Case Report

Recurrent keratocystic odontogenic tumor of right maxillary sinus involving the right infraorbital rim

ABSTRACT

Keratocystic odontogenic tumor (KCOT) is a benign odontogenic tumor with an aggressive behavior and high recurrence rate. The most common site of predilection is the posterior mandible. In contrast, KCOTs occurring in the maxillary region are relatively rare. However, the maxillary involvement poses a greater and increased threat, due to proximity to vital structures such as maxillary sinus, orbital floor, and infratemporal fossa. This report presents such a case of KCOT involving the maxillary sinus eroding the floor of the orbit and provides an account of the factors that need to be considered during management.

Keywords: Infraorbital rim, keratocystic odontogenic tumor, maxilla, maxillary sinus

INTRODUCTION

The term “odontogenic keratocyst (OKC)” was coined by Philipsen in 1956. Owing to its locally aggressive behavior and higher tendency of recurrence, the World Health Organization in the year 2005 has renamed it to keratocystic odontogenic tumor (KCOT) rather than a cyst.[1]

KCOT shows a bimodal age distribution with its first peak in the second and third decade and the second peak in the fifth and sixth decade of life.[2] It is more commonly seen in males with a M:F ratio of 1.3:1[3] and has an increased site predilection for the posterior body and ramus of the mandible, with the maxillary involvement being very rare (<1%).[4] Diagnosis of KCOT is confirmed by histopathology though radiographic features may be suggestive of it.

A wide array of treatment modalities is available from marsupialization to enucleation, primary closure, packing open with adjuvant therapy such as cryotherapy or camoy’s solution, marginal or radical resection. However, the success of the treatment depends mainly upon the site involved, size of the lesion, the proximity of the vital structures, and appropriate surgical procedure with a regular clinical follow-up.[5]

Here, we present a rare case of KCOT involving the anterior maxilla and the right maxillary sinus with perforation of the infraorbital rim, and the orbital floor emphasizes the importance of interdisciplinary cooperation involved in customization of the treatment protocol intraoperatively to preserve the form and function of vital structures.

CASE REPORT

A 45-year-old female patient reported to the outpatient Department of Oral and Maxillofacial Surgery, Adhiparasakthi Dental College and Hospital, with a chief complaint of painless swelling on the right side of the face for the past...
6 months with watering of the eyes for 1 month. The swelling was initially smaller in size and had gradually increased in size to its present state. The patient gives an alleged history of a surgery done at the same site in 2005. However, no further details or records were available with the patient in this regard. There was no history of any trauma, tooth extraction, nasal obstruction, or epistaxis.

On extraoral examination, a swelling of about 30 mm × 22 mm was present in the right maxillary region which was oval involving the lower eyelid and obliteration of the nasolabial fold [Figure 1]. The swelling was nontender, nonfluctuant and had well-defined margins. The overlying skin was not attached to the swelling and was freely movable; the skin over the swelling had no abnormal color or surface changes. On palpation, the swelling was firm in consistency, and there was no paresthesia. There was no restriction of extraocular muscle movement, and bilateral pupillary light response was normal. There was no significant enlargement in the regional lymph nodes.

On intraoral examination [Figure 2], tenderness was elicited in the right vestibule between the canine region and buccal frenum of the maxilla. Radiographic examination of orthopantomograph (OPG) revealed well-defined multilocular radiolucent lesion with sclerotic border present in the right maxilla extending between right maxillary molar (16) and left maxillary canine region (23) mesiodistally and superiorly involving the right orbit with discontinuity in the right infraorbital margin. No evidence of root resorption was present. Computed tomographic scan revealed two well-circumscribed lesions present in the anterior maxilla and the maxillary sinus region measuring 2.24 cm × 1.90 cm and 3.71 cm × 2.71 cm, respectively, [Figure 3a and b] and occupying the right maxillary sinus [Figure 3c]. There was also deviation of nasal septum toward the left side and also obstruction of the right osteomeatal complex. The cystic lesion involved the lateral, medial walls of the right maxillary sinus. The roof of the sinus was completely eroded due to the lesion along with the infraorbital rim but not penetrating into the orbital tissues and the cavity [Figure 3b].

Aspiration of the right maxillary region yielded a serous fluid with a brownish tinge confirming the cystic nature of the lesion. Incisional biopsy was performed in relation to 13–15 regions under local anesthesia and was sent for histopathological examination. Microscopical feature of the specimen was that of nonspecific inflamed cyst.

Under general anesthesia, a high vestibular incision was placed from the nonpathological site of the left maxillary
canine to maxillary right first molar region. White keratinaceous material exudated from the anterior incisional area [Figure 4]. Mucoperiosteal flap was raised; thorough curettage was done in relation the anterior region. The other cystic lesion in the right maxilla was enucleated by blunt dissection [Figures 5-7] to prevent damage to the orbital contents as there was no bony separation between the right orbital tissue and the sinus lesional area. Irrigation was done with normal saline. The eyeball was postoperatively supported by surgical gelfoam with the support of 2% povidone-iodine-soaked roller gauze pack which was placed in the enucleated right maxillary sinus region. Even though the cystic lesion which was involving the infraorbital rim, it was not involving the orbital cavity or the soft tissues; as there were no clinical signs of diplopia, enophthalmos, or any restricted movement of the eyeball, the procedure did not warrant for an immediate reconstruction, but still due to the high recurrence nature of KCOT, we subjected the patient on close follow-up and considered for reconstruction using calvarial graft on later stages. Surgical site closure was done using 3-0 vicryl suture.

The enucleated mass was sent for histopathological evaluation. On the 5th postoperative day, the entire roller gauze pack was removed from the surgical site. The histopathological examination of the H- and E-stained section shows a parakeratinized stratified squamous epithelium of six to eight cell thicknesses with surface corrugation. The basal layer showed palisading arrangement of the cells, and the epithelial-connective tissue junction was flat with the absence of rete ridges. The underlying connective tissue wall showed parallelly arranged collagen fibers. Strands of odontogenic epithelium were also noticed in the connective tissue wall at few places. Inflammatory cells were very minimal in nature. These features were suggestive of KCOT [Figure 8]. On follow-up, the 1st week postoperatively patient had slight pain, and wound gaping was evident at preoperative biopsy site in relation to the right maxillary premolar region (14 and 15) for which wound debridement and resuturing done with COE pack. At 1-month postoperative, the patient had no symptoms, wound healing was satisfactory, and the patient is suggested for regular follow-ups at every 3-month interval.

**DISCUSSION**

In the early 19th century, Mikulicz described a lesion similar to OKC as a part of familial trait affecting the jaws. In 1926, the term “cholesteatoma” was used to describe a cyst or open cavity filled with keratin. Robinson in 1945 described a primordial cyst mainly due to its origin from primordial structures of the tooth such as enamel organ or dental lamina.

KCOT is a benign unicystic or multicystic intraosseous neoplasm of odontogenic origin which arises from the dental lamina both in the mandible and maxilla.

Many researchers have found that KCOT occurring from the second to third decade of life and mostly the mandible is affected, especially in the ramus part of the mandible being the
most common site. However, KCOT occurring in the maxilla and involving the maxillary sinus is an unusual presentation; here, we report a case where a female patient had a recurrence of cystic lesion of the right maxillary sinus in her fourth decade of life; there was no proper record of the previous surgical history for the patient, but we concluded this with presenting complaint which had recurred after 11 years.

The ratio for KCOT predilection between males and females is 1.3:1, and the occurrence rate of KCOT in the maxilla has been recorded <1%. In previous studies, the KCOT was completely restricted in the maxillary sinus without alveolar bone or erupted teeth association. An interesting aspect in our case, there was the erosion of roof, lateral, medial wall, and floor of the right maxillary sinus, and there was no root resorption, no alveolar process involvement.

In all KCOTs recorded till now, the major chief complaint was swelling present which would expand without pain or without any discharge. In our case, the patient had an oval-shaped swelling in the right maxillary sinus region, which was of slow-growing nature, with no signs of pain. The patient had epiphora in the right eye, due to obstruction in the nasolacrimal duct.

The recurrence rates of KCOTs have always been placed on the higher rate because of its thin cystic lining, size, shape, and placement of the lesion in inoperable regions where enucleation is not favorable for surgeons. Usually, KCOT reoccurs after 5 years postoperatively, and it can extend up to 15 years according to many literatures. In our case, the recurrence duration was 11 years.

Radiographic features in literatures suggest unilocular or multilocular lesions; long-standing lesion shows scalloped margin with regional resorption of the cancellous bone. In our case report, OPG reveals radiolucency present in the anteroposterior direction crossing the midline with sinus involvement and minimal intraoral expansion in the right maxillary sinus region. Computed tomography shows expansive nature of the cystic lesion invading the roof, lateral and medial walls of maxillary sinus, and medial and lateral pterygoid plates being not affected and also displacing the inferior rectus muscle superiorly.

There are large rooms for controversies in the treatment of KCOT. The treatment modalities are basically divided into conservative and aggressive. Conservative management includes enucleation with and without curettage and marsupialization. On the contrary, aggressive management includes peripheral ostectomy, electrocauterization, resection, and chemical cautery with carnoy’s solution.

In the case reported here, an enucleation and simple curettage of the cystic cavity margins were done in the anterior maxillary region extending from left maxillary lateral incisors to the right maxillary canine. On the other hand, the larger lesion occupying the entire right maxillary sinus was decompressed and enucleated into, and the overlying mucosa was also excised. During enucleation, the surgical plane was identified using the finger dissection which had a great help in identifying the pathological tissue and the normal tissue as the surgical site being the orbital cavity was of vital importance.

Oral surgeons and pathologist often find great difficulty in instituting the appropriate treatment modalities for each case of KCOT because it is challenging to determine the true nature, identification, and management of diseases affecting the maxillofacial regions.

The previous literature shows where degenerative changes of the cystic epithelium secondary to intense inflammation in the wall; those cases were diagnosed by pathologist as inflamed OKC.
Due to its developmental origin, usually, KCOT does not show much of inflammatory component, and it is not a characteristic feature of KCOT. However, it has been reported in the previous literature as KCOT for few cystic lesions with inflammatory component. Similarly, in the present case report, preoperative histopathological report suggestive of inflamed cyst which showed some areas of ulceration with loss of epithelium which is filled with fibrinopurulent exudate.

Pindborg and Hansen in 1963 suggested some basic histological features for KCOT; these include epithelial lining which is thin and uniform in thickness with no evidence of rete ridges, well-defined basal cell layer with cuboidal or columnar cells often arranged in palisaded manner, and thin spinous layer with direct transition from the basal cell layer with the exhibition of intracellular edema seen in spinous layers of cells. There may be keratinization which can be either parakeratotic or orthokeratotic with corrugated keratin layers. The fibrous tissue wall is also thin and uninflamed.

Our postoperative histopathological report suggested the presence of encapsulated cystic lesion lined by an epithelium, with a uniform thickness of 6–8 cell layers with corrugated parakeratinized surface, with palisaded basal cell layer and contained columnar cells with hyperchromatic nucleus. Inflammatory cell infiltrate was seen in underlying connective tissue with strands of odontogenic epithelium also seen in connective tissue. The connective tissue capsule was composed of thick parallelly arranged collagen fibers lined by fibroblast.

**Genetic mutations**

Protein patched homolog gene (PTCH) mutation as found in many other tumors such as basal cell carcinomas and squamous cell carcinomas which has been reported in both syndromic and sporadic KCOT. It is a tumor suppressor gene occurring in 9q22.3-q31, which inhibits the oncogene smoothened (SMO) by forming a receptor complex. Upon binding to the ligand sonic hedgehog gene, PTCH releases the inhibitory effect on SMO and thus the oncogenic activity continues.

The PTCH gene inactivation occurs by means of two literature hypotheses. The first literature involves inheritance of one mutant allele; the second literature involves the mutation of the other allele resulting in loss of heterozygosity and thus degeneration of cyclin D1 and P53. Intracystic injections of SMO protein antagonist have the greatest potential as a future treatment option.

**CONCLUSION**

Multiple surgical approaches are available in treating KCOT such as decompression, marsupialization, enucleation with or without adjunct (Carnoy’s solution, cryotherapy), and resection. Depending on the site of occurrence of KCOT in the maxilla, we can treat conservatively by enucleation without the use of carnoy’s solution. Hence, the aggressive behavior and high reoccurrence rate is always a challenging task for the maxillofacial surgeons in managing KCOT occurring in the maxilla. Herewith, we did a conservative management for KCOT with decompression, enucleation, and curettage.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**Acknowledgment**

We extend our heartfelt thanks to our student Hema Ravi, CRRI, Adhiparasakthi Dental College, for the extended support.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

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