO (smoothened) for the SHH (sonic hedge hog) ligand. The growth-signal-transduction is inhibited by PCTH binding to SMO. This inhibition is released by SHH binding to PTCH. When the normal functioning of PCTH is lost, the proliferation stimulating effects of SMO are permitted to predominate [8]. However the 2017 classification reverted back to the well accepted terminology of OKC because there was substantial evidence that PTCH gene mutation can also occur in non neoplastic lesions like dentigerous cysts [9]. Moreover, many researchers challenged the neoplastic process of OKC as marsupilisation causes resolution of the cyst [10]. And keratocystic odontogenic tumor was carried forward as a synonym of OKC.

Most of the OKC'S arise from the cell rests of dental lamina or from the basal cells of oral epithelium and are thus are primordial-origin odontogenic keratocysts. The remaining 40% arise from the reduced enamel epithelium of the dental follicle and are thus dentigerous- odontogenic keratocysts as in our case. The clinical identification is crucial in determining the treatment as the recurrences are more

frequently seen after treatment of primordial type of OKC. OKCs may occur at any age but the highest incidence is generally in the second and third decades of life. There is a slight male predilection .Approximately 20-45 % of OKC'S are associated with unerupted tooth .And about 70% of the cases involve mandible especially the molar, angle and ramus regions in particular. Here in our case all the three distinctive characters can be appreciated.

The characteristic radiographic feature of OKC'S include unilocular or multilocular radiolucency with distinctly corticated, often scalloped, borders.

OKC'S tend to grow in antero posterior direction within the medullary cavity of the bone and may cause an obvious bone expansion [11.] A hazy radiolucent lumen can be seen on a conventional radiograph which is suggestive of a dense proteinacious material such as keratin. Resorption of the roots of the erupted tooth is rare. Displacement of teeth adjacent to the cyst occurs more frequently than resorption.

The histopathologic features of OKC are more specific. Diagnostic features include a uniform cyst lining, hyperchromatic and palisaded basal cells, wavy parakeratin production and a flat interference between the epithelium and connective tissue wall. One of the most peculiar characacteristic features of OKC is the appearance of satellite cysts/ island of odontogenic epithelium [12] which was seen in our case .the high recurrence rate can be attributed to the satellite cysts that are retained during enucleation; thin, fragile cystic walls can be left intact.

The treatment options for OKC'S range from simple conservative treatment to more aggressive treatment that may include enucleation, with or without curettage, or marsupialization. Though conservative treatment preserves the anatomic structures they have a high risk for recurrence. The aggressive treatment includes peripheral ostectomy, chemical curettage with carnoys solution, or *enbloc* resection.recurrence rate of OKC'S range from 32 to 62% on an average. This is due to the persistence of satellite cells in the cyst wall. In such cases cornoys solution acts as a fixative agent , and can be used in odontogenic cysts and tumors show a reduction in recurrence rate to 9%.And a long term follow up of 5-10 years is generally recommended.

Conclusion:

Odontogenic keratocyst is an unique entity in among odontogenic cysts that has feature similar to those of dentigerous cysts , which is a common provisional diagnosis for OKC, the best way for proper diagnosis is accurate clinical, radiographic and trans-surgical observations with a biopsy specimen examination; this approach will help determine the most effective treatmentthereby preventing recurrence.

Figure 1: Extra oral clinical photograph showing swelling in the left lower half of the face.

Figure 2: Intraoral clinical picture showing a diffuse swelling irt 36, 37

Figure3: Panoramic radiograph showed a well defined radiolucency irt left molar ramus area.

Figure 4a and 4b: Computed tomography revealed expansile corticated and scalloped cystic lesion.

Figure5a: Histopathologic section shows stratified squamous epithelium showing parakeratosis

With prominent basal layer without rete ridges.

Figure 5b : Histopathologic section shows sub-epithelium islands of squamous epithelium (daughter cysts)

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Figure 1:

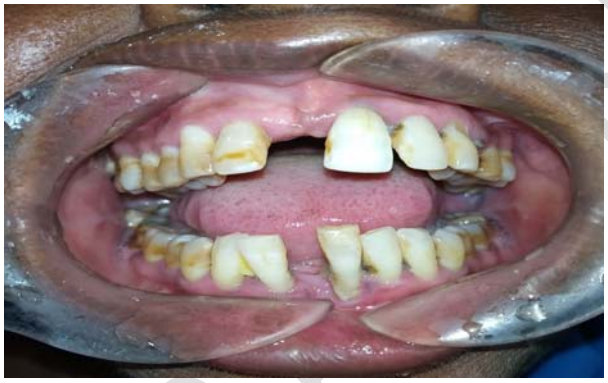

Figure 2:

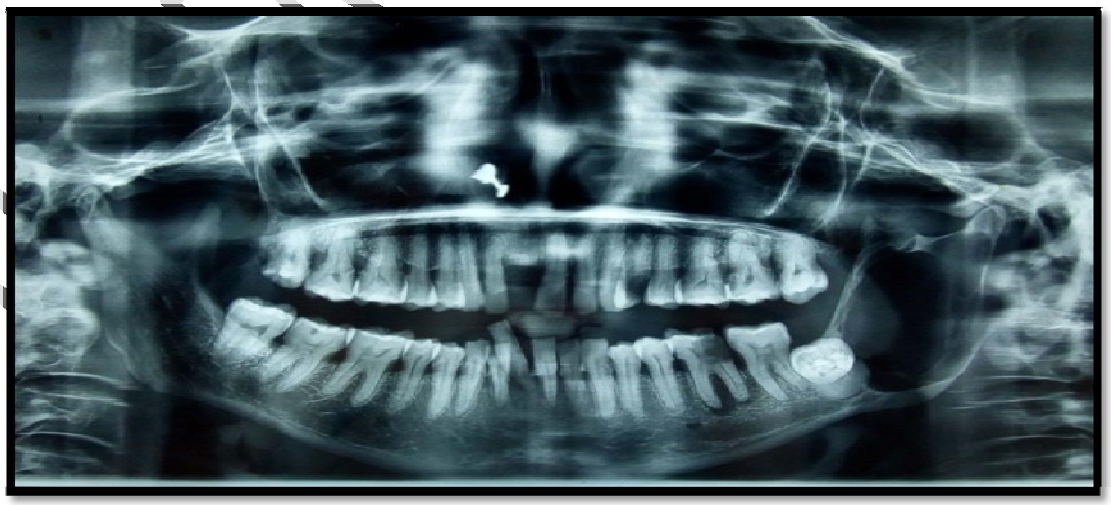

Figure 3

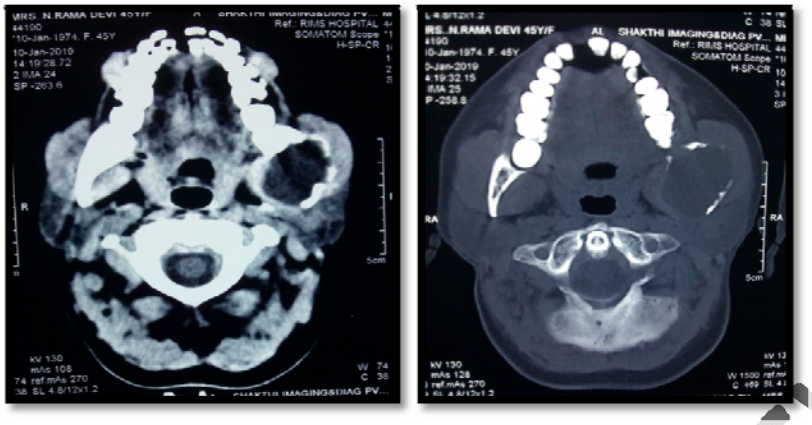

Figure 4a and 4b:

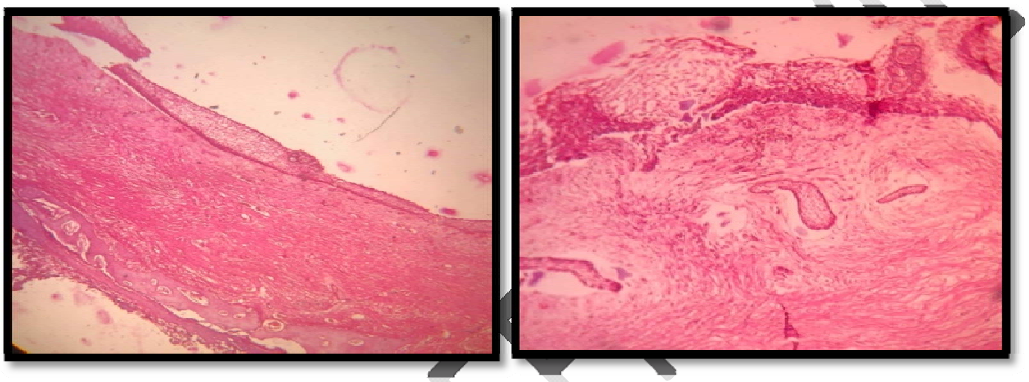

Figure 5a and Figure 5b.: