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Case Report

Orthokeratinized Odontogenic Cyst: An Unusual Histopathological Presentation

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Abstract

Introduction: The orthokeratinized odontogenic cyst (OOC) is an uncommon developmental cyst that occurs between the second to fifth decades, more commonly in males. It is a solitary lesion that mostly occurs in the mandible rather than the maxilla. Histologic features include a thin, uniform epithelial lining with orthokeratinization and a prominent granular layer below a noncorrugated onion-skin-like surface.

Case Presentation: A 40-year-old man presented with pain and swelling in the left mandibular canine molar area. The panoramic radiograph revealed a well-defined radiolucency extending from the left mandibular canine to the left mandibular molar, with scalloped projections between the teeth roots. Microscopic examination showed a cystic lesion lined by an orthokeratinized stratified squamous epithelium, and a prominent granular layer beneath the cornified layer was seen. The features were those of an OCC. **Conclusions:** From the demographic and radiographic perspectives, the features of OOCs can be similar, but more variation can be found on routine histopathological analyses.

Keywords: Odontogenic Cyst, Jaw, Developmental

1. Introduction

The orthokeratinized odontogenic cyst (OOC) is an uncommon developmental cyst that the world health organization (WHO) defined as a type of odontogenic keratocyst (OKC) in 1992 (1). It was first described by Schultz as a dermoid cyst in 1927, then Philipsen termed it a variant of the odontogenic keratocyst (OKC), which is less aggressive (2). In 1981, this cyst was specified by Wright as an orthokeratinized variant of OKC (3), and Li et al. later suggested the descriptive term of orthokeratinized odontogenic cyst (4).

The OOC occurs during the second to fifth decades of life, more commonly in males (5). It is a solitary lesion that mostly occurs in the mandible rather than the maxilla. It usually appears in the posterior mandible as a small, non-expanding, asymptomatic lesion (2). Radiographically, the cyst is usually described as a well-defined unilocular radiolucency. In two thirds of cases, the cyst is associated with an impacted tooth, usually an unerupted mandibular third molar (6). The histologic features include a thin, uniform, epithelial lining with orthokeratinization, and a prominent granular layer below a noncorrugated onion-skin-like surface (5). The hypocellular spinous cell layer is made up of polyhedral to flattened cells with an eosinophilic cytoplasm beneath the granular layer. The basal layer cells are usually flattened or cuboidal, without any palisaded or polarized nuclei (7). Here, an interesting case of OOC with unique histopathologic features is reported.

2. Case Presentation

A 40-year-old man presented with pain and swelling in the left mandibular canine molar area to the dental faculty at Hamadan university, Iran, in 2007. The panoramic radiograph revealed a well-defined radiolucency extending from the left mandibular canine to the left mandibular first molar, with scalloped projections between the tooth roots (Figure 1). The lesion was removed by surgical excision under general anesthesia, and the obtained material was analyzed for its histopathological features.

Microscopic examination showed a cystic lesion lined by an orthokeratinized stratified squamous epithelium with approximately 5 - 8 layers of cells. A prominent granular layer was seen beneath the cornified layer. The features indicated an OCC. Interestingly, the basal layer cells exhibited flat morphology in some areas (Figure 2), but a prominent palisaded basal layer and nuclear polarization were seen in other areas (Figure 3). Other epithelial changes included lichenoid tissue reactions, with band-like lymphocyte infiltration, hydropic degeneration, and focally hyperplastic rete ridges with different saw-toothed and club-

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Figure 1. Panoramic Film Showing a Radiolucent Area in the Left Side of the Mandible



Note the sclerotic border and the scalloping superior aspect of the cyst between the roots of the teeth.



Figure 3. High-Magnification (\times 400) Image Showing the Basal Epithelial Layer With Palisading, and Keratohyalin Granules on the Luminal Surface

shaped patterns (Figure 4). In addition, the basal layer hyperplasia suspended from the epithelium was obvious, revealing a basal cell carcinoma (BCC) (Figure 5). As with all inflamed odontogenic cysts, a hyperplastic squamous cell epithelium was also found. Mild dysplasia and pseudoepitheliomatous hyperplasia due to secondary inflammation were also seen. PAS staining for the detection of fibrinous deposition was negative.

3. Discussion

In the present case, unusual histopathological findings were observed in the OOC. Although lichen planuslike changes were seen in this case, in ordinary lichen planus the infiltration of lymphocytes is denser, and varying degrees of orthokeratosis and parakeratosis with hypergranulosis can be seen (8). Additionally, a PAS-positive basement membrane, which is found in oral lichen planus (8), was not observed in this case. The present cyst showed hyperchrome and palisaded basal layer cells in some areas, as well as a few clusters of basaloid cells with a palisaded outer layer, resembling BCC. However, a palisaded basal layer is also a histopathological finding of OKC (9). Some investigators believe that OKC is a neoplasm, which they prefer to call a keratocystic odontogenic tumor (KCOT) (9). According to the WHO classification, oral cystic lesions lined by orthokeratinized epithelium are not included in the spectrum of KCOT, as the lining epithelium of OOC lacks the typical features of OKC and has lower proliferative activity (10). It has been suggested that KCOT may



Figure 2. Low-Magnification (×100) Image Showing a Thin Epithelial Lining With Keratohyalin Granules, and a Thick Layer of Orthokeratin on the Luminal Surface



Figure 4. Low-Magnification (\times 100) Image Showing Hyperorthokeratosis, Saw-Toothed Rete Ridges, and Band-Like Infiltration of Lymphocytes Immediately Adjacent to the Epithelium



Figure 5. Low-Magnification (\times 100) Image Showing a Few Clusters of Basaloid Cells Suspended From Orthokeratinized Epithelium

arise from the dental lamina, but OOC may arise from the oral epithelium under the influence of the dental papilla or only the oral epithelium (11), and for OOCs associated with an impacted tooth, a different histogenesis has been suggested (12). On the other hand, a previous study showed that most OOCs and dermoid cyst specimens express CK10. This finding also suggests that OOCs may originate from the oral epithelial component, whereas KCOTs originate from dental lamina (13). This would explain the occurrence of KCOTs in the posterior mandible, where the dental lamina is more active at the age that patients develop this lesion (1). In addition, the unusual histopathological findings of the present case (lichenoid changes and basilar hyperplasia) can be explained by the suggested origin of the OOC. Additionally, less significant expression of p63 in OOCs compared to KCOTs suggests a lower proliferative and self-renewal potential for OOCs (1, 14), which may explain the different clinical behaviors between OOCs and OKCs.

OOCs occur across age groups varying from 20 to 50 years, and more commonly in males (3). The present case was a 40-year-old man, confirming this trend. The mandible appears to be more frequently affected than the maxilla, especially the posterior region, and indeed in this case, the posterior region of the mandible was involved. In one series, swelling, often associated with pain, was the most common symptom (4). In the current case, the patient had swelling and pain that might have been due to inflammation. Radiographically, an OOC usually appears as a unilocular radiolucency, as in the current case. OOCs are frequently associated with an impacted tooth, but in the present case, no such association was found. The differen-

tial diagnosis of an OOC includes other radiolucent lesions of the jaw. When the cyst is small, it might be diagnosed as a dentigerous cyst or paradental cyst, but when the lesion reaches a large size, odontogenic tumors, such as ameloblastoma and KCOT, should be ruled out. OOCs have radiographic characteristics similar to those of ameloblastomas and KCOTs; however, OOCs do not cause root resorption, which is a frequent characteristic of ameloblastomas and KCOTs (5). OOCs also do not tend to recur (2). The current case was treated with total surgical excision, and no recurrence was detected 8 years later.

In conclusion, from the demographic and radiographic perspectives, the features of OOCs can be similar, but more variation can be found on routine histopathological analyses.

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Footnotes

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