Bilateral Keratocystic Odontogenic Tumor Invading Maxillary Sinuses: A Case Report

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Keratocystic odontogenic tumor (KCOT) is an odontogenic tumor which has the highest rate of recurrence. This report describes a rare case of bilateral ciliated epithelium-lined odontogenic keratocyst in the maxilla of a 15-year-old male. Panoramic radiography showed two lytic lesions on both sides of the maxilla associated with impacted third molars. Computerized tomography image revealed the involvement of both maxillary sinuses by the lesion which was destroying both sinuses' floors and posterior walls. Histopathologically, the keratinized epithelial-lined cyst of OKC was changed to a ciliated columnar hyperchromatic epithelium, suggesting the fusion of both these epithelia rather than a metaplastic transformation. The patient was treated by surgical enucleation of the cyst and was followed for one year. In this case report the biological behaviour of odontogenic keratocysts and its treatment options are discussed.

Keywords: Keratocystic odontogenic tumor; Odontogenic tumor; Paranasal sinuses

Introduction

Keratocystic odontogenic tumor (KCOT) is a benign uni- or multicystic, intraosseous tumor of odontogenic origin, with a characteristic lining of parakeratinized stratified squamous epithelium and potential aggressive, infiltrative behavior.The odontogenic keratocyst was first described by Philipsen in 1956 (1). The cyst, accounting for between 3% and 11% of all jaw cysts (2), is an important clinical entity because of its great tendency to recur. The traditional designation is odontogenic keratocyst (OKC), which stresses the benign behavior of this lesion. However, the term KCOT is recommended by the WHO as it better reflects the neoplastic nature of the lesion (3).

It is generally agreed that KCOT arises from odontogenic epithelium (4). Computed tomography (CT) scans may be valuable in evaluation of the patients with multiple nevoid basal cell carcinomas (NBCCs)-related KCOTs (5) as it is helpful in detecting cortical perforation and assessment of soft tissue involvement. In addition, contrast enhanced magnetic resonance imaging (MRI) may provide more detailed information (4). However, for exact diagnosis, histopathological examination is generally necessary (6-8). KCOT is more frequently observed unilaterally in the angle of the mandible, extending in anterior-superior direction particularly in the region of mandibular third molar (4, 9). KCOT occurs in different ages with a peak in the second and third decades (4). The mean age of affected patients is approximately 34.5 years, and the incidence is approximately 1.3 times higher in males than females (10). Patients should be carefully followed up after treatment as daughter cysts are common in KCOT and it has high rate of recurrence. It is known that the KCOT has a tendency to recur after enucleation (11), particularly within the first five years, since complete enucleation of a keratocyst is often difficult. Although complete resection of a keratocyst with the associated tooth decreases the recurrence rate but is often associated with morbidity. Recent studies have shown that marsupialization is a better alternative for the treatment of keratocysts (12).

Previously, some cases of massive and bilateral occurrence of KCOT in mandible (3, 13, 14) or unilateral lesion in maxilla (15-17) were reported. Management of KCOT in maxillary region and subsequent rehabilitation is challenging because by the onset of symptoms it would have progressed considerably beyond the confines of the maxilla. To the best of the authors' knowledge, there are few reports of nonsyndromic bimaxillary KCOT in the literature. Therefore in the present case, we reported a bilateral invasive KCOT in maxilla in an underage boy which was challenging in diagnosis and management.

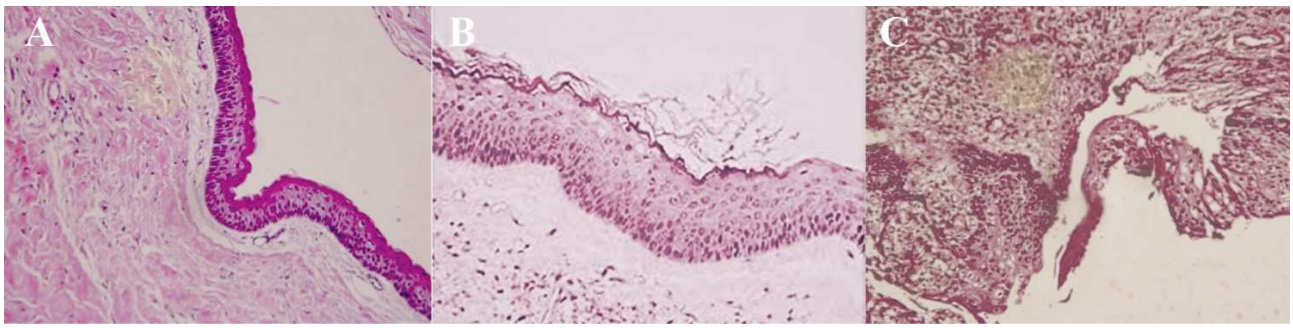

Figure 1. (A) The cyst lining shows few layers of a corrugated parakeratinized stratified squamous epithelium; (B) Palisading layer of cubidal and culumnar epithelial cells in basal layer; (C) Inflammatory cells in the connective tissue around the cyst lining

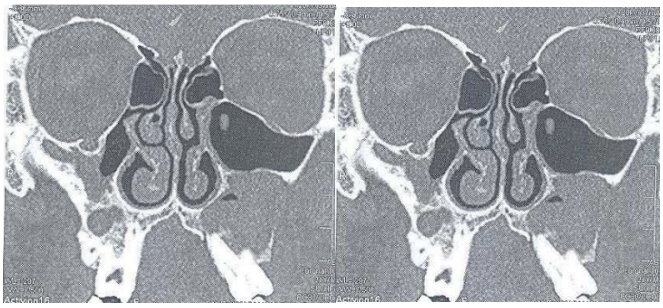

Figure 2. (A) View of Maxillary sinuses involvement and sinus wall perforation in CT scan image; (B) CT scan image after surgical procedures

Case Report

A 15-year-old Iranian boy complaining of facial swelling in the left maxillary region for three months was referred to Department of Oral and Maxillofacial Surgery of Taleghani Hospital, Tehran, Iran in December 2010. The patient had no similar familial history and was in good physical health. Three months prior to referral to this department, facial swelling was much less and a biopsy was obtained by an oral and maxillofacial surgeon and the histological evaluations confirmed diagnosis of dentigerous cyst.

The borders of the lesion were extended to nasal bone, zygomatic bone and infraorbital region. The orbital rims were palpable and visible lymphadenopathy over the left submaxillary premolar area was evident. He had slight tenderness on the region and the oral mucosa was erythematous with bony expansion and the swelling in the buccal vestibule was bony hard. A surgical scar from the posterior portion of the left maxillary vestibule from the previous biopsy was evident. No discharge was detectable from the lesion visually and parotid milking test was normal. Signs of Gorlin syndrome were notevident so this syndrome was excluded. Other major differential diagnosis included ameloblastoma, dentigerous cyst, and aneurysmal bone cyst.

The patient underwent the enucleation and curettage and the specimen was sent for histology. Microscopically the cyst was lined by few layers of a corrugated parakeratinized stratified squamous epithelium. The basal layer showed a palisading layer of cubidal and culumnar epithelial cells with polarized and hyperchromatic nuclei (Figure 1A, B). Epithelial lining of the cyst was rather thin and epithelium-connective tissue interface was flat with no epithelial ridge formation. Although no inflammatory cell was seen in the fibrous connective tissue inflammatory cells were detected in some regions of the loose connective tissue (Figure 1C).

CT scans showed two unilocular radiolucent lesions in both maxillary sinuses extending to the sinus roof and one tooth was involved in each sinus. The CT scans showed third molar displacement in both maxillary sinuses. The lesion had perforated the medial and posterior wall of both sinuses (Figure 2).

The general approach to treating OKC is enucleation and curettage (18). Alternative therapies such as marsupialisation and resection are also valid but have limited indications. The indications for resection are limited to two conditions: 1. multiple recurrences, and 2. large multilocular cysts, in which enucleation and curettage procedure may lead to an almost continuity loss (18). The treatment included an enucleation and curettage and no evidence of recurrence was detected in five year follow-up.

Discussion

In 1956 Philipsen described an odontogenic cyst with keratinized epithelial lining and some typical characteristics were described by Pindborg and Hansen, such as uniform thickness, a keratinized pattern with a corrugated surface, palisadedbasal cells with hyperchromatic nuclei and daughter cysts in the cystic wall (19).

The available evidence demonstrated the two main sources of the epithelium: the dental lamina or its remnants and extensions of the basal cells from the overlying oral epithelium (5). Recent studies have demonstrated the role of the protein patched homolog gene (PTCH) in the etiology of KCOTs (20-26). PTCH gene has been mapped to chromosome 9q22.3-q3122 and probably acts as a tumor suppressor (27). Studies on NBCCS and sporadic KCOT have expressed evidence of a two-hit mechanism in molecular level in the pathogenesis of these tumors demonstrating allelic loss, at two or more loci of 9q22 (28, 29) that leads to overexpression of bcl-1 and TP53 in the NBCCS. This supports the concept of neoplastic nature of KCOT (29). There is also abundant evidence that the PTCH gene might be a significant factor in the development of sporadic KCOT (21, 23, 25, 27). Furthermore, preliminary results have shown over-expression and amplification of genes located in 12q. Some linings may show some features of epithelial dysplasia (30) but malignant transformation to squamous cell carcinoma is rare (31). The keratocyst may be solitary or multiple. The latter usually indicates one of the stigmata of the inherited nevus basal cell carcinoma syndrome (NBCCS). Characteristically, multiple KCOT of the jaws is associated with the nevus basal cell carcinoma syndrome. Our case however had no related lesions fitting the components of this syndrome. It is important therefore to report this case because of its oddity and perplexing clinical presentation (32).

Unlike other odontogenic cysts, the odontogenic keratocyst has local aggressive behaviour producing a high rate of recurrence in up to 62.5% of cases. Adjacent teeth may be displaced but root resorption occurs rarely (4). The maxilla is involved in 23.5% of odontogenic keratocysts (10). Two cases of ciliated epithelium in odontogenic keratocysts in the maxilla was reported previously (33, 34). Although the origin of this epithelium seems to remain unknown, it has been suggested that it is a true metaplasia (35). In the present study, a case of odontogenic keratocyst in the maxilla partially lined with ciliated epithelium is presented. The CT examination disclosed a lesion in the left and right maxilla invading both sinuses floor. The microscopic examination revealed a cyst covered by two different epithelia. It is reasonable to consider that this microscopic characteristic suggests an aggressive biological behaviour of the odontogenic keratocyst.

In some rare cases KCOT involves and invades maxillary sinuses which are an uncommonly affected. Similar to the present case, previous KCOTs involving maxillary sinuses were all associated with unerupted third molar and were treated by surgical enucleation (15-17). Routine treatments for for KCOT include marsupialization, enucleation and curettage, and enucleation with chemical cautery (Carnoy's solution), thermal (cryotherapy), mechanical (peripheral ostectomy) cautery of surrounding tissue and osseous resection with or without continuity defect (11, 13, 14). Treatment options for aggressive and massive KCOTs are not very broad. Previously, a bilateral massive KCOT involving the whole body and symphysis of the mandible of an 11 year old girl treated with enucleation and open dressing was reported (3). Other reports also show that enucleation is more suitable for bilateral KCOTs of the mandible comparing to marsupialization and cryosurgery (13, 14). Although aggressive treatments have been recommended for large KCOTs (11), one should consider esthetic outcomes of the treatment and its effect on patient's life style; especially for young patients (36-38). The treatment method applied in the current study was surgical enucleation of KCOT with excision of mucosa in perforated regions to ensure removal of the residual lesion or daughter cysts adherent to soft tissue. In addition, the structure of the facial bone was preserved to provide better esthetic for the patient.

Conclusion

More detailed evaluation of clinical and radiological records and further molecular and histopathological studies will be useful in providing a comprehensive understanding of the biological behaviour of perplexing clinical presentations and invasion of odontogenic keratocysts to maxillary region.

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Conflict of Interest: 'None declared'.

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