Introduction

The identification and description of odontogenic keratocyst (OKC) tumors were done in 1876 as odontogenic developmental cysts of epithelial origin and later in 1956 it was characterized by Phillipsen.[1,2] Pindborg and Hansen in 1962(2) mentioned the histologic criteria required to diagnose OKC. As the origin of this cyst was thought to be the primordium of the tooth the initial terminology was given as “primordial cyst.” Later, in 1992, the World Health Organization preferred the term “odontogenic keratocyst” for such cysts with a keratinized lining in the histologic grading of odontogenic tumors.[3] OKC is best known for its repetitive occurring potential, invasive nature, and its sporadic association with the nevoid basal cell carcinoma syndrome (NBCCS). Three histologic forms were known initially which includes an orthokeratinized variant, a parakeratinized variant, and combination of these two variants. The classification of the orthokeratinized variant as a discrete clinical entity of “orthokeratinized odontogenic cyst” is due to its less invasive clinical nature and repetitive pattern of this variant.

The percentage of OKCs versus other cysts of the jaws was given by different authors as follows; Pindborg and Hansen (1963) - 7%, Brannon (1976) - 11%, Hjorting-Hansen et al. (1969) and Toller (1972) - 9%, Pindborg and Hansen (1969) - 11% and Brannon (1976) - 9%.

Case Report

A 62-year-old patient came to the Department of Oral Medicine and Maxillofacial Radiology with a chief complaint of pain in lower left back tooth region since 1 and ½ months. The pain was persistent, throbbing, dull and continuous which aggravated during lying down and relieved on taking medications. Medical and allergy history were not significant. The patient had the habit of bidi smoking since 20 years around 10-15 bidis/day.

Extraoral examination revealed no gross facial asymmetry [Figure 1a]. On palpation, a bony hard swelling was present extending 2 cm away from the distal aspect of 36 which was not tender. Intraoral examination of soft tissue showed a healing extraction socket site present irt 38 with normal appearance of the overlying alveolar mucosa. On palpation overlying mucosa appeared smooth, firmly adhere to underlying periosteum with no evidence of bony expansion on buccal and lingual side of 38. The tooth 37 was tender on percussion [Figure 1b].

Examination of the hard tissue showed the presence of stains and calculus and generalized attrition. With the above findings of the site of lesion and history a provisional diagnosis of keratocystic odontogenic tumor (KCOT) of the left mandible was given.
An orthopantomogram (OPG) was taken which revealed missing 38 and presence of well-defined multilocular radiolucency on the left mandibular region measuring about 8 cm × 4 cm extending from mesial aspect of 34 to sigmoid region anteroposteriorly and superiorly from body of ramus to lower border of the mandible inferiorly with loss of lamina dura of 36, 37 [Figure 2].

Computed tomography scan was done to determine the exact location of the borders of the tumor [Figure 3].

Fine needle aspiration cytology was done, and sample showed clear scanty fluid which was watery in consistency. Histopathological report of the H and E stained cytological smear showed few cells with hyperchromatic nucleus in clusters, few keratin, and few inflammatory cells suggestive of keratin-filled lesion.

Biopsy from multiple hard and soft tissue lining was taken which showed parakeratinized stratified squamous cystic lining with surface corrugation and flat interface of underlying connective tissue wall with the separation of the epithelium from the capsule showing tombstone appearance [Figure 3].

After relevant investigations, a final diagnosis of OKC was given.

Treatment included surgical excision of the lesion with a wide margin of 1 cm, and Carnoy’s solution applied after excision. Reconstruction was done using Free Fibula osteocutaneous flap [Figure 4]. An OPG was taken postoperatively [Figure 5].

Discussion

The OKC is a unique and prevalent clinical and histologic lesion with aggressive nature. It usually arises in the dental lamina, but some suggest a probable origin from basal cell component.[5] About 70% or more cases involve the mandible, especially in the third molar, angle, and ramus areas. Next, most common site of occurrence is the maxillary third molar followed by mandibular premolar and maxillary canine region.[2,6]

It accounts for 10% of all jaw cysts and shows the prevalence in wide age groups. Symptoms include pain, swelling and drainage, especially with larger lesions and half of all these lesions are noticed as incidental radiographic findings.[7] Due to its potential to grow within the medullary bone, they

Figure 1: (a) Extraoral examination revealed no gross facial asymmetry. (b) Intraoral picture

Figure 2: Panoramic radiograph well defined multilocular radiolucency on the left mandibular region

Figure 3: Biopsy specimen and computed tomography image showing extent of lesion

Figure 4: Carnoy’s solution application and fibula reconstruction

Figure 5: Postoperative radiograph and extraoral picture
tend to become extremely large with undue clinical signs or symptoms.

Radiological examination of most of the cases reveals a well-defined radiolucency with thin corticated margins with the majority of these being unilocular, but larger lesions present as multilocular lesions.\cite{7,4}

Various percentage of radiologic patterns of OKCs as shown in Table 1.

20-40% of OKCs are usually found with an unerupted tooth and bear a resemblance to a dentigerous cyst. Root resorption is relatively rare with OKCs.\cite{8}

The OKC shows highly distinct histopathological features with uniform cyst lining, wavy parakeratin formation, hyperchromatic, and palisaded basal cells and a flat interface between the epithelium and underlying connective tissue wall. When the cyst is inflamed, these classic microscopic features are often completely lost, which can lead to an inappropriate diagnosis. NBCCS (also known as Gorlin-Goltz syndrome or jaw cyst-basal cell nevus-bifid rib syndrome) should be considered when a patient presents with multiple OKCs. NBCCS is an inherited genetic condition which occurs due to mutation of the PTCH1 gene. Palmar and plantar pits, bifid ribs, multiple basal cell cancers of the skin and calcified falx cerebri are the other findings of this syndrome. The lesions found with NBCCS are comparatively less aggressive than traditional basal cell epithelioma, because of this the designation “nevoid,” or having biologic behavior more similar to a nevus was mentioned.\cite{6}

About 25-30% of OKC lesions tend to recur unlike most other cysts of odontogenic origin, and most recurrent cases are seen during the first 5-7 years after treatment.\cite{9} Up to date, there are no practical instruments or techniques available to surgeons to help predict the recurrence of the lesions. The probable etiology for the cause of recurrence includes inappropriate expulsion of the original cyst lining, development of a new lesion from residual epithelial islands and or genotypic variations between lesions.\cite{9} Because of these probable reasons, the treatment protocol for OKCs is still controversial. The clinician should treat these lesions in such way to reduce both the risk of recurrence and morbidity of the patient.

**Treatment**

Treatment modalities for OKC commonly used with a conservative approach for large lesions are marsupialization and decompression. They are preferred to preserve bone, teeth, and preventing damage to other vital structures, and also for decreasing the chances of pathologic fracture. They are the treatment modalities recommended for pediatric and poor surgical patients. The principle behind these procedures is to decrease the cystic osmotic pressure by exposing it to the surrounding oral cavity. It is helpful in a bone deposition at the periphery of the lesion and also a progressive reduction in the cyst size.\cite{10} The most reliable and single-step procedure which leads to ultimate complete resolution of the cystic lesion is marsupialization.

Two-step procedure in the management of OKC’s is decompression which involves the placement of a surgical drainage tube, the following enucleation after the cyst has reduced to a manageable size.

Enucleation is the complete and intact removal of a cystic lesion by surgically husking it from the surrounding tissues. 17-56% of the recurrence rate of enucleation alone has been reported.\cite{11} Because of this, many surgeons prefer a combination of enucleation and adjunctive therapies to eliminate any residual cyst lining and islands within the cyst wall.

Adjunctive therapy includes the application of Carnoy’s solution which destroys cyst remnants by using chemical cautery. 18% reduction in the recurrence potential when both combination of enucleation with adjunctive treatment performed.\cite{11}

The surgical approach for OKC’s is resection. It is defined as the surgical excision of a section of the involved maxillary or mandibular jaws. If a rim of uninvolved bone is left behind it is known as marginal resection, whereas segmental resection involves the removal of a complete jaw without any continuity to the adjacent structures. This procedure is considered as an aggressive treatment modality which most commonly helps in eliminating the chances of recurrence, but somehow it is regarded as a radical approach in the treatment of OKC which often leads to the morbidity of affected patients.

An ample amount of research is going on over the past few decades regarding the genetic and molecular factors which are involved in the pathogenesis of OKCs. The cyst was previously considered as developmental in origin, but now based on available evidence suggests that it should be included as a benign neoplasm.\cite{12,13}

Due to the presence of higher levels of the biologic proliferation markers Ki67, proliferating cell nuclear antigen in OKCs compared to other dentigerous cysts and radicular

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<th>Author names</th>
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<td>Park and Kim (1985)</td>
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cysts, indicating the proliferation of the lining epithelium in the pathogenesis of OKC.\textsuperscript{4,14,15}

The WHO in 2005 in view of its evidence of neoplastic origin officially assorted the term OKC as KCOT.\textsuperscript{16} This term KCOT is the most current and precise nomenclature, but practically these both terms OKC and KCOT are synonyms and indistinguishable.

Most of the oral pathologists, including those at the University of Tennessee still using the nomenclature OKC in biopsy reports to decrease dilemma among the professionals who may not be used to the term KCOT. Currently, there are no supplementary advanced laboratory techniques available associated in the diagnosis of OKC except the traditional routinely used hematoxylin and eosin stained microscopic slides. Perhaps continued research in this regard will successfully help in identifying an accurate diagnostic tool which aids pathologists and surgeons in sorting out the aggressive and recurrent lesions and thereby guiding the treatment outcome and prognosis.\textsuperscript{17}

Summary

The present case of OKC was noticed in a 62-year-old male patient on the left mandible showing the clinical and radiographic presentation, diagnosis, treatment, and follow-up. Research is still going on appropriate treatment modalities for OKCs because of it is genetic and molecular basis of pathogenesis. Surgeons should thoroughly examine each case individually and provide with different treatment options to the patients.

References

5. Neville BW. Oral and Maxillofacial Pathology. 3\textsuperscript{rd} ed. St. Louis, MO: Elsevier/Saunders; 2009.