Case Report
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A Large Orthokeratinized Odontogenic Cyst of the Maxilla: A Rare Case Report

Mohammad Mehdizadeh [a], Ali Lotfi [b], Saede Atarbashi-Moghadam [b]

[a]Associate Professor, Dept. of Oral and Maxillofacial Surgery, School of Dentistry, Qom University of Medical Sciences, Qom, Iran.
[b]Assistant Professor, Dept. of Oral and Maxillofacial Pathology, School of Dentistry, Shahid Beheshti University of Medical Sciences, Tehran, Iran.
Correspondence to Saede Atarbashi-Moghadam (email: S_atarbashi@sbmu.ac.ir).
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Objectives Orthokeratinized odontogenic cysts (OOCs) are relatively uncommon odontogenic cysts, lined by an orthokeratinized epithelium. These cysts show different clinical behaviors from the more common odontogenic keratocysts (OKCs), and it is important to distinguish these two types of cysts. Commonly, OOCs manifest as well-defined, unilocular, radiolucent lesions, with a high frequency in the mandibular region. In this case report, we aimed to describe a large maxillary OOC with prominent tooth displacement.
Case The patient was a 14-year-old boy with significant painless swelling in the upper jaw. The panoramic radiograph revealed a well-defined, unilocular radiolucent area from midline to the first right maxillary molar with prominent tooth displacement. The lesion was completely excised under local anesthesia. The histopathological sections demonstrated a cystic lesion lined by an orthokeratinized stratified squamous epithelium with a prominent granular cell layer.
Conclusion Based on the present results, OOCs can be large in size and may be characterized by ballooning expansion and tooth displacement. Therefore, knowledge of the clinical and radiographic features of these uncommon odontogenic cysts can facilitate an accurate diagnosis.
Keywords Odontogenic cysts; Maxilla; Jaw

Introduction

Orthokeratinized odontogenic cysts (OOCs) are relatively uncommon developmental odontogenic cysts, characterized by an orthokeratinized stratified squamous epithelial lining. [1-8] It is known that OOC is clinicopathologically different from the more common parakeratinized odontogenic cyst (OKC). [1, 5] Generally, OKC shows mutations in the protein patched homolog 1 (PTCH1) gene, which results in abnormal cell proliferation in the OKC epithelium. [1, 3, 4] Nevertheless, PTCH1 mutations have not been distinguished in OOC. [5, 6] According to a study by Wang et al. [8], although histopathological examination can be used to differentiate these two cysts, diagnosis is postoperative. On the other hand, radiographic examination is a preoperative diagnostic tool, which can facilitate proper treatment planning.

Evidence suggests that OOCs are more likely to show swelling than OKCs. They are also more significantly associated with impacted teeth compared to OKCs. [9] Additionally, multiple OOCs[1] and hybrid OOC with ameloblastoma have been described in the literature. [5] On the other hand, solitary or multiple lesions are not associated with Gorlin syndrome. [1] In a previous study, OOC with an associated OKC component and ghost cell pilomatrixoma-like keratinization and calcification was reported in a patient with Gardner syndrome. [10] The treatment of choice for OOC is enucleation with a low recurrence rate, although malignant transformation has also been detected. [11] In this case report, we describe a maxillary OOC in a 14-year-old boy.

Case Report

A 14-year-old boy was referred to our private oral pathology service center for the evaluation of painless swelling of the right maxilla and tooth displacement in the corresponding area for at least six months. The patient had no history of trauma or medical problems. An intraoral examination demonstrated buccopalatal expansion with bony hard consistency, extending from the midline to the first right maxillary molar with intact mucosa (Figure 1).

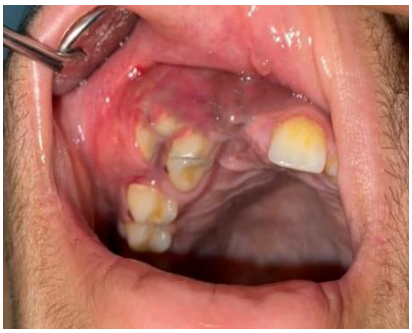

Figure 1: Intraoral photograph shows bucco-palatal expansion and tooth displacement.

The involved teeth were found to be vital. There was no cervical lymphadenopathy. The panoramic radiograph showed a well-defined, corticated, unilocular radiolucent lesion from midline to the first right maxillary molar with prominent tooth displacement (right canine and premolar teeth). The floor of the maxillary sinus was also pushed upward (Figure 2).

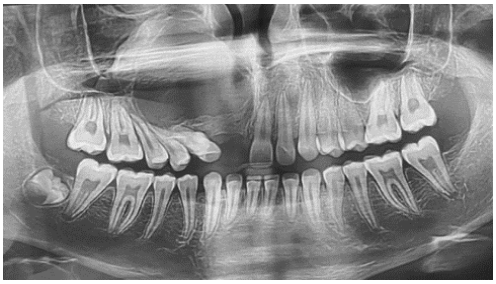

Figure 2: Cropped panoramic radiograph reveals a well-defined corticated unilocular radiolucency from midline to right first molar with prominent tooth displacement.

Based on the clinical and radiographic findings, odontogenic lesions, including calcifying odontogenic cyst and adenomatoid odontogenic tumor, were considered as differential diagnoses. The lesion was completely enucleated under local anesthesia. On gross examination, the lesion was creamy white, and the cyst contained a cheesy material. Microscopic sections showed a cystic lesion, lined by an orthokeratinized stratified squamous epithelium with a prominent granular cell layer (Figure 3). The underlying connective tissue demonstrated mild infiltration of chronic inflammatory cells. According to the microscopic and radiographic features, the diagnosis of OOC was confirmed. The follow-up showed that the patient was disease-free for six months postoperatively.

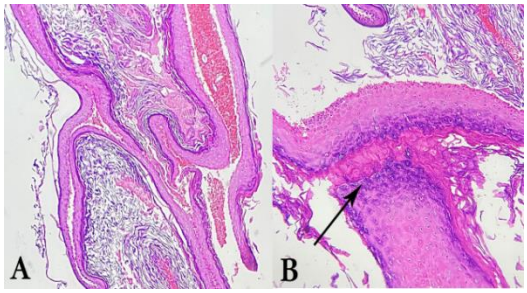

Figure 3: Microscopic sections show A) A cystic lesion lined by orthokeratinized squamous epithelium (H&E, ×100). B) Prominent granular cell layer (black arrow) (H&E, ×400).

Discussion

OOC is a relatively uncommon developmental odontogenic cyst of the jaw, with lower aggressiveness than OKC. [8, 12] It is characterized by a high male predilection, occurring mainly at an average age of 35.3 years (age range, 10-75 years). [5] Our patient was in the second decade of his life. The OOCs are commonly found in the mandible and posterior regions [5]; however, our case showed maxillary involvement. Evidence indicates that more than half of OOCs are associated with unerupted teeth and resemble dentigerous cysts on radiographs. [5, 8] The majority of OOCs are well-defined unilocular radiolucent lesions, and the rate of cortical bone expansion is higher in these cysts compared to unilocular OKCs. Moreover, OOCs are more likely associated with cortical bone destruction. [2] Both fusiform and ballooning patterns of buccolingual expansion have been reported in OOCs.[8] However, few cases containing calcified foci have been reported, which may be related to dystrophic calcification in a cholesterol granuloma. [5]

Wang et al. [8] found that features, such as root resorption and tooth displacement, are not observed in OOC. Nevertheless, in another study, root resorption and tooth displacement were detected in 12.3% and 9.2% of OOCs, respectively. [5] In our case, considerable expansion and a ballooning growth pattern were detected, along with prominent tooth displacement. The histopathological evaluation shows an orthokeratinized stratified squamous epithelium with a prominent granular cell layer and flat or cuboidal basal cells. [1, 5, 8] The corrugated surface and palisading of the basal cell layer, which are characteristics of OKCs, are not seen in OOCs. [1, 5] On the other hand, histiocytic lining, epithelial spheres, sebaceous differentiation, and papillary morphology have been described in OOCs; these features indicate the multi-potentiality of the odontogenic epithelium in OOCs. [5]

In previous studies, the expression of Ki67 and cyclin D1 markers was lower in OOCs than OKCs, representing a low proliferation rate. [2, 13] Thosaporn et al. [14] found that IPO-38, a proliferative marker, was more significantly expressed in OKC than OOC. Generally, the walls of OOCs rarely contain satellite or solid islands, which may be responsible for the lower recurrence rate of OOC compared to OKC. [5] The treatment of choice for OOC is enucleation of the cyst, and the prognosis is known to be excellent. [15] These lesions rarely recur, with an estimated frequency of 2.7%; however, the possibility of malignant transformation in recurrent OOCs needs to be considered.[5]

Conclusion

Although a microscopic evaluation can simply differentiate OKC and OOC, it is a postoperative diagnostic tool, while a clinicoradiographic evaluation is a preoperative diagnosis, which can aid us in proper treatment planning. Generally, OOC, which may be characterized by ballooning expansion and tooth displacement, can be large in size. Therefore, knowledge of the clinical and radiographic features of this uncommon odontogenic cyst can facilitate an accurate diagnosis.

Conflict of Interest

No Conflict of Interest Declared■

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