Case Report

Mucosal keratocyst of buccal mucosa: A rare entity

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Abstract

The odontogenic keratocyst (OKC) is a developmental odontogenic cyst that is important due to its specific histopathologic features and clinical behavior. It is well known that the OKC arises from cell rests of the dental lamina. This cyst is important as it shows a different growth mechanism and biologic behavior from other common odontogenic cysts. In rare cases, OKCs occur in sites other than intraosseous sites. The most common location of peripheral OKC is the gingiva, but mucosal, epidermal and even intramuscular sites have also been described. The origin of peripheral OKCs is still under controversy. We, hereby, add a case of peripheral OKC located in the soft tissue of the buccal mucosa to the scarce literature consisting of only a few case reports and small case series of peripheral odontogenic cysts.

Keywords: Buccal mucosa, mucosal keratocyst, odontogenic keratocyst

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INTRODUCTION

An odontogenic keratocyst (OKC) is a developmental odontogenic cyst, comprising of about 3%–12% of all odontogenic cysts.[1] The odontogenic OKC is different from other cysts of the head and neck region. This is due to its ability to attain a large size before the occurrence of any clinical signs and symptoms, its high recurrence rate and an association with nevoid basal cell carcinoma syndrome. The histogenesis of intraosseous OKCs can be explained by the sources of odontogenic epithelium, that is, the dental lamina and its remnants and the extensions of basal cells from the overlying epithelium. The pathogenesis of OKCs associated with basal cell nevus syndrome is the involvement of the Hedgehog signaling pathway due to PTCH-1 inactivation.[2] Cysts and tumors which arise from the epithelium and its remnants associated with tooth development are termed as "odontogenic." These lesions usually occur centrally within the bone. There have also been case reports of OKCs in other, nonosseous locations. Most of these involve the gingiva, but mucosal, epidermal and even intramuscular sites have also involved. The origin of OKCs in areas other than intraosseous sites of the mandible or maxilla (mucosal OKCs) is still a controversy.[3] In this article, we report a case of peripheral OKC located in the soft tissue of the buccal mucosa in a 61-year-old male patient.

CASE REPORT

A 61-year-old male patient reported with a complaint of swelling in the right cheek region. The patient noticed the swelling 4 years back. He was generally healthy and had no medical history. On examination, a diffuse swelling was noticed on the right buccal region extending from the zygoma to the angle of the mandible.

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On inspection, a well-defined solitary swelling of approximate size 2.5 cm × 2.5 cm was noted, extending from below the zygoma to the lower border of the lower lip superoinferiorly [Figure 1]. The swelling was in line with the outer canthus of the eye to 2 cm in front of the external auditory meatus anteroposteriorly. The swelling was firm in consistency, nontender, slightly compressible nodular mass. Intraorally, a nodular mass of approximate size 2.5 cm × 2.5 cm was noted in the right buccal mucosa, extending from the lower lip posteriorly [Figure 2]. Overlying mucosa appeared normal and the swelling was firm and nontender on palpation.

On performing ultrasonography, a well-defined homogeneously hyperechoic focal lesion of approximate size 4.3 cm × 3.7 cm × 3.4 cm was noted on the right cheek. The right parotid gland showed normal sonomorphology. There was no evidence of duct obstruction. Based on sonographic findings, a differential diagnosis of a cystic lesion (most likely a mucocele) was made.

Figure 1: Clinical image showing swelling on the right side of the face extending from the zygoma to the lower border of the lip

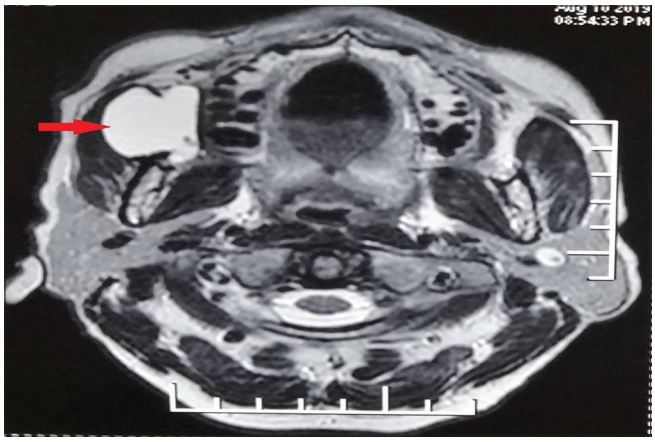

Figure 3: Imaging axial magnetic resonance imaging (T1-weighted imaging): A well-circumscribed nonenhancing T1 hyperintense cystic lesion deep to the masseter muscle (arrow head)

Magnetic resonance imaging (MRI) findings revealed the presence of a well-defined lobular T2 and T1 hyperintense lesion measuring 5.6 cm × 2.2 cm × 3 cm, extending deep into the right masseter muscle. MRI findings were suggestive of a benign cystic lesion with high-protein content [Figure 3]. Based on MRI findings, a differential diagnosis of lymphangioma/lymphatic cyst was made.

Intraoral aspiration revealed brownish liquid with numerous squamous cells, neutrophils and lymphocytes in the cytological examination.

The lesion was excised intraorally under general anesthesia through an incision along the anterior border of the mandibular ramus. The specimen obtained was 4 cm × 4.5 cm × 1.8 cm in size, dark gray in color with a firm consistency with an irregular surface [Figure 4]. The specimen was routinely processed and stained with hematoxylin and eosin.

Microscopically, the tissue sections showed a cystic lumen lined by parakeratinized epithelium of 5–6 cell layer

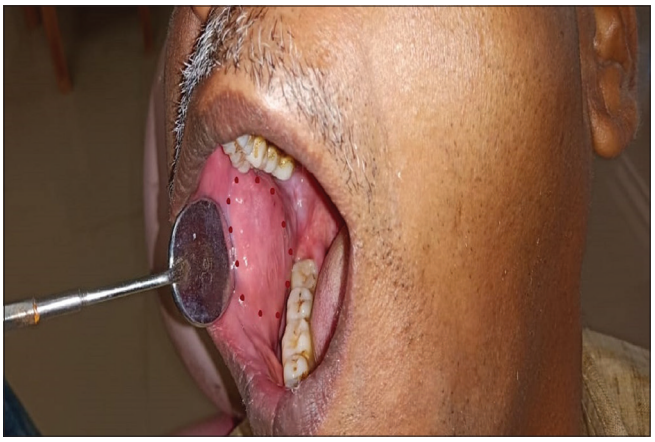

Figure 2: Intraoral view showing swelling at the right buccal mucosa (dotted area)

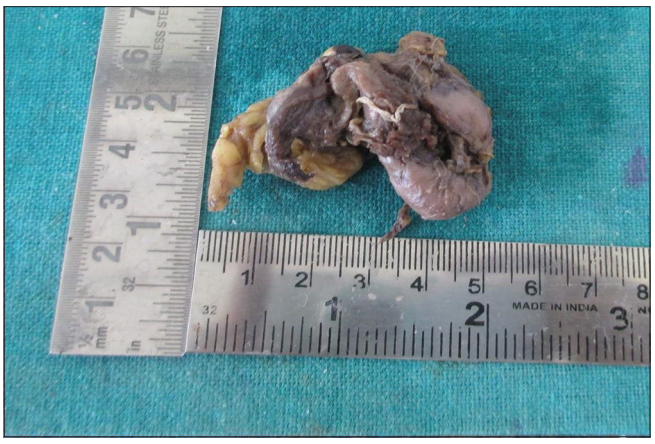

Figure 4: Gross image of a 4 4.5 cm × 1.8 cm in size, lobulated soft-tissue mass with an irregular surface

thickness. The epithelium connective tissue interface was generally smooth and flat with no rete ridges. The parakeratin layer showed surface corrugations. The basal cells were cuboidal to columnar in shape with nuclear palisading. The cystic lining in some areas exhibited numerous infoldings of the epithelium into the connective tissue wall. The underlying stroma was moderate to densely collagenous with sparse diffuse collection of chronic inflammatory cells, chiefly lymphocytes [Figure 5a and b]. All these features were compatible with OKC. There was no recurrence noted during a follow-up for 6 months.

DISCUSSION

The term "cyst" was derived from a Greek word *kysits* which means "sac" or "bladder." Cysts are relatively common and may be encountered virtually in any organ or tissue in the body. The head and neck region and the jaws are common sites for the occurrence of cysts. The frequency of occurrence of cysts within the jaws may be attributed to the peculiar embryology of the facial skeleton. The epithelium and epithelial residues capable of producing cysts are associated with teeth.[4] OKC is one of the most common odontogenic cysts. The two peculiar characteristics of OKCs due to which they have received special attention in the literature are: (1) their relatively high recurrence rate and (2) their tendency to grow within the medullary spaces of the bone in the anteroposterior direction.[1] Thus, the possibility of detection of OKC at an early stage is less.

The exact epidemiology of this lesion has not been established due to the limited number of reported cases. Nine cases of soft-tissue/peripheral OKCs located in the buccal mucosa have been documented in the literature, out of which only one of the cases was diagnosed in a female patient.[5] The mean age of occurrence was 49 years with a range of 16–74 years.[2,3,5-8]

The most common location of peripheral OKC is the gingiva, but mucosal, epidermal and even intramuscular sites have also been described. They are more common in males than females and tend to involve the mandible far more frequently than the maxilla.[9]

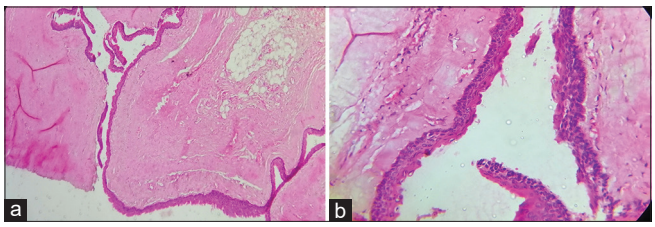

Figure 5: Histological findings. (a) H&E stained section (×10 view): The cyst wall is lined with parakeratinized epithelium with a corrugated surface. (b). H&E stained section (×40 view). Note the 5–6 cell layer epithelium with palisading of the basal cell layer

The origin of peripheral OKCs is still under controversy. The mechanism by which OKC can develop in the buccal mucosa is still not well known. One of the possible explanations is that the remnants of the dental lamina may get displaced in the buccal mucosa and persist during embryogenesis.[10] A recent embryological description of the developmental relationship between deciduous dentition and the oral vestibule may give an important clue. Accordingly, the remnants of the dental lamina entrapped in the buccal tissue during embryogenesis may develop a keratocyst with the features of OKC. Other odontogenic lesions such as ameloblastoma and odontoma have also been reported to develop in the buccal mucosa.[11]

Histologically, the lining epithelium is highly characteristic and is composed of:

- A parakeratinized surface which is typically corrugated, rippled or wrinkled
- A remarkable uniformity of the thickness of the epithelium, usually ranging from 6 to 10 cells thick
- A prominent palisaded, polarized basal layer of cells often described as having a "picket fence" or "tombstone" appearance.[9]

The intraosseous OKC associated with buccal mucosa is usually managed by total enucleation. In case of OKC affecting gingiva or palate, peripheral osteotomy may also be performed. Due to the less aggressiveness of peripheral OKC, more conservative line of treatment is employed, as in the present case.[1,3] The recurrence rate of peripheral OKCs, excluding the gingival cases, is 12.5%.[5] This is much lower compared to the recurrence rate of intraosseous OKCs which is about 10%–62%.[3]

Peripheral OKC should be differentiated from various similar lesions. OKCs occurring in the soft tissues may clinically resemble the gingival cyst of adults (GCA). It has been stated by many investigators that both these cysts (OKC and GCA) originate from the remnants of the dental lamina. Traumatic implantation of epithelium is said to be a cause of GCA, but in case of peripheral OKCs, the history of traumatic insult or surgical intervention has not been recorded so far. GCA occurs mainly on the lower anterior gingiva. In contrast to that of OKC, the lining epithelium of GCA usually consists of 1–3 layers, resembling reduced enamel epithelium.[12]

The main histopathologic differentials for OKC of buccal mucosa include epidermoid cyst, cutaneous

keratocyst (CKC), trichilemmal cyst and steatocystoma. There are some differences between OKC and an epidermoid cyst. Unlike the epidermoid cyst, the location and etiopathogenesis of peripheral OKC is unrelated to the lines of embryonic fusion. Epidermoid cysts contain large amounts of keratin and are lined by orthokeratinized stratified squamous epithelium with prominent stratum granulosum. The orthokeratinized lining epithelium of epidermoid cysts may resemble the lining of orthokeratinized odontogenic cyst (OOC). However, there are no separate case reports regarding peripheral OOCs, which may be probably because they were initially regarded as a variant of OKCs.[5]

CKC, which is generally encountered in patients with nevoid basal cell carcinoma syndrome, can also develop independently, but are rare. Nevertheless, the ectopic development of a CKC in the buccal mucosa can also occur. The corrugated lining of CKC resembles that of OKC, making it difficult to distinguish from OKCs.[13] They most commonly occur in the extremities and their cystic content shows thick brownish material containing clotted blood. The lining epithelium of CKC comprises several layers of squamous epithelial cells without a granular cell layer and skin appendages. CKC may also show the presence of mural daughter cysts.[14]

OKCs can be differentiated from trichilemmal cyst and steatocystoma by their catagen pattern and the presence of pilosebaceous units, respectively.[3,14] Trichilemmal cysts contain a cheesy, foul-smelling thick material and they mainly occur on the scalp. The lining epithelial cells exhibit no clearly distinguishable intercellular bridges. The lumen of trichilemmal cyst comprises of eosinophilic homogenous material. It can be also be distinguished from OKC by the presence of the sebaceous structures.[13]

CONCLUSION

We, hereby, add a new case of OKC involving buccal mucosa in a 61-year-old male patient to the very limited number of reports in the literature. The diagnosis of peripheral OKC can be established by proper clinical and histological evaluation. The present case of the peripheral OKC involving buccal mucosa also exhibited similar findings. Although buccal mucosa is a Rare site for occurrence of mucosal keratocyst, its occurrence at this location can be explained by displaced and persistent dental lamina rests in the buccal mucosa during odontogenesis in the embryo. The present case was surgically excised with no possible recurrence on follow-up for 6 months.

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.

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Conflicts of interest

There are no conflicts of interest.

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