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Odontogenic Keratocyst Diagnosis and Management: A Case Report

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Abstract:-Odontogenic keratocyst (OKC) is a developmental, non-inflammatory, chronic cystic lesion of odontogenic origin which is considered aggressive due to its genetic abnormalities and biological behavior being consistent with neoplastic progression. OKC clinically appears as a swelling with or without pain, occurring in the jaw bones with greater predilection towards mandible. Radiographically appearing as unicystic or multicystic radiolucency with anteroposterior growth of the lesion in the medullary spaces. Histologically the presence of parakeratin and the 'picket fence' appearance of the basal cell layer is unique to this lesion. Various treatment modalities have been put forth but no specific treatment has been regarded as the best. The goal of this paper is to create awareness about the aggressive nature of the OKC and to throw a light on its management.

Keywords:- *OKC, Multicystic, Mandible, Dentigerous Cyst, Aggressive.*

I. INTRODUCTION

Odontogenic keratocyst was first described in the year 1956 by Philipsen. In terms of treatment options and recurrence rate in the world literature, this pathological entity has evoked much controversies and debates. This lesion was reclassified as Keratocystic odontogenic tumor [KCOT] in 2005 by the World Health Organization. [1] KCOT was placed under the cyst category and was renamed odontogenic keratocyst (OKC) in the year 2017 by WHO on the release of a new head and neck tumor classification. [2]

It is a developmental cyst of the jaw with an odontogenic origin (dental lamina and its remnants) that exhibits aggressive and infiltrative clinical behavior and recurrence caused by the mutation of Patched gene (PTCH gene). OKCs are described as unicystic or multicystic tumours within the bone which have a parakeratinized stratified squamous epithelium cystic lining. [3] There have been reports of well-established OKCs transforming into primary intraosseous carcinoma (PIOSCC) or an Ameloblastoma. [4]

Epidemiologically, OKC accounts for 7.8% of all cysts of the jaw and the incidence varies from 4-16.5% with peak incidence in the 2nd and 4th decade of life showing predilection to males (male: female ratio is 1.6:1). As OKC has high recurrence rate and aggressive in nature, it is very important to know the presentation and clinical feature of the lesion for early diagnosis and management. Here OKC in a 29 years old male patient with minimal extraoral and intraoral presentation but aggressive involvement of the angle and ramus is presented. [5] In this article we presented a case with management on OKC.

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II. CASE REPORT

A medically fit 29-year-old male patient reported to our dental OPD with a chief complaint of swelling on the right side of the face since 10 days. The swelling was gradually increasing in size and was not associated with fever. The patient also gave a history of pain in the lower right back tooth region which was associated with pus discharge a year ago. The pain was mild in intensity, dull aching, and continuous in nature, non-radiating, non-aggravating and relieved on intake of anti-inflammatory drugs.

Extraoral examination revealed slight facial asymmetry due to diffuse swelling on the right side of the face **[Figure 1].** The swelling extended from the ala tragus line to the lower border of the mandible, majorly from the outer canthal line to the posterior border of the ramus. A restricted mouth opening of 21 mm was seen along with mild paresthesia on the right side of the face **[Figure 2].**

The patient was advised a panoramic radiograph which revealed multilocular radiolucencies on the right mandibular ramus and angle region extending up to the coronoid process with the thinning of the lower border of the mandible near the mandibular angle region **[Figure 3].**

The Cone Beam Computed Tomography scan revealed multilocular radiolucency separated by sclerotic septa. Perforation was noted on the lingual cortical plate with an intact buccal cortical plate **[Figure 4].**

Based on the given history, thorough clinical examination followed by the radiographic findings a diagnosis of odontogenic keratocyst (OKC) was considered.

Biopsy was done and the specimen was sent for histopathological examination which revealed cystic lining containing a parakeratinized stratified squamous epithelium with a thickness varying from 5- 10 layers. The lumen comprising of keratin material. Underlying connective tissue is composed of collagen fibers arranged parallel to the epithelium. Cholesterol clefts and dense amounts of inflammatory cells were noted. Hemorrhagic areas were seen **[Figure 5].** And the diagnosis of odontogenic keratocyst was confirmed.

After which the patient underwent segmental resection of the right mandible followed by reconstruction using customized reconstruction plates. Post-op review of patient after 4 months shows no recurrence of the lesion.

III. DISCUSSION

WHO termed the lesion as Keratocystic odontogenic tumor (KCOT) due to the neoplastic nature exhibited by the same. In 2017, OKC was reclassified back into the cystic category according to the new WHO classification of head and neck pathology. [1] It was named Odontogenic keratocyst (OKC) due to the keratin being produced by the cystic lining. It is an intraosseous lesion with a parakeratinized cystic lining. It is a rare and distinctive developmental odontogenic cyst that originates from the dental lamina remnants in the maxilla and mandible before the completion of odontogenesis. The lesion contains cheesy material and clear fluid that resembles keratin debris.

Occurring at all ages OKC shows a peak incidence in 2nd and 4th decade of life with male predilection (male: female is 1.6:1). In our presented case, the patient is a 29-year-old male who is within the above-mentioned peak age category.

Mostly seen in the mandible compared to the maxilla with the ratio of 2:1. In the mandible, a common site of occurrence is usually the angle–ascending ramus region (69-83%). In our reported case, radiographic investigation reveals the occurrence of the lesion in the mandibular angle – ramus region.

If the cyst involves the anterior mandible the lesion crosses the midline and in maxilla it involves the maxillary antrum, nasal floor, premaxilla, and maxillary third molar region. OKC can also arise from temporomandibular joints. [5]

Clinically, OKC manifests as swelling with or without pain, discharge, displacement of teeth and occasionally paresthesia of the lower lip. [1] This relates to the aforementioned case where the patient presented with diffuse swelling and pain, and gave a history of pus discharge. Paresthesia of the right side of the face was present and no displacement of the tooth was seen.

Radiographically, OKC is distinguished as a well-defined, unilocular or multilocular (25-40%) radiolucent lesion with smooth margins. This lesion may involve impacted tooth (25-40%), expansion of the cortical plates (buccal > lingual) with or without perforation. [5,1] In the presented case, the panoramic radiograph reveals a well-defined multilocular lesion with smooth margins and CBCT reveals no expansion but perforation of the lingual cortical plate on the right mandibular region with an intact buccal cortical plate.

A specific characteristic of OKC is the cystic growth in the anteroposterior direction through medullary spaces of the bone due to which there is a minimal bone expansion in the initial stages. Expansion of buccal cortex in maxilla and mandible is 30% and 50% respectively. [1]

Histologically, OKC can be diagnosed by its specific features. A stained section may present with a ‘ribbon-like’ 8-10 layer thick stratified squamous epithelium, absence of rete-ridges, ‘Picket fence or Tombstone appearance’ of well-defined basal cell layer consisting of cuboidal or columnar cells, mostly parakeratinised surface keratinization which is corrugated and rippled with artefactual separation between the basement membrane and the overlying epithelium. Other histological findings can be the presence of cholesterol crystals, satellite cysts, daughter cysts and Rushton bodies. [15]

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In the presented case, the stained section revealed the presence of the above mentioned features with the absence of Rushton bodies.

The lesion has various histological and radiological differential diagnosis. The former includes Ameloblastoma, Central giant cell granuloma, Myxoma, Odontogenic cysts and the latter includes Dentigerous cyst, Radicular cyst, Residual cyst, Primordial cyst and Globulomaxillary cyst.

Depending upon the patient's age, extend of the lesion, location of cyst, involvement of the adjacent structures and histological variant of the lesion these treatment modalities can be put forth. Conservative treatment methods are enucleation and marsupialization. [6] Enucleation is the complete removal of the cyst along with the cystic lining, due to thin friable envelop of OKC it is difficult to completely remove the cystic lining which leads to recurrence as high as 30%. [7,14] In marsupialization, the cyst is decompressed and the cystic lining is exposed into the oral cavity by creating a window of 1cm into the cyst and suturing the lining with the oral mucosa transforming the cyst into a pouch. [12,13] Due to the pathological tissue left in-situ, the recurrence rate is 33%. [14]

Whereas aggressive treatment modalities are peripheral osteotomy, chemical curettage with Carnoy's solution, cryotherapy and resection. [6] Peripheral osteotomy is done following enucleation in which a rotary instrument is used in reduction of peripheral bone followed by filling the defect with Iodoform gauze having a recurrence rate of 18%. [9,14]

Chemical curettage with Carnoy's solution also follows enucleation, in which Carnoy's solution consisting of 3ml chloroform, 6ml absolute alcohol, 1ml of glacial acetic acid and 1g of ferric chloride is used as the cauterizing agent having a recurrence rate of 9%. [8,9,14]

Liquid nitrogen is the devitalizing agent used in cryotherapy. This method is also an adjunct to enucleation. In OKC the agent kills the epithelial remnant or satellite cyst but because of its aggressive behavior the recurrence is 38%. [10,11,14]

The treatment method with the lowest recurrence rate of 0% is Resection which is either segmental (without maintaining the continuity of the bone a segment of maxilla or mandible is removed) or Marginal (with maintaining the continuity of the bone a segment of maxilla or mandible is removed). Reconstructive measures are necessary to restore the aesthetics and function. [8,14]

The use of combination therapy where the enucleation is followed by chemical curettage using Carnoy's solution and peripheral osteotomy can be employed. The recurrence rate incases of these treatment option is as low as 8%. [14] In this method after enucleation the defect is rinsed using saline and then a gauze soaked in carnoy's solution is used to pack it and left in place for 3 minutes. Thus the cystic wall is completely removed, following which peripheral osteotomy is conducted and the overlying attached gingiva is excised. Finally, the defect is packed with Vaseline- lodoform gauze. [9] In the presented case, taking into consideration the age, location, severity, extent of the lesion and the surrounding structures being affected the patient underwent segmental resection of the right side of mandible and customized reconstruction plates were used to rehabilitate the jaw function and aesthetics.

Recurrence rates in Odontogenic Keratocyst range from 25-60%. Meticulous surgical treatment shows a reduction in recurrences. The mean age for recurrence has been determined as 4 years for males and 7 years for females. Thus long-term follow-up is crucial. Recurrence of OKC can be associated with incomplete removal of cystic lining, epithelial lining being thin and friable, perforation of bone, adherence to adjacent soft tissue, development of new OKC in the adjacent area from the remnants of dental lamina, satellite cysts or daughter cysts grow into an OKC. [5]

Syndromes associated with multiple OKC are Nevoid Basal cell carcinoma syndrome which consist of features like bifid ribs, ectopic calcification of the falx cerebri, Gorlin goltz syndrome which is multisystemic, Marfans syndrome with classical feature of long limbs, fingers and toes, Ehlers danlos syndrome which has features like joint hypermobility and many other syndromes like Noonan's syndrome, orofacial digital syndrome and Simpson golabi – behmel syndrome can be ruled out with respect to the presented case due to the absence of syndrome-specific features.[2,16]

IV. CONCLUSION

The lack of evidence to support hypothesis such as colonality leads to Odontogenic Keratocyst no longer be considered as a neoplasm. Though, OKCs being considered a cyst or neoplasm, still remains as an area of debate. The most effective treatment for this lesion is indefinite, even though there are various treatment modalities for the management of this lesion. The clinician should decide on if it's advisable to carry out an incisional or excisional biopsy depending on the size, location and behavior of the lesion. With the implementation of appropriate imaging modalities like Panoramic radiograph and CBCT, the most accurate diagnosis and the best suitable treatment option can be drawn out.

The management of OKC should focus on proper diagnosis, conservative treatment as far as possible, using adjuvants like Carnoy's solution, peripheral osteotomy, cryosurgery if available and long-term follow-up up to 5 years to rule out any recurrences.

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FIGURE LEGENDS

FIGURE 1: Patient's profile with the swelling on the right mandibular region.
FIGURE 2: Restricted mouth opening.
FIGURE 3: OPG showing multilocular radiolucency extending from the right angle of the mandible to the coronoid process
FIGURE 4: A- 3D view showing radiolucency on the right ramus. B- Axial view showing radiolucency in the ramus. C & D- Coronal and a panoramic view showing multilocular radiolucency with Intra bony septa
FIGURE 5: Histopathological section

FIGURES

Fig 1

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Fig 2

Fig 3

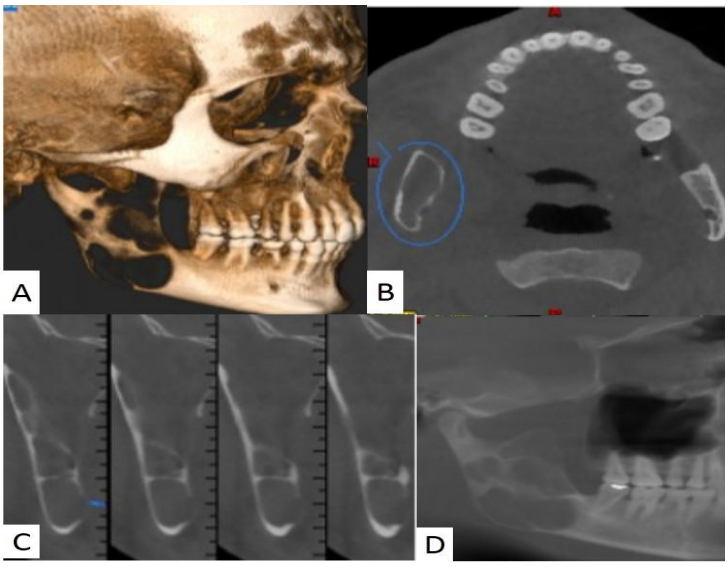

Fig 4

Fig 5