Case Report / Olgu Sunumu

Maxillary sinus odontogenic keratocyst presenting with orbital pain and inferior rectus dysfunction

Orbital ağrı ve inferior rektus işlevsizliği ile seyreden maksiller sinüs odontojenik keratokist

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ABSTRACT

Keratocystic odontogenic tumor (KCOT) is a benign odontogenic tumor with a potentially aggressive and infiltrative behavior. In this article, we report a 22-year-old male patient with swelling over the right side of cheek and over the palate on the same side and orbital pain since two months. Medical history of the patient was not significant. Computed tomography scan showed a well-defined cystic mass occupying anterior and superior wall of maxilla from premolar region to the incisor region of opposite side and destruction of palate and inferior orbital wall. Cyst was excised via lateral rhinotomy incision completely with intact wall. Histopathological examination showed features of KCOT. Keratocystic odontogenic tumor is well-known for its tendency to recur, potential aggressive behavior, and defined histopathological feature. Keratocystic odontogenic tumor occurrence in the maxilla is unusual and its appearance in the maxillary sinus is very rare.

Keywords: Diagnosis; keratocystic odontogenic tumors; treatment.

ÖZ

Keratokistik odontojenik tümör (KOT) olasılıkla agresif ve infiltratif davranışlı bir benign odontojenik tümördür. Bu yazıda, yanağın sağ tarafında ve aynı tarafta damağın üzerinde şişkinlik ve iki aydır orbital ağrısı olan 22 yaşında bir erkek hasta bildirildi. Hastanın tıbbi geçmişi anlamlı değildi. Bilgisayarlı tomografi taraması maksillanın anterior ve superior duvarını karşı tarafın küçük azı dişi bölgesinden kesici diş bölgesine kadar kaplayan, sınırları belli bir kistik kitle ve damakta ve inferior orbital duvarda tahribat olduğunu gösterdi. Kist lateral rinotomi ile duvar sağlam kalacak şekilde tamamen eksize edildi. Histopatolojik inceleme KOT nitelikleri gösterdi. Keratokistik odontojenik tümörün nüksetme eğilimi, olası agresif davranışı ve tanımlanmış histopatolojik niteliği iyi bilinmektedir. Maksillada KOT oluşumu olağandışıdır ve maksiller sinüste görünümü çok nadirdir.

Anahtar Sözcükler: Tanı; keratokistik odontojenik tümörler; tedavi.

Keratocystic odontogenic tumor (KCOT) is a benign odontogenic tumor with potentially aggressive and infiltrative behavior.[1,2] Keratocystic odontogenic tumor is defined as a benign unicystic or multicystic intraosseous neoplasm of odontogenic origin with a characteristic lining of parakeratinized stratified squamous epithelium.[1,2] The cyst was termed

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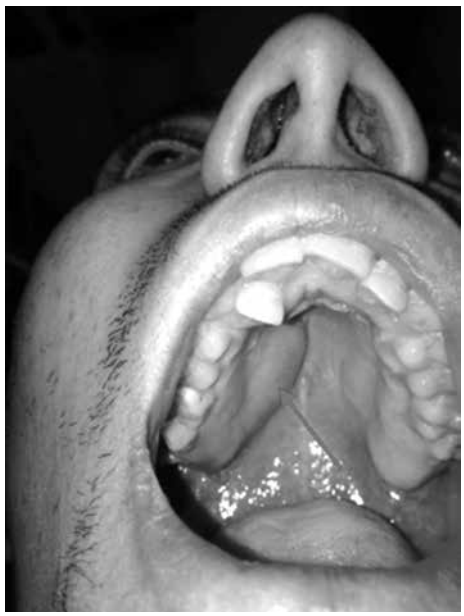

Figure 1. *Intraoral view of mass.*

"odontogenic keratocyst" by Philipsen HP in 1956 and the World Health Organization changed the name of odontogenic keratocyst into KCOT in 2005.[3] Keratocystic odontogenic tumors comprise approximately 11% of all cysts of the jaws. Occurring at any age, these tumors are more common in males than females, at an approximately 2:1 ratio. These tumors are very aggressive locally, with recurrence rates ranging from 3-60%.[1-4] Keratocystic odontogenic tumor predominantly develops in the mandible or maxilla bones and occasionally on the gingiva as a peripheral type of manifestation.[1-4] Computed tomography (CT) is useful for diagnosing and assessing cysts.[5] Treatment of KCOT remains controversial. The challenges lie in minimizing both the risk of recurrence and the surgical morbidity. Simple enucleation with or without curettage and marsupialization are conservative treatments for KCOT. Aggressive treatment includes peripheral ostectomy, chemical curettage using Carnoy's solution, and resection.[1-6]

The aim of this paper is to report a case of KCOT located in the maxillary sinus. Differently from the other rare reports presented in the literature concerning KCOT in the maxillary sinus and in our case the lesion presenting with orbital pain and inferior rectus dysfunction. The lesion was surgically removed and the patient has been followed up for two years showing no signs of recurrence.

CASE REPORT

A 22-year-old male patient presented with orbital pain and swelling over the right side of cheek and over the palate on the same side continuing for two months. He had no history of any trauma, tooth extraction or nasal obstruction or no other significant medical history. On clinical examination, a swelling was observed on the right hard palate and right cheek (Figures 1-3). Ophthalmologic examination showed inferior rectus dysfunction. He had no other symptoms such as pain, drainage or neurosensory deficit. He reported that the swelling had gradually increased in size over a period of two weeks.

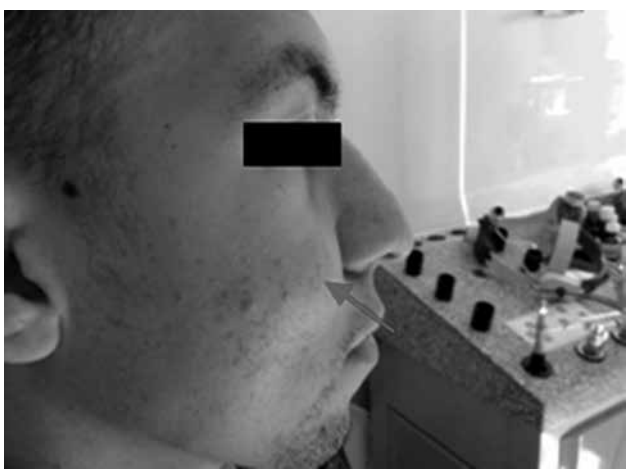

Figure 2. *Lateral view of patient.*

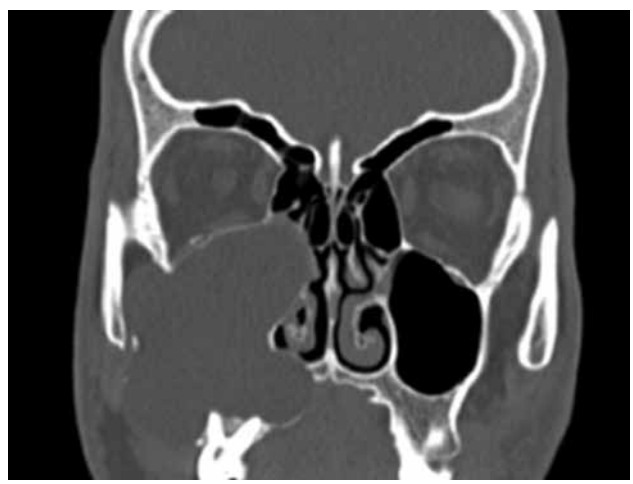

Figure 3. *Mass occupying anterior wall of maxilla from premolar region to incisor region of opposite side and destruction of palate and inferior orbital wall of same side.*

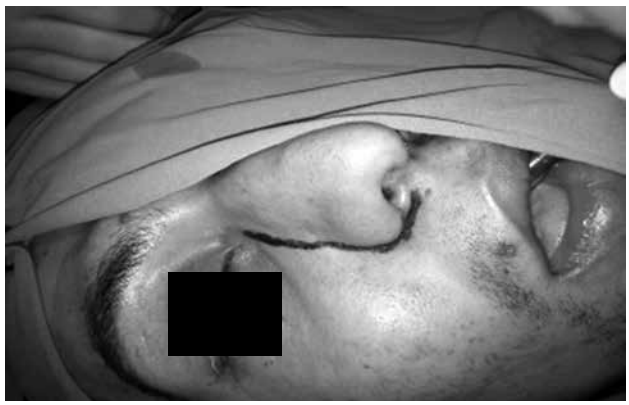

Figure 4. *Intraoperative view of patient.*

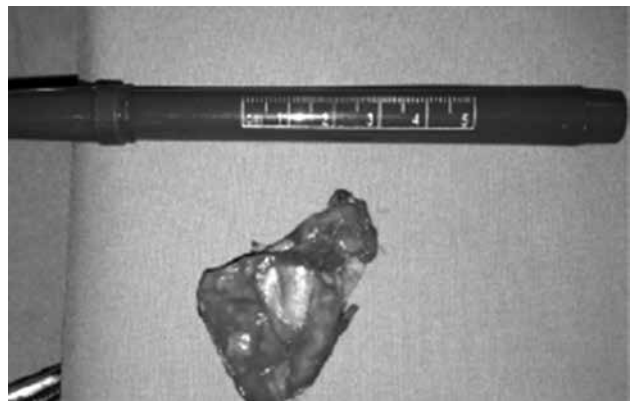

Figure 5. *Excised specimen of keratocystic odontogenic tumor.*

A written informed consent was obtained from the patient.

Coronal CT scan showed a well-defined cystic mass occupying the anterior wall of maxilla from premolar region to the incisor region of opposite side and destruction of palate and inferior orbital wall of the same side (Figure 3).

We approached the cyst by lateral rhinotomy incision (Figure 4). Skin was elevated. We saw that the front wall of the maxillary bone was extremely enlarged and thinned by mass. Then, a small piece of bone from the medial and front wall of the maxillary bone was removed. From the pop-up window, contents of the cyst were poured to obtain a wide space for maneuver by needle. Cyst was excised completely with intact wall (Figure 5). Cyst wall was tightly adherent to the underlying palatal mucoperiosteum. Histopathologically, our case showed parakeratinized lining and a capsule which was slightly inflamed with chronic inflammatory cell infiltration. A diagnosis of inflamed odontogenic keratocyst was established. The patient has been followed up for two years showing no signs of recurrence.

DISCUSSION

Keratocystic odontogenic tumors of the maxillary region are developmental cysts arising from cell rests of the dental lamina, the oral epithelial lining of the developing tooth follicle.[4] Discovery of increased mitotic activity in the cyst epithelium, potential for epithelial budding from basal layer or daughter cysts in the cyst wall, presence of chromosomal abnormalities, and role of mutation of the patched tumor suppressor gene in the etiology of KCOT resulted in its reclassification and renaming as keratocystic odontogenic tumor.[3] Keratocystic odontogenic tumor may occur at virtually any age, but the highest incidence is generally in the second and third decades of life.[4,7-9] Several authors have also noted a second peak between the fifth and eighth decades.[9,10] The sex distribution may be equal, or with male predominance.[4,7-9]

In our patient, the lesion was radiographically radiolucent, which can be seen in ameloblastomas, dentigerous cysts or odontogenic keratocysts; however, there was no associated impacted tooth or root resorption. Therefore, a dentigerous cyst or ameloblastoma was less likely. In the CT scan, the lesion appeared to be aggressive and destructive as seen by its large size, with an extension towards the inferior orbital wall. It is very rare and unusual for the maxillary KCOT to show orbital pain and inferior rectus dysfunction.

Keratocystic odontogenic tumor has a predilection for occurring in the mandible (75.58%) as compared to maxilla.[11,12] Maxillary KCOT tends to exhibit a smooth, round border while mandibular ones have scalloped border. In mandible, majority occur in third molar-ramus area, followed by first and second molar, and then followed by anterior mandible. In maxilla, the most common site is the third molar area followed by the cuspid region.[11,12] The lesion may be single or multiple, the latter case being more common in patients with nevoid basal cell syndrome. Clinically, they are asymptomatic but may present as toothache, swelling, and bone

destruction. Histologically, they are classified as parakeratotic and orthokeratotic, depending upon the type of keratin production.[5] Some clinical and molecular studies showed that parakeratinized and orthokeratinized KCOTs were significantly different in molecular area as well as the recurrence rate, with orthokeratinized KCOTs having a lower recurrence rate than parakeratinized KCOTs.

To our knowledge, the literature does not present any consensus on a uniform treatment plan for KCOT, and the recommended surgical management varies from marsupialization to *en bloc* resection since KCOT requires complete removal.[13] Enucleation with or without continuity defect are the treatment of choice. Presence of residual epithelium or an epithelial remnant after enucleation of the cyst and presence of satellite cysts in the cyst's wall are contributory factors for recurrence. Because of the high recurrence rates, the author believes that further studies should investigate the possible benefits of supplementary treatments, particularly the Carnoy's solution. Postoperative follow-up with regular radiographic examination is important with KCOTs because of the potential for recurrence. Keratocystic odontogenic tumors usually recur within five years after surgery; however, they may also recur more than 15 years later.[14]

In conclusion, the present case report emphasizes that any cystic lesion in the maxillary region should be evaluated cautiously. Although KCOTs are rare in the maxilla, CT scan and biopsy should be performed to evaluate the lesion and its extension particularly in the second decade when radicular cyst would be a more probable diagnosis.

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