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Case Report

An unusually extensive orthokeratinized odontogenic cyst: A case report

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ABSTRACT

While odontogenic keratocysts (OKC) are fairly common and well-known, orthokeratinized odontogenic cysts (OOCs) are quite rare accounting for less than 1% of odontogenic cysts. The entity is characterized by the presence of a predominant orthokeratinized lining. OOC is much less aggressive as compared to OKC and simple enucleation is discerned as adequate treatment with minimal chances of recurrence. The present case report describes an extensive OOC that involved most of the angle and ramus of the mandible in a 25-year-old female.

1. Introduction

Odontogenic keratocyst (OKC) is well-known by surgeons and pathologists comprising about 11% of all the odontogenic cysts. On the other hand, an entity closely related histopathologically to OKC yet exhibiting a much different clinical course called 'orthokeratinized odontogenic cyst' (OOC) is relatively infrequent accounting for less than 1% of odontogenic cysts.[1]

In 1927, OOC was first described as an orthokeratinized variant of OKC which is still considered so by many even today.[2] Wright described OOC as an entity distinct from OKC in 1981. The World Health Organization (WHO), in 1992, defined OOC as an uncommon orthokeratinized type of OKC, and the trend continued in the second edition of the WHO classification of head and neck tumors.[3]

It was only in the third edition released in 2005, that OOC was excluded from the definition of OKC and the latter was reclassified as those cysts with a predominantly parakeratinized lining. Cystic lesions lined by orthokeratinizing epithelium were considered distinct but were separated from the spectrum of OKCs. At present the WHO considers OOC as a distinct entity and has cautioned against considering it as a variant of OKC.[4]

OOC is quite rare with prevalence about one-eighth of that of OKC.[3] While the latter is an aggressive lesion that tends to cause massive areas of destruction by proliferating in between the marrow spaces in an anteroposterior direction, the former is a relatively docile one.[5] The present case report describes an unusual case of extensive OOC that involved the majority of the angle and ramus of the mandible.

2. Case Report

A 25-year-old female complained of discomfort in the lower left back region of jaw since one year. The patient's medical history was unremarkable. Extra-oral as well as intra-oral examination of the patient did not reveal any significant findings in the region except for a missing mandibular left third molar.

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Orthopantogram showed a large, well-defined unilocular radiolucent lesion involving the angle and lower two-thirds of the ramus of the mandible (Figure 1A). The mandibular left third molar was impacted, vertically inverted, and displaced up to the sigmoid notch. A provisional diagnosis of unicystic ameloblastoma was considered while the differential diagnosis included other odontogenic cysts and tumors, particularly odontogenic keratocysts and conventional ameloblastoma.

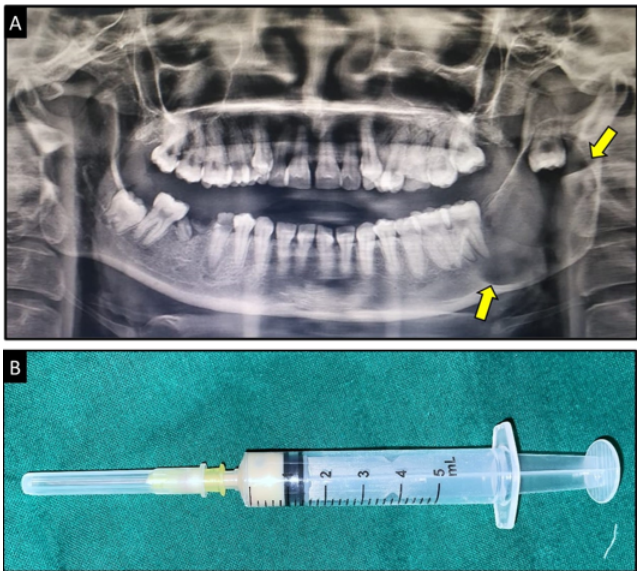

Fig. 1: A: Orthopantomogram showing a well-defined unilocular radiolucent lesion with severely displaced mandibular left third molar; **B:** Creamy aspirate obtained from the lesion.

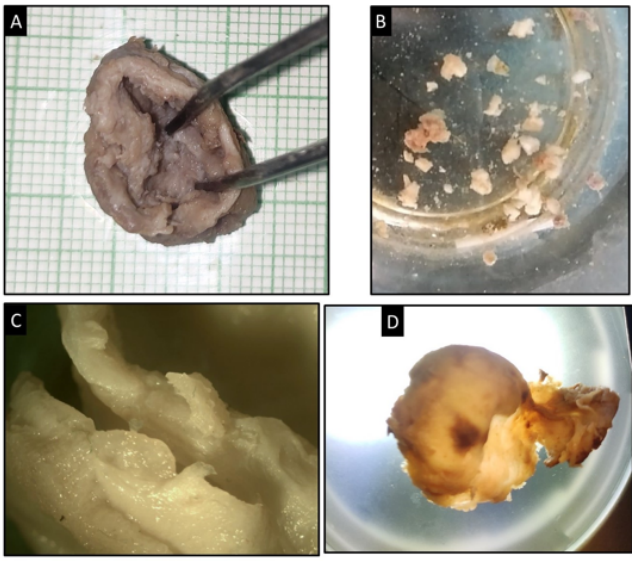

Fig. 2: A: Enucleated cystic lesion; **B:** Exuberant flakes of keratin; **C:** Lesion observed under steromicroscope (4.5x magnification); **D:** Transillumination of the excised cyst

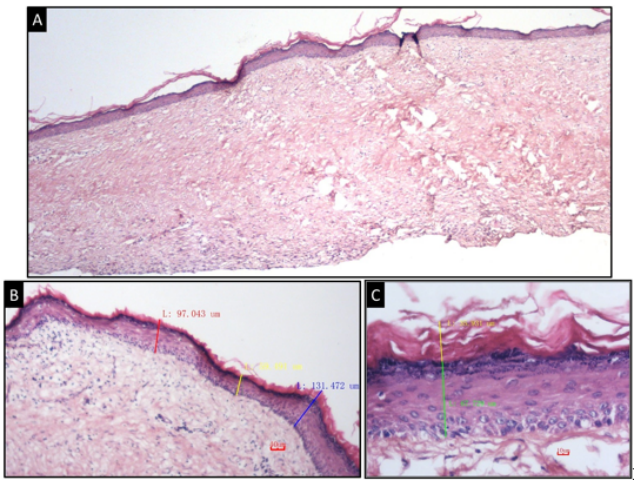

Fig. 3: A: Scanner view exhibiting cystic lumen lined by epithelium and capsule (H and E, original magnification x40); **B:** Orthokeratinizied lining varying from 60 to 130 μm in thickness (H and E, original magnification x100); **C:** Prominent stratum granulosum and stratum corneum with a thickness about half the thickness of the other layers of the epithelium combined (H and E, original magnification x400)

The lesion was exposed clinically by creating a window in the bone under local anesthesia. A cystic lesion was noted which elicited about 1.5 ml of creamy-white material having a cheesy consistency on aspiration (Figure 1B). The mandibular left second and third molars were extracted and the lesion was enucleated in toto.

Macroscopically, a cystic lesion of size 40 x 35 mm was noted. The excised specimen was whitish in color, had a smooth surface texture, and was firm in consistency (Figure 2A). The lumen of the cyst consisted of exuberant amounts of cheesy white material suggestive of keratin flakes (Figure 1B). Observation of the excised cyst stereomicroscope revealed that a 3 to 5 mm thick capsule having a smooth surface texture surrounded a central lumen (Figure 1C). Transillumination did not reveal any nodular ingrowths or calcifications within the lesion (Figure 1D).

Histopathology revealed a cystic lumen lined by orthokeratinized stratified squamous epithelium of 6 to 9-layer thickness with a prominent stratum granulosum (Figure 3A). Micromorphometry showed that the epithelial lining varied from about 60 μm to 130 μm (Figure 3B). The thickness of the stratum corneum was found to be about 35 μm, which was more than half the thickness of the other layers of the epithelium combined (Figure 3C). The interface between the lining epithelium and the underlying capsule was flat. The connective tissue capsule comprised parallelly arranged dense bundles of collagen fibers interspersed with few fibroblasts. Only scattered foci of mild inflammatory cell infiltrate were noted in the connective tissue.

Based on the clinical, radiographic, and characteristic histopathological findings, the final diagnosis was imparted as OOC.

3. Discussion

OOC is presently defined as 'an odontogenic cyst that is entirely or predominantly lined by orthokeratinized stratified squamous epithelium.'[4] This definition clearly implies the extent of orthokeratinization in the cystic lining required to diagnose a lesion as OOC. Establishing predominance of orthokeratinization is crucial as many other cysts particularly dentigerous cysts and OKC may exhibit focal areas of the same.[6]

While the present case of OOC occurred in a female, the available data states that it exhibits a male predilection.[7] OOCs show a peak incidence in the third to fourth decades as noted in the present case.[8] The lesions are generally asymptomatic and may even be discovered only during routine radiographic examination.[9] In the present case, it was only the slight discomfort felt by the patient that pointed toward the existence of the lesion which was revealed by radiographic investigation.

The mandibular posterior region is most frequently involved as noted in the present case.[4] The radiographic (well-defined unilocular radiolucency)[10] as well as histopathological features (uniform completely orthokeratinized epithelial lining) observed were also similar to those most frequently seen in OOCs. Besides gender, all the other clinicoradiopathologic features of the present case were characteristic of an OOC.

While OKCs tend to cause extensive destruction in an anteroposterior direction, OOCs are much less aggressive, making the extensive size attained in the present case an unusual phenomenon.[4] Even so, no cortical plate expansion or destruction was noted despite the lesion being quite extensive, which further points toward the unagressive nature of the lesion. Enucleation is considered as an adequate treatment for OOC and recurrence has rarely been described in less than 2% of cases.[4,10] No evidence of disease was noted after a follow-up of 18 months in the present case.

4. Conclusion

OOC is a much rare entity and is, therefore, not well-known to surgeons who may even consider it similar to an OKC. It is on the part of pathologists to distinguish the two and also guide the surgeons in formulating the treatment plan.

5. Conflict of Interest

None.

6. Source of Funding

None.

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