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Case Report

Odontogenic keratocyst- Mimicking residual cyst in maxilla

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ABSTRACT

OKC was classified as cystic lesion by WHO in 1971 & 1991, based on aggressive nature, growth pattern, clinical, histological and immunohistochemical nature in 2005 they again classified it as benign lesion, however in 2017 WHO head and neck pathology reclassified it as cystic lesion. It more commonly occurs in posterior mandible and rarely occurs in maxilla, in this case occurrence of OKC in maxillary posterior region is very rare with distinctive expansion and lifting of maxillary sinus floor without perforating in edentulous area makes it more difficult to detect and justify from residual cyst. Here a 65 years old patient came with chief complaint of pus discharge from upper left posterior region since 7 months, having a small opening in edentulous ridge, which provisional diagnosis was given as residual cyst later after excision of lesion and histopathological analysis it was given as OKC.

1. Introduction

Philipsen in 1956, first described the term 'odontogenic keratocyst' (OKC), as developmental cyst of jaw.[1] Histologic criteria to detect OKC was given by Pindborg and Hansen.[2] Intially it was given as odontogenic keratocyst, but later on WHO classified it as Keratocystic odontogenic tumour because of its aggressive nature and growth pattern and mutational changes,[3] it is non inflammatory and arises from cell rest of dental lamina. Now in 2017 again WHO reclassified it as OKC.[4] They are usually seen in mandibular posterior regions and rarely in maxilla, about 70-80% occur in the lower jaw, about 90% occur posterior to the canines and 50% in the ascending ramus of the mandible, very rarely in the maxilla.[5]

WHO gave new classification in which they changed the designation to OKC from keratocystic odontogenic tumour (KCOT) and described it as a benign uni or multilocular, intraosseous tumour of to odontogenic origin, with aggressive and invading behaviour".[3] Usually there are no significant clinical symptoms, although mild swelling and pain can be seen likely due to secondary infections.[5] As they are often confused with other cysts and tumours because of vague appearances and similar features it is important to diagnosis. Similar in our case OKC occurred in edentulous area with history of long term presence but it cannot be proved clinically or radiological as OKC but on histopathology it was proved. So complete diagnosis and proper treatment is necessary to reduce recurrences.

2. Case Report

A 65 years old female patient reported to the department with chief complaint of pus discharge from upper left posterior region of jaw since 7 months. Initially patient was asymptomatic, later on sudden onset of swelling with pus discharge in that area. Swelling was smaller in size and gradually increased to present size. Patient gives history of extraction 2 years back after which she had experienced mild pain during chewing for which she has taken medication from local dentist, patient is diabetic and hypertensive since 2 years and under medication.

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2.1. On examination

(Figure 1 a and b) Mild facial fullness and tenderness over left side of cheek. Intraoral (Figure 2) single diffuse swelling present in maxillary left posterior region which is of 3 x 3cms in size obliterating buccal vestibule, Anteroposteriorly from 24 to 28 region, Supero-Inferior from alveolar ridge to depth of vestibule, expanding the ridge overlying surface appears normal and surrounding too. Intraoral examination showed a tiny draining sinus is seen in crest of edentulous ridge in 27 region.

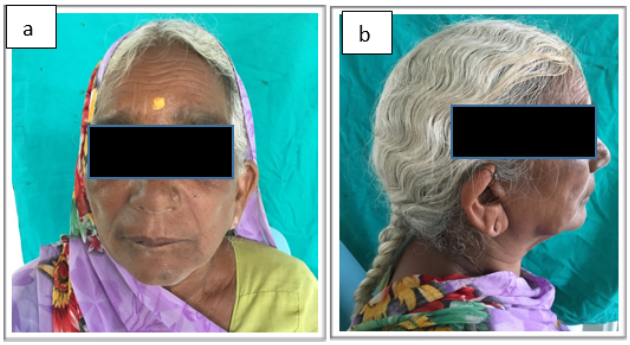

Fig. 1: (a),(b) front and lateral profile showing mild facial fullness left cheek

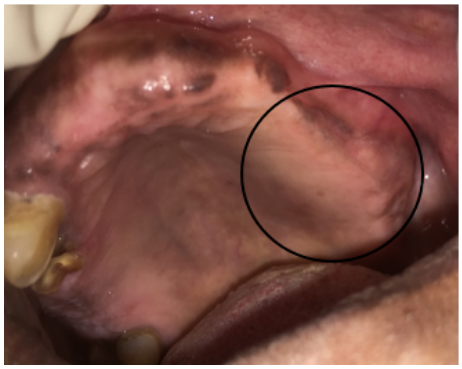

Fig. 2: Showing diffuse swelling in left buccal vestibule

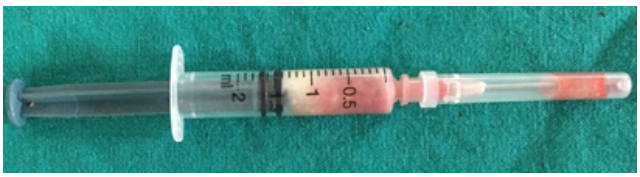

Fig. 3: Aspiration showing cheesy fluid with blood.

On palpation swelling is firm in consistency, mildly tender, compressible, egg shell crackling and expansion felt. Clinically present 13, root pieces-14,15 and mesioproximal caries with 17. Single left submandibular lymph node is palpable which is soft, mobile, mildly tender. So provisional diagnosis is given as residual cyst of left maxillary region.

Patient is advised for IOPA, OPG, CBCT, routine investigation, aspiration and biopsy. Aspiration- (Figure 3) Milky white cheesy liquid with RBCs aspirate on investigation revealed keratocyst. IOPA (Figure 5) and OPG reveals unilocular radiolucency surrounded by corticated border which is interrupted at base and opening in soft tissue which is hazy and appears like involving maxillary sinus.

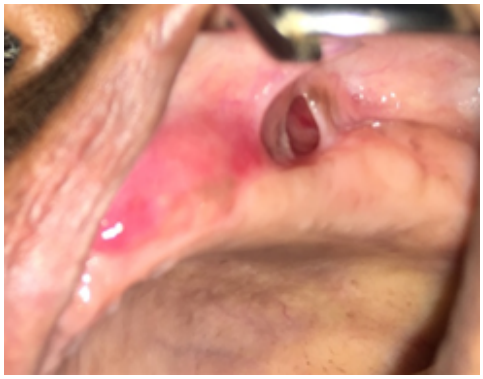

Fig. 4: Showing after excision of lesion, opening in alveolar ridge

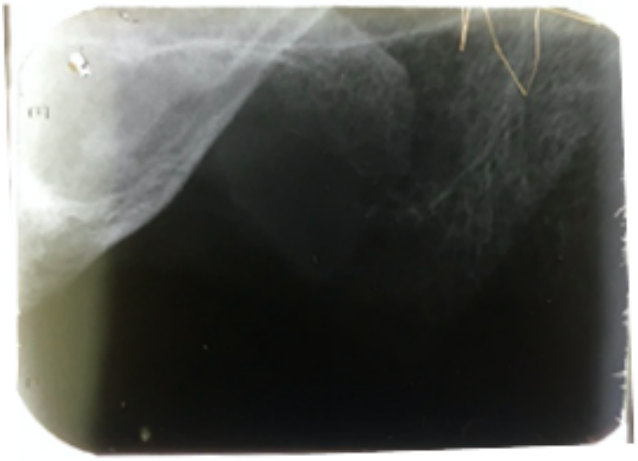

Fig. 5: Iopa showing interrupted floor of maxillary sinus

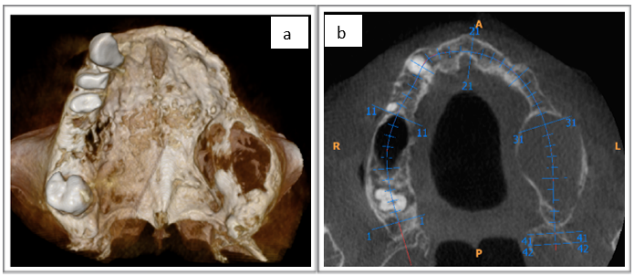

Fig. 6: a and b reveals large lesion involving left maxillary sinus with anteroposterior and buccopalatal expansion

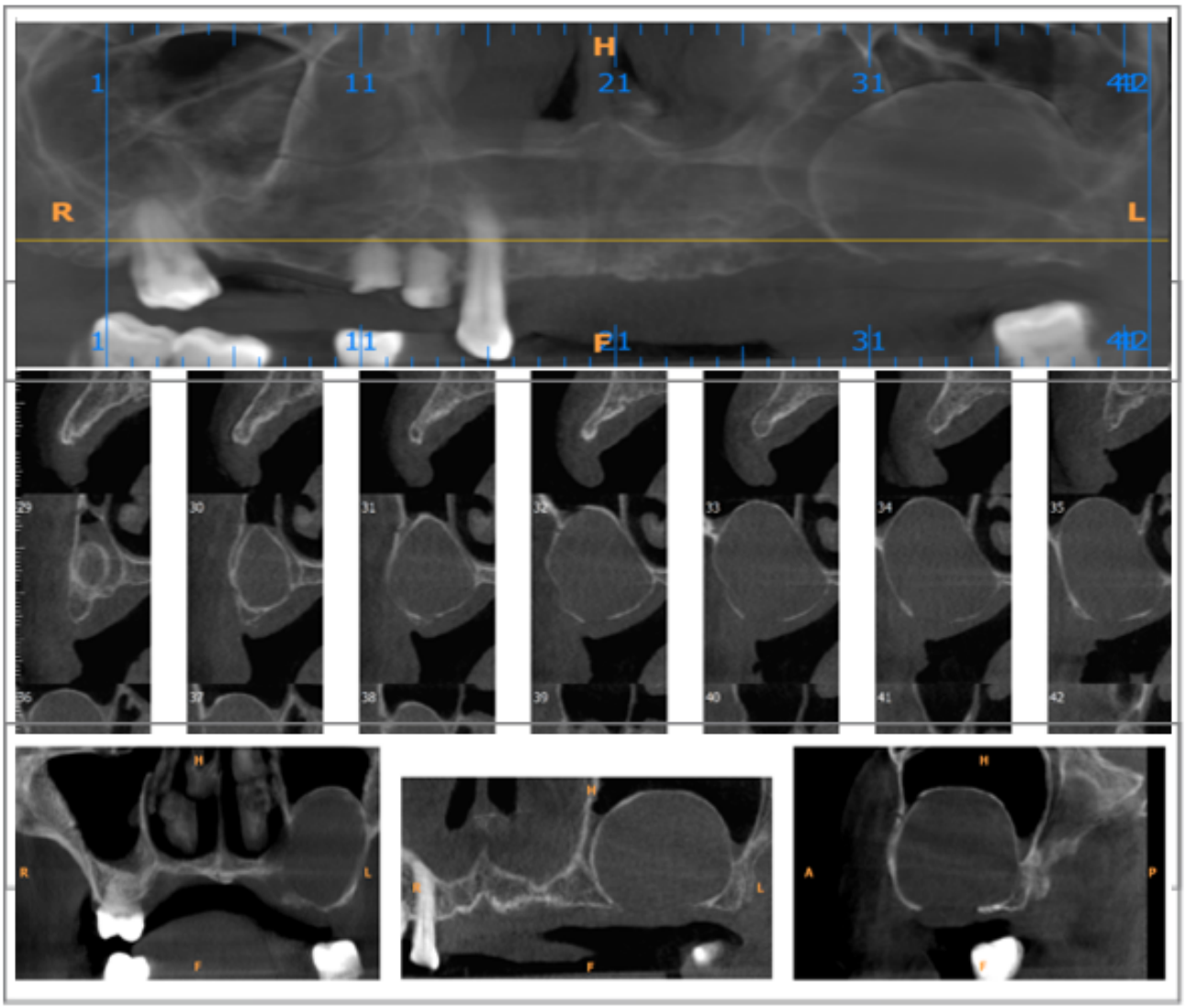

Fig. 7: CBCT sections showing unilocular hyperdense area in maxillary sinus encroaching and obliterating it except superior aspect.

CBCT(Figures 6 and 7) reveals well defined, corticated, unilocular hyperdense areas in left maxillary sinus measuring around 26.7mm in vertical height and 33.3mm in mediolateral dimensions. Antero-posterior extensions are approximately 31.4mm in length. Missing 17, 16, 12, 11, 21, 22, 23, 24, 25, 26 and 27 is observed. Hyperdense area obliterating left maxillary sinus from all aspects except the superior aspect of left maxillary sinus. Floor of left maxillary sinus is displaced superiorly due to the extension of the lesion. Coronal Sections depicts the well defined border of lesion involving and encroaching the lower part of left maxillary sinus. Buccal and palatal bone in left posterior region in relation with edentulous area of 26, 27 and 28 is lost. The alveolar crest in the edentulous region is destructed in relation with 26 and 27, given as residual cyst in relation to left maxilla.

Curettage & enucleation of the lesion followed by application of Carnoy's solution used for reducing the recurrence rate under general anaesthesia (Figure 4). Excised tissue specimen was submitted for the histopathological examination. On histopathological examination it was confirmed as OKC. Patient was kept on follow up for 6 months and no recurrent lesion seen.

3. Discussion

In the earlier literature, the OKC was described as a cholesteatoma (Hauer, 1926; Kostecka, 1929). Forssell (1980) concluded that the first account of this lesion was given by Mikulicz in 1876, described it as a dermoid cyst.[1]

The epithelium of the keratocyst is strongly believed to arise from either the dental lamina or the residue of the dental lamina (cell rests of Serre).[4]

OKC are relatively common developmental odontogenic cysts and account for 10–12% of all jaw cysts.[6] The term Odontogenic Keratocyst (OKC) was first coined by Philisen in 1956 (Eryilmaz et al., 2009) and its characterstic features was first described by Pindborg and Hansen in 1963.[1] Odontogenic keratocyst (OKC) is so named because keratin is produced by the cystic lining. It is a Parakeratin lined cyst-like lesion within bone. OKC is the one of the rare and distinctive developmental odontogenic cyst which from the dental lamina, containing clear fluid and a cheesy material resembling keratin debris.[7]

Its journey of nomenclature is as follows:

1. Dental cyst (John Hunter 1774),
2. Dermoid Cyst (Mikulicz 1876)
3. Primordial cyst (Robinson 1945)
4. Keratocystoma (Shear) Odontogenic keratocyst (Philipsen 1956 & Pindborg and Hansen 1963)
5. Benign neoplasm (Toller 1967)
6. Odontogenic keratocyst (WHO 1971)
7. True benign cystic epithelial neoplasm (Ahlfors 1984)
8. Odontogenic keratocyst (WHO 1992)
9. Keratocystic odontogenic tumor (Benign neoplasm) (WHO 2005)
10. Odontogenic keratocyst (WHO 2017)[1,8]

The keratocystic odontogenic tumor is more common in males than females with ratio of 1.6:1 and occurs over a wide age range. White more common, it is typically diagnosed during the second to fourth decade. In our case, the patient was in his sixth decade.[9] Peripheral OKCs have female predominance with male: female ratio of 2.2:1.[7]

KCOTs (Figures 8 and 9) mostly occur in the body of the mandible, most commonly in the molar region and vertical ramus.[9] However in present case, there was involvement of maxillary posterior region which is edentulous and lifting of the maxillary sinus seen. In our case shows a rare site of occurrence in a female. Literature suggests that less than 1% of the KCOT cases occur in the maxilla with maxillary antrum involvement. In the maxilla, the canine region is the most common location for KCOTs.[10]

The clinical features include pain or without pain, soft-tissue swelling, displacement of teeth, expansion of bone in anteroposterior in medullary space of bone, drainage, and various neurological manifestations, such as, paresthesia of the lip or teeth in case of lower jaw involvement. The maxillary KCOT tends to be secondarily infected with greater frequency than the mandibular ones, due to its vicinity to the maxillary sinus.[10] In this case too, the lesion was secondarily infected, with the presence of mild pain, a sinus opening and pus discharge. Expansion of buccal cortex in 30% of maxillary and 50% of mandibular regions.[3] Syndromes associated with multiple OKC are Nevoid Basal cell carcinoma syndrome (NBCCS), Gorlin goltz syndrome, Marfans syndrome, Ehlers danlos syndrome, Noonans syndrome, Orofacial digital syndrome, Simpson golabi-behmel syndrome.[10,11]

Radiographically most KCOTs are unilocular or multilocular, presenting a well-defined peripheral rim (corticated), with scalloped borders, root resorption, displacement of teeth with involvement of impacted tooth. Buccal bone expansion more compared to lingual with or without perforation. Small lesion have minimal expansion due to its growth pattern in antero posterior direction when it increases it involved buccal and lingual bones too.[3,7,10] In the present case, the radiographic evaluation revealed unilocular radiolucency in left maxillary posterior region which is aggressive osteolytic lesion, involving edentulous area causing perforations in the buccal and palatal cortical plates in the 24, 25, 26, 27 region, lesion encroaching and lifting maxillary sinus floor without perforating it and maintaining its border.

Radiological Types of keratocyst

1. Replacement type: Cyst which forms in the place of normal teeth.
2. Envelopmental type: Cyst which envelopes adjacent impacted tooth.
3. Extraneous type: Cyst which occur in ascending ramus away from the teeth.
4. Collateral type: Cyst which occurs adjacent to the root of teeth which are indistinguishable radiologically from lateral periodontal cyst.[8]

KCOT: In 1967, Toller suggested that the OKC should be considered as benign tumour because of its clinical nature.[1,4] In 1984, Ahlfors et. al[12] suggested that "if we consider OKC as benign neoplasm, the question of modified treatment would be raised." Since, many published reports in these years have influenced WHO to again classify the lesion as a tumour.

1. Behaviour: KCOT is localised catastrophic and high chances of recurrence.
2. Histopathology: Studies such as that by Ahlfors and others 10 show the basal layer of the KCOT budding into connective tissue. In addition, WHO notes that mitotic figures are frequently found in the suprabasal layers.[2]
3. Genetics: PTCH ("patched"), a tumour suppressor gene involved in both NBCCS and sporadic KCOTs, occurs on chromosome 9q22.3-q31.[13–16] Normally,PTCH forms a receptor complex with the oncogene SMO ("smoothened") for the SHH ("sonic hedgehog") ligand. PTCH binding to SMO inhibits growth-signal transduction. SHH binding to PTCH releases this inhibition.[14] If normal functioning of PTCH is lost, the proliferation-stimulating effects of SMO are permitted to predominate.
4. Recent studies revealed that the etiology of NBCCS and KCOTs has a "2-hit mechanism," with allelic

MALIGNANT TUMOURS

Odontogenic carcinomas

Metastasizing (malignant) ameloblastoma

Ameloblastic carcinoma - primary tumour

Ameloblastic carcinoma - secondary tumour (dedifferentiated)

Intraossoeus

Ameloblastic carcinoma - secondary type (dedifferentiated)

Peripheral

Primary intraosseous squamous cell carcinoma - solid type

Primary intraosseous squamous cell carcinoma derived from keratocystic odontogenic tumour

Primary intraosseous squamous cell carcinoma derived from odontogenic cysts

Clear cell odontogenic carcinoma

Dhost cell odontogenic carcinoma

Odontogenic sarcomas

Ameloblastoma fibrosarcoma

Ameloblastoma fibrodentino - fribro-odontosarcoma

BENIGN TUMOURS

Odontogenic epithelium with mature fibrous stroma without odontogenic ectomesenchyme

Ameloblastoma

Ameloblastoma, unicystic type

Ameloblastoma, extraneous/ peripheral type

Metastatisizing amelobalstoma

Squammous odotnogenic tumour

Calcifying epithelial odontogenic tumour

Adenomatoid odontogenic tumour

Odontogenic keratocyst

Odotnogenic epithelium with odontogenic ectomesenchyme, with or without hard tissue formation

Ameloblastomic fibroma

Ameloblastic fibrodentinoma

Ameloblastic fibro-odontoma

Odontoma

- odontoma, complex type

- odontoma, composite type

Odontoameloblastoma

Calcifying cystic odontogenic tumour

Dentinogenic ghost cell tumour

Mesenchyme and/ odontogenic ectomesenchyme with or without odontogenic epithelium

Odontogenic fibroma

Odontogenic myxoma/ myxofibroma

Cementoblastoma

Bone-related lesions

Ossifying fibroma

Fibrous dysplasia

osseous dysplasia

Central giant cell granuloma

Cherubism

Aneurysmal bone cyst

Simple bone cyst

Other tumours-

Melanotic neuroectodermal tumour of infancy

Fig. 8: Showing WHO 2005 classification in which OKC is included in benign lesion as KCOT and 2017 reclassified as cystic lesion as OKC.

Odontogenic carcinomas

Ameloblastic carcinoma

Primary intraosseous carcinoma

Sclerosing odontogenic carcinoma

Clear cell odontogenic carcinoma

Ghost cell odontogenic carcinoma

Odontogenic carcinosacrcoma

Odontogenic sarcomas

Benign epithelial odontogenic tumours

Ameloblastoma

Ameloblastoma, unicystic type

Ameloblastoma, extraneous/ peripheral type

Metastatisizing amelobalstoma

Squammous odontogenic tumour

Calcifying epithelial odontogenic tumour

Adenomatoid odontogenic tumour

Benign mixed epithelial & mesenchyme odontogenic tumours

Ameloblastic fibroma

Primordial odontogenic tumour

Odontoma

Dentinogenic ghost cell tumour

Benign mesenchymal odontogenic tumour

Odontogenic fibroma

Odontogenic myxoma/ myxofibroma

Cementoblastoma

Cemento-ossifying fibroma

Giant cell lesions and simple bone cyst

Central giant cell granuloma

Peripheral giant cell granuloma

Cherubism

Aneurysmal bone cyst

Simple bone cyst

Hematolymphoid tumour

Solitary plasmacytoma of bone

Odontogenic cysts of inflammatory origins

Radicular cysts

Inflammatory collateral cysts

Odontogenic and non odontogenic developmental cysts

Dentigerous cyst

Odontogenic keratocyst

Lateral periodontal cyst and botyroid odontogenic cysts

Gingival cyst

Glandular odontogenic cyst

Calcifying odontogenic cyst

Orthokeratinized odontogenic cyst

Nasopalatine duct cyst

Malignant maxillofacial bone and cartilage tumours

Chondrosarcoma

Mesenchymal chondrosarcoma

Osteosarcoma

Benign maxillofacial bone and cartilage tumours

Chondroma

Osteoma

Melanotic neuroectodermal tumour of infancy

Chnondroblastoma

Chondromyxoid fibroma

Osteoid osteoma

Osteoblastoma

Desmoplastic Fibroma

Fibro-osseous and osteochondromatous lesions

Ossifying fibroma

Familial Gigantiform cementoma

Fibrous dysplasia

Cementosseous dysplasia

Ostoechondroma

Fig. 9: Showing WHO 2005 classification in which OKC is included in benign lesion as KCOT and 2017 reclassified as cystic lesion as OKC.

loss at 9q22.42,43.[13] Tumour suppressor gene is inactivated in "2 hit mechanism". The first hit is a mutation in one allele, which, although it can be dominantly inherited, has no phenotypic effect. The second hit refers to loss of the other allele and is known as "loss of heterozygosity" (LOH). In KCOTs, this leads to the dysregulation of the oncoproteins cyclin D1 and p53. Lench and others[15] indicate that LOH in the 9q22.3-q31 region has been reported for many epithelial tumours, including basal cell carcinomas, squamous cell carcinomas and transitional cell carcinomas; they note that LOH is, "by definition a feature of tumorigenic tissue".[17]

Numerous surgical modalities have been suggested for the treatment of KCOTs, including, enucleation with primary closure, enucleation with open packing, marsupilization, enucleation with use of Carnoy's solution or cryotherapy, with a marginal or radical section. Although KCOTs are stated to be the most aggressive and recurrent form of tumors, there are cases where KCOTs have been treated by enucleation. In the present case also it has been noted that there has been complete healing following the enucleation with carnoys's solution procedure. Nevertheless, a long-term follow-up is required to check recurrence.

1. Enucleation - 30%.
2. Enucleation + carnoy's solution - 9%..
3. Enucleation + peripheral ostectomy - 18%
4. Enucleation + carnoy' solution + peripheral ostectomy - 8%.
5. Enucleation + cryotherapy - 38%.
6. Marsupialization - 33%.
7. Marsupialization + cystectomy - 13%.
8. Resection - 0%.

Recurrence rates with different treatment modalities (Madras and Lapointe, 2008).[4,11]

Histological features - Pindborg, phillipsen and Henriksen (Pindborg et al., 1962)[18] suggested series of histological features for the diagnosis of OKC which includes:

1. Stratified squamous epithelium lining which is thin and having wavy appearance – 8-10 layers thick.
2. Lacks of rete ridges/pegs / epithelial extensions.
3. Well defined basal cell layer having cuboidal or columnar cells arranged in palisaded fashion described as "picket fence or tombstone appearance."
4. A thin spinous cell layer which often shows direct transition from basal cell layer (artifactual separation of epithelium from basement membrane) and spinous cell layer intracellular oedema.
5. Surface keratinisation which is corrugated and rippled and mostly parakeratosis (keratinized cells with nuclei.
6. Cystic wall composed of fibrous connective tissue which is thin and usually uninflamed.
7. Others findings are satellite cysts, daughter cysts (7-30%), solid epithelial proliferation, odontogenic rests basal layer budding may be seen. Fibrous connective tissue wall may get mineralised and may include cholesterol crystals and Rushton bodies.

The mutliple factors could be responsible for reoccurrence of okc are :

1. After incomplete removal of cystic lining
2. Very thin and friable nature of epithelial lining,
3. Higher level of cell proliferative activity in the epithelium
4. Budding in the basal layer of the epithelium
5. Bony perforation
6. Adherence to adjacent soft tissue
7. Supra-epithelial and Sub-epithelial split of the epithelial lining
8. Parakeratinization of the surface layer
9. Remnants of dental lamina epithelium not associated with original OKC and development of new OKC in the adjacent area.
10. Growth of new OKC from satellite cyst /daughter cyst/remnants/cell rest.[4,8]

Recurrence of OKC/KCOT: Recurrence rate was found to vary from 0% to about 62%, depending on the kind of treatment management and follow-up period.[8]

4. Conclusion

Although we have many diagnostic tools to research articles still OKCs are routinely seen but their diagnosis is as difficult as previously, because of its appearances in both multi and unilocular fashion with occurrence in wide age group to any site and due to varying pattern its difficult to precise it. Its difficult to give exact diagnosis and treat it, as high chances of occurrences is there. So it should be considerate in differential diagnosis of cystic lesion and tumours.

5. Source of Funding

None.

6. Conflict of Interest

There is no conflict of interest.

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