Case report

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

Abstract: Odontogenic keratocyst (OKC) 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 and 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 history, clinical appearance, and 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 clinical behavior and histopathologic features. The term odontogenic keratocyst was first given by philipsen in 1956[(1).] OKC's most commonly occur in the second and 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- ascending ramus region.

The clinical & radiographic features of OKC are indefinite; while some may be associated with pain, swelling or drainage, most of them are asymptomatic. OKC'S commonly occur in the tooth bearing areas(82%) and some of the cases show an association with atleast one impacted tooth (27%-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 presented to our department on account of swelling and pus discharge in the left lower back tooth region of 4 months duration. History revealed that the swelling was initially small in size but gradually increased to the present size with associated history of difficulty in swallowing and foul taste. Past medical history revealed that the patient had been blind since 15 years.

On extra oral examination slight facial asymmetry was present due to the presence of a solitary 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 in relation to 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 and non tender, on application of pressure a white creamy exudate oozed out of the area distal to 37. Aspiration of the swelling yielded a cream coloured hazy fluid (Figure 3).

A provisional diagnosis of odontogenic keratocyst of the left mandible was made with a differential diagnosis of dentigerous cyst.

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 to 0.5 cm 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 4). Computed tomography revealed a cystic lesion with scalloped and well corticated borders (Fig 5a & 5b). Radiographic findings were suggestive of odontogenic keratocyst with a differential diagnosis of unicystic ameloblastoma, and odontogenic myxoma.

An incisional biopsy was done under local anesthesia. Histology revealed stratified squamous cell epithelium with parakeratosis and prominent basal layer without rete ridges. Sub-epithelium showed fibro collagenous stroma with islands of squamous epithelium (daughter cysts) (Fig6and 6b). A definitive diagnosis of odontogenic keratocyst involving the left angle-ramus area was made.

Discussion:

The history of odontogenic keratocyst dates back to 1826 when Mickuliz first described it as a part of familial condition affecting the jaws. In 1926 it was referred to as "cholesteatoma."- meaning a cystic or "open" mass of keratin squames with a living "matrix". Later in 1945 Robinson mentioned this cyst as primordial cyst as they arose from remnants of the dental lamina or the enamel organs before enamel formation has had taken place. However it was not until 1956 the cyst has got the name odontogenic keratocyst by philipsen [1]. Since then the terminology has been a matter of dispute due to the distinct clinical, radiological, histopathologic features of OKC and gained a special attention since last two decades. Some investigators classify OKC as a benign tumor but the aggressive nature of OKC has put the dilemma as to whether it is a cyst or neoplasm. In 1967 TOLLER suggested that OKC is to be named as benign neoplasm [(4)].Shear used the term "keratocystoma" citing the aggressive nature of the odontogenic keratocyst and finally labeled it as a benign cystic neoplasm OKC was reclassified and renamed as keratocystic odontogenic tumor (KCOT) in the WHO classification of head and neck tumors in 2005[(5)]. 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 [(6)]. 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 [(7)]. But there was substantial evidence that PTCH gene mutation can also occur in non neoplastic lesions like dentigerous cysts[(8).] Moreover, many researchers challenged the

neoplastic process of OKC as marsupialization causes resolution of the cyst [9]. Hence the 2017 WHO classification reverted back to the well accepted terminology of odontogenic keratocyst-OKC[10].

Most of the OKC'S arise from the cell rests of dental lamina or from the basal cells of oral epithelium and are thus primordial-origin odontogenic keratocysts[11]. 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 the mandible especially the molar, angle and ramus region. Here in our case all the three distinctive characteristics can be appreciated.

OKC'S tend to grow in antero posterior direction within the medullary cavity of the bone and may cause an obvious bone expansion [12] A hazy radiolucent lumen can be seen on a conventional radiograph which is suggestive of a dense proteinacious material such as keratin. Resorption of roots of the erupted teeth is rare with displacement of teeth adjacent to the cyst occuring more frequently than resorption.

And more attention is given to the histological features which help to ensue a definitive diagnosis. 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 characteristic features of OKC is the appearance of satellite cysts/ island of odontogenic epithelium [13] which was seen in our case. High recurrence rate can be attributed to the satellite cysts that are retained during enucleation; thin, fragile cystic walls can be left intact.

Unicystic ameloblastoma and orthokeratinized odontogenic cyst (OOC) though present with the same clinical and radiographic features, Histologically Unicystic ameloblastoma has ameloblastic epithelial lining which is pathognomic of this cyst. The suprabasilar areas often loosen up giving the appearance like a stellate reticulum[14]. OKC shares similar characteristics with Orthokeratinized odontogenic cyst with

respect to age of occurrence and site, yet these two lesions differ in their biological activity[15]. OKC can be differentiated from Orthokeratinized odontogenic cyst on various features like older age group, more antero-posterior extension without expansion, characteristic histopathological features different from Orthokeratinized odontogenic cyst, parakeratinized layer, high recurrence rate, association with basal cell nervous syndrome.

The treatment options for OKC'S range from simple conservative treatment like enucleation, (with or without curettage), or marsupialization. Marsupialization is a technique relying on incomplete removal of the cyst lining. Opening a window into the cyst forms an invagination of the oral cavity or the maxillary antrum.It relies on the principle that decompression halts expansion of the cyst and appositional growth of bone occurs, and the former cyst lumen becomes smaller with time. Many modification of the procedure have been taken place over the time like usage of decompression tubes, marsupialization catheter. One such method has been demonstrated by COSTA F.W.G et al where he used a segment of polyethylene suction tube, prepared according to the radiographic size of the lesion. Using a disposable needle, a hole is drilled near the extremity, large enough to allow the passage of a 0.8-mm orthodontic stainless steel wire. With the aid of a needle holder, one end of the wire is shaped into a loop and the other end is inserted through the hole in the tube, pulled back, and twisted. The tooth crown is etched with acid and the loop is attached to the dental surface with composite resin. Advantage of this technique over other methods was that it provides greater stability and minimizes the need for additional surgical interventions, when compared to traditional methods where decompression devices were attached to the surrounding structures with sutures. This provides insufficient stability in case of surgical wound dehiscence. Poor adjustment increases the likelihood of device-related complications.[16]

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. Carnoy's solution is composed of 3 ml of chloroform, 6 ml of absolute ethanol, 1 ml of glacial acetic acid, and 1 g of ferric chloride, is often used as a complementary treatment of lesions with high recurrence rates such as odontogenic keratocyst. The action of this solution is given by chemical cauterization, promoting a superficial necrosis of about 1.5mm of depth after 5 minutes of

bone cavities exposure. In a study conducted by Albuquerque et al on surgical treatment with or without carnoy's solution in aggressive tumors of odontogenic origin the authors found a beneficial effect of the Camoy's solution in reducing the recurrence rate in several cases of jaw aggressive odontogenic tumors. This emphasizes the importance of carnoy's solution when used in conjunction with conservative procedures like enucleation.[17]

Surgical intervention included resection of the diseased mandible – hemimandibulectomy (figure 7, 8) followed by reconstruction with titanium condylar plates (figure 9) by giving 2 cm surgical margin proximally.

Conclusion:

Odontogenic keratocyst is a unique entity among odontogenic cysts, due to its varied clinical, radiological and histopathologic features. This case report re-iterates the importance of histology in differentiating a odontogenic keratocyst from other odontogenic cysts of the mandible. Hence the correlation of histopathologic findings with clinical and radiographic features is of paramount importance to achieve a correct definitive diagnosis, as most of these lesions have a prognostically different biologic behaviours and the final diagnosis helps to proceed with the appropriate treatment procedure.

Consent Disclaimer:

As per international standard or university standard, patient's consent has been collected and preserved by the authors.

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 in relation to 36, 37

Figure3: showing dense creamy exudate on aspiration

Figure 4: Panoramic radiograph showed a well defined radiolucency in relation to left molar ramus area.

Figure 5a and 5b: Computed tomography revealed expansile corticated and scalloped cystic lesion.

Figure6a: Histopathologic section shows stratified squamous epithelium showing parakeratosis

With prominent basal layer without rete ridges.

Figure 6b: Histopathologic section shows sub-epithelium islands of squamous epithelium (daughter cysts)

Figure 7: showing the exposed cyst involving the angle- ramus up to the condyle of the mandible.

Figure 8: showing the resected mandible along with the condyle

Figure 9: showing reconstruction of the defect with titanium reconstruction plates.

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Figure 1:

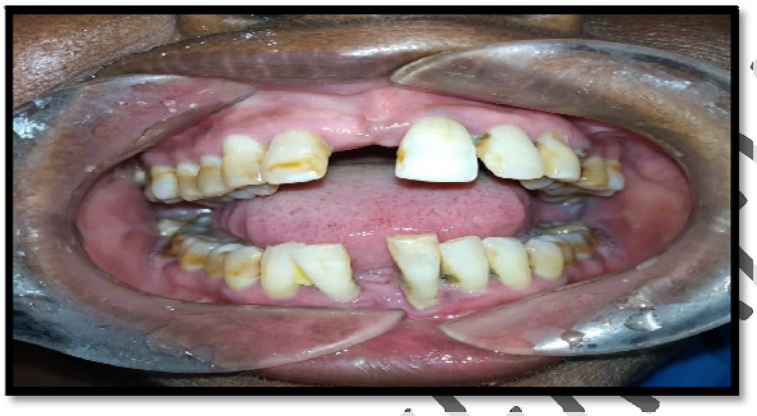
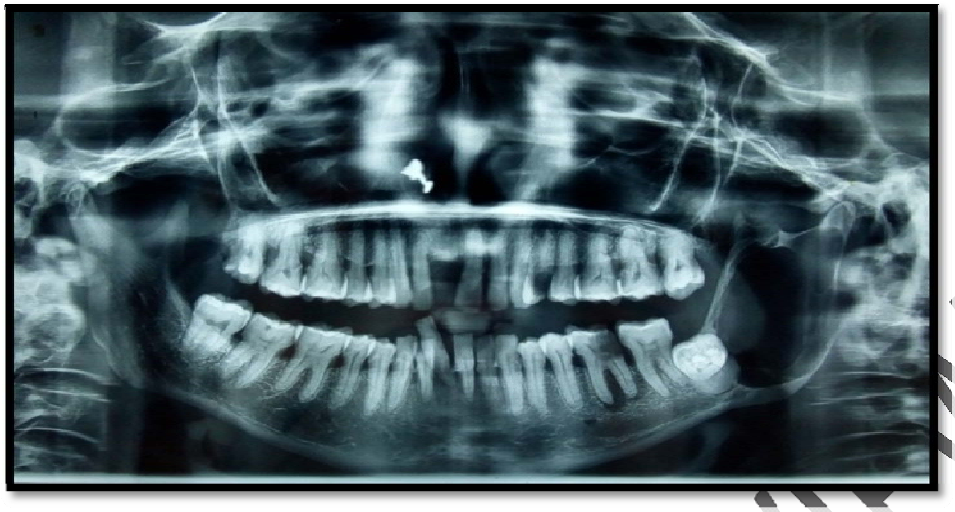


Figure 2:

Figure 3:



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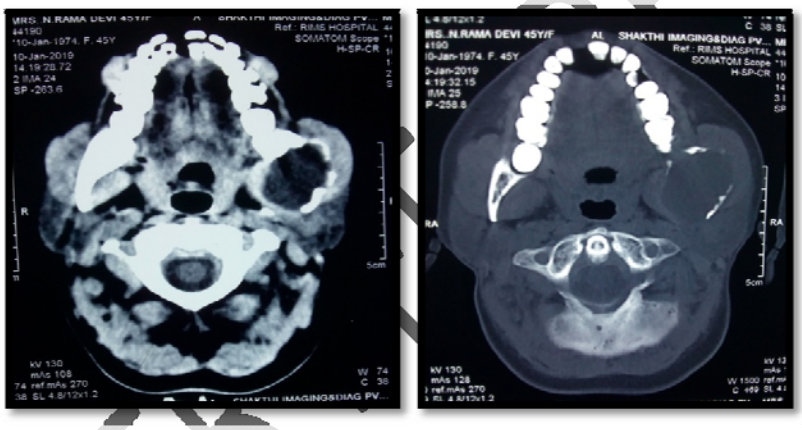

Figure 5a and 5b:

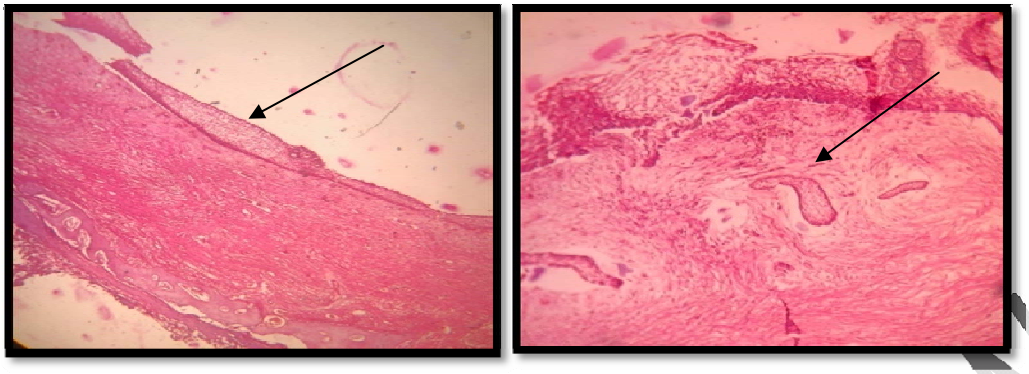

Figure 6a and Figure 6b.:

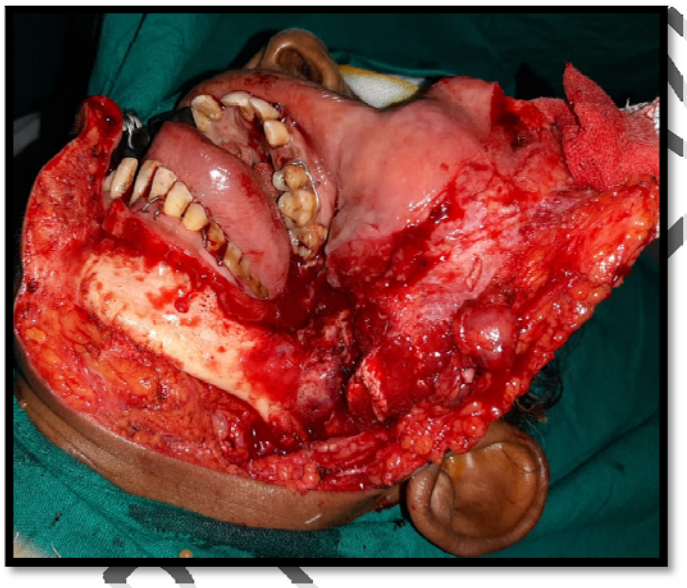

Figure 7

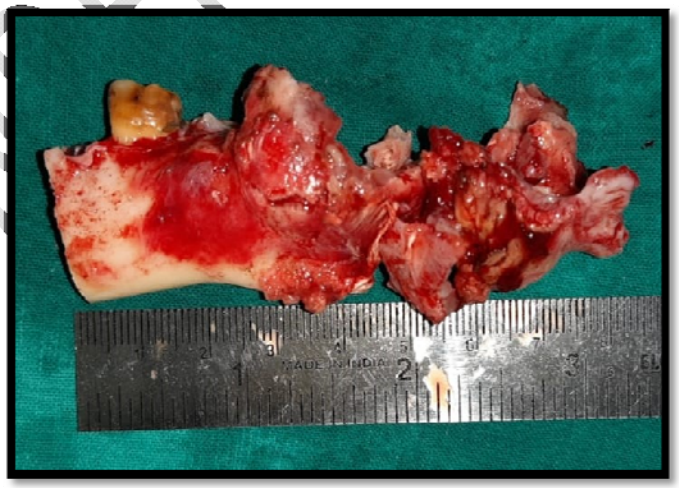

Figure 8

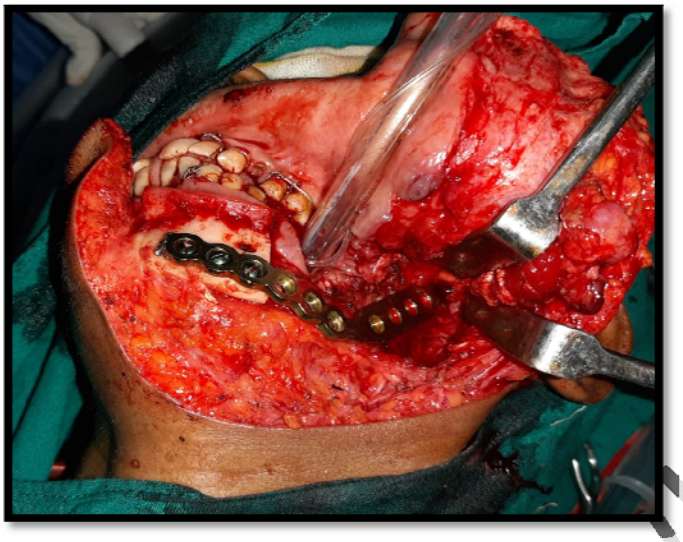

Figure 9