Expect the unexpected – A rare case of Orthokeratinized Odontogenic Cyst and its surgical management

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Μια σπάνια περίπτωση ορθοκερατινοποιημένης οδοντογενούς κύστης και η χειρουργική της διαχείριση

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Case report
Αναφορά περιστατικού

SUMMARY: Orthokeratinized Odontogenic Cyst (OOC) is a rare, developmental odontogenic cyst of the dental lamina. It was initially defined as the uncommon orthokeratinized variant of the Odontogenic Keratocyst (OKC), until the World Health Organization's (WHO's) classification in 2005 and 2017, where it was separated from the Keratocystic Odontogenic Tumor (KCOT) and has been included as a separate entity from the category of developmental odontogenic cysts respectively. It presents as a unilocular radiolucent lesion involving the posterior mandible and is frequently related to impacted teeth, often similar to other odontogenic cysts. Due to low local aggressiveness and less proliferative activity, it has to be differentiated from the other cysts in terms of surgical management. Here we report a rare case of OOC involving the maxilla along with an impacted canine and discuss the surgical management and why a secondary surgical intervention is unnecessary.

KEY WORDS: orthokeratinized odontogenic cyst, maxilla, impacted canine, surgical management, odontogenic cyst

ΠΕΡΙΛΗΨΗ: Η Ορθοκερατινοποιημένη Οδοντογενής Κύστη (OOC) είναι μια σπάνια, αναπτυξιακή οδοντογενής κύστη της οδοντικής ταινίας. Αρχικά ορίστηκε ως η ασυνήθιστη ορθοκερατινοποιημένη παραλλαγή της Οδοντογενούς Κερατινοκύστης (OKC), έως την κατάταξη του Παγκόσμιου Οργανισμού Υγείας (ΠΟΥ) το 2005 και το 2017, όπου διαχωρίστηκε από τον Κερατινοποιούμενο Οδοντογενή Όγκο (KCOT) και έχει συμπεριληφθεί ως ξεχωριστή οντότητα από την κατηγορία αναπτυξιακών οδοντογενών κύστεων. Παρουσιάζεται ως μονήρης ακτινοδιαφανής βλάβη στην οπίσθια περιοχή στην κάτω γνάθο και συχνά σχετίζεται με έγκλειστα δόντια, συχνά παρόμοια με άλλες οδοντογενείς κύστες. Λόγω της χαμηλής τοπικής επιθετικότητας και της λιγότερης πολλαπλασιαστικής δραστηριότητας, πρέπει να διαφοροποιηθεί από τις άλλες κύστες όσον αφορά τη χειρουργική αντιμετώπιση. Εδώ αναφέρουμε μια σπάνια περίπτωση OOC στην άνω γνάθο σχετιζόμενη με έγκλειστο κυνόδοντα και συζητάμε τη χειρουργική διαχείριση και γιατί δεν απαιτείται δεύτερη ευρύτερη χειρουργική επέμβαση.

ΛΕΞΕΙΣ ΚΛΕΙΔΙΑ: ορθοκερατινοποιημένη οδοντογενής κύστη, άνω γνάθος, κυνικός σκύλος, χειρουργική αντιμετώπιση, οδοντογενής κύστη

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Paper received: 9/4/2021 - Accepted: 16/7/2021

Παρελήφθη: 9/4/2021 - Έγινε δεκτή: 16/7/2021

INTRODUCTION

The orthokeratinized odontogenic cyst (OOC) is a rare, developmental odontogenic cyst originating from the cell rests of the dental lamina (1). It was first described by Schultz in 1927 as an orthokeratinized variant of the odontogenic keratocyst which was then called as keratocystic odontogenic tumors (KCOT) (2). In 1981, Wright termed it as orthokeratinized variant of odontogenic keratocyst (OKC) (3) But historically, it was Jeannel in 1885 who first treated a case of dermoid cyst which subsequently was an OOC (9). However, it was not until after Schultz's description in 1927 it had gained familiarity. Later in 1998, Li suggested the term "orthokeratinized odontogenic cyst" based on the histogenesis and is the most accepted term as of today. The summary of OOC terminology timeline is described in Table I. OOC frequently occurs between the third and fourth decades of life (10). The mandibular molar and ramus region are more commonly involved compared to maxilla (90.6% versus 9.4%) with a mandible-maxilla ratio of cases were 9.17:1, higher than that reported for KCOTs (11).

Radiographically, the OOC appears as a well-defined, unilocular or multilocular, radiolucent lesion frequently associated with an impacted teeth (12). Though OOC is similar to many odontogenic cysts & tumors such as adenomatoid odontogenic tumors, ameloblastoma, dentigerous cysts clinically and radiographically. The low aggressive cyst expansion, minimal biological activity and low recurrence with a unique histopathology sets it apart from other odontogenic lesions. Here we present a rare case of OOC involving the anterior maxilla of a young male patient which was found incidentally and discuss about its surgical management.

CASE REPORT

A 25 year old male patient presented with the chief complaint of crowded teeth and wanted to get it aligned.

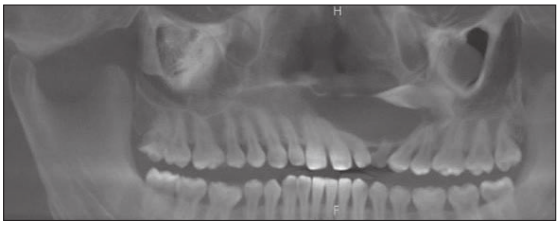

Fig. 1: Panoramic radiograph showing Unilocular radiolucent lesion in left body of mandible.

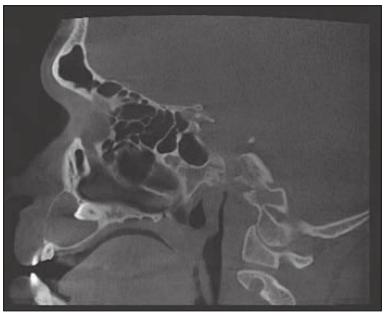

Fig. 2: Sagittal CT section showing well defined radiolucent unicystic lesion measuring 2 x 2 x 1.5cm with cortical expansion and associated impacted canine tooth within the anterior maxilla. No erosion of nasal floor.

The patient had a habit of thumb sucking till 7 years of age and has discontinued the habit, there was no other contributory history. On extra oral examination, there was no facial asymmetry or facial swelling. Intra-oral examination incidentally revealed a swelling over the premaxillary region involving 22, 23 and 24 measuring 3 x 1.5 cm which was soft on palpation. Expansion of buccal cortical plate was evident with crepitus felt at some areas. On further questioning, it was found that the patient had developed the swelling 3 month back but did not report it since it was asymptomatic. A panoramic radiograph taken revealed a well-defined radiolucency 2 x 3 cm in diameter with corticated border below the root apex of retained deciduous teeth 63. An impacted tooth was also observed within the radiolucency [Fig-

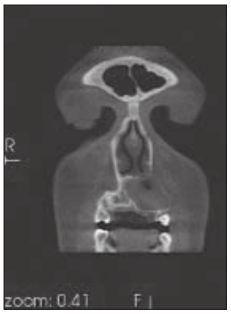

Fig. 3: Coronal CT Section showng expansile radiolucency.

Table 1
Chronology of OOC

Year	Description	Author
1885	Treated the first case of OOC as dermoid cyst	Jeannel [4]
1926	Reported a known case of OOC as Cholesteatoma	Hauer [5]
1927	Described the entity as dermoid cyst	Schultz [2]
1945	Described it as a variant of OKC	Philipsen [3]
1981	Described as an orthokeratinized variant of OKC	Wright JM [3]
1998	Termed as orthokeratinized odontogenic cyst based on histology	Li [6]
2005	OOC was considered a separate entity after reclassifying parakeratinized type of the cystic lesion as keratocystic odontogenic tumor (KCOT)	WHO [7] WHO [8]
2017	It was considered as a separate entity in the category of developmental odontogenic cysts	

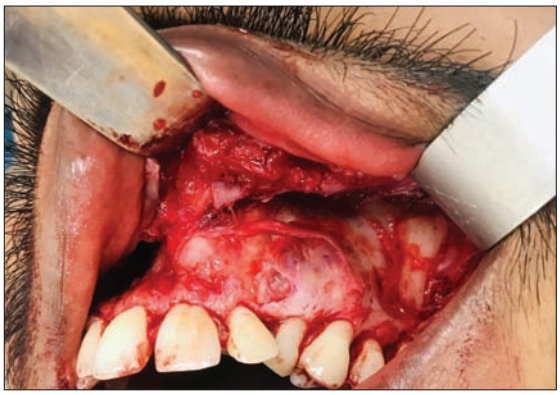

Fig. 4: Mucoperiosteal flap reflection.

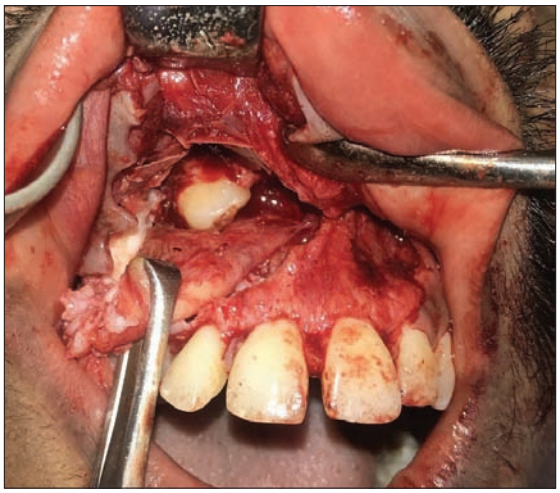

Fig. 5: Enucleation of the cystic lesion revealing the impacted canine.

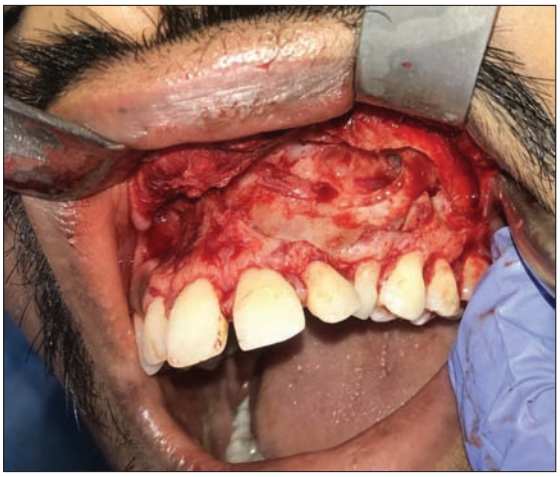

Fig. 6: Post surgical enucleated field.

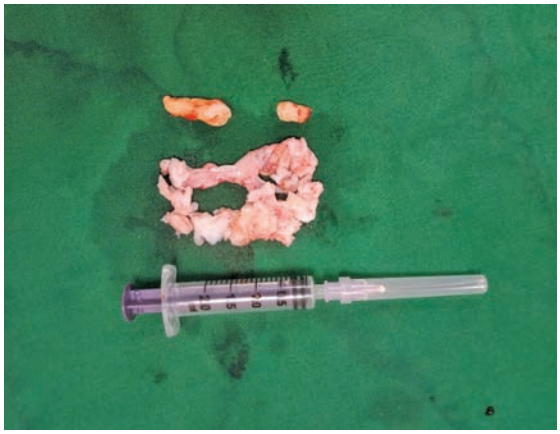

Fig. 7: Gross specimen of enucleated cyst and negative aspiration.

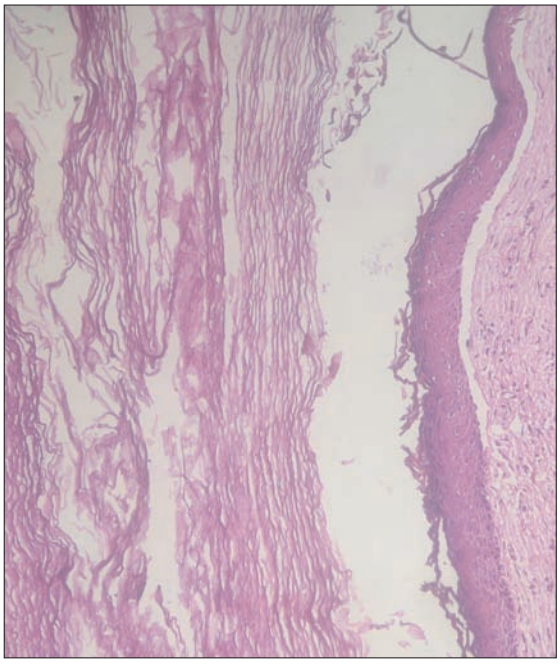

Fig. 8: Histopathological picture showing cystic cavity lined by an orthokeratinized stratified squamous surrounded by a fibrous cystic wall. The epithelium was 4-5 cell layers in thickness in most areas with a prominent stratum granulosum layer. (Hematoxylin & eosin staining, 10x).

ure 1]. To further study the expansion of the lesion. A computed tomography scan (CT Scan) was taken and it showed an expansile, osteolytic lesion with buccal cortical plate perforation [Figure 2 & 3]. A diagnosis of adenomatoid odontogenic tumor (AOT) with impacted canine was made with differential diagnosis of dentigerous cyst, OKC, unicystic ameloblastoma and OOC. No preoperative biopsy was planned as clinical, radiological and cyst aspiration findings were suggestive of benign cystic lesion. Surgical enucleation with peripheral ostectomy was planned. All laboratory findings were within normal range. Under local anesthesia, surgical exploration with full mucoperiosteal flap reflection was done [Figure 4], the impacted canine was seen and the cystic lining was surgically enucleated along with removal of the impacted canine followed by Peripheral osteotomy and extraction of the retained deciduous canine [Figure 5 & 6]. The reflected flap was then sutured and post-operative drug regimen consisting of antibiotics

& analgesics were prescribed. The patient healing was satisfactory and uneventful. The gross examination of the excised specimen revealed a thin cystic sac with luminal surface [Figure 7]. Microscopic examination of the specimen revealed shows a cystic cavity lined by an orthokeratinized stratified squamous surrounded by a fibrous cystic wall. The epithelium was 4-5 cell layers in thickness in most areas with a prominent stratum granulosum layer. The cystic wall was composed of densely arranged collagen fibres and fibroblasts. There was also evidence of inflammatory cell infiltrate predominantly lymphocytes with blood vessels in the cystic wall. These histopathologic feature was suggestive of Orthokeratinized Odontogenic Cyst [Figure 8]. Intraoral periapical radiograph taken a year after the procedure showed significant bone formation in the enucleated sites with no signs of recurrence [Figure 9].

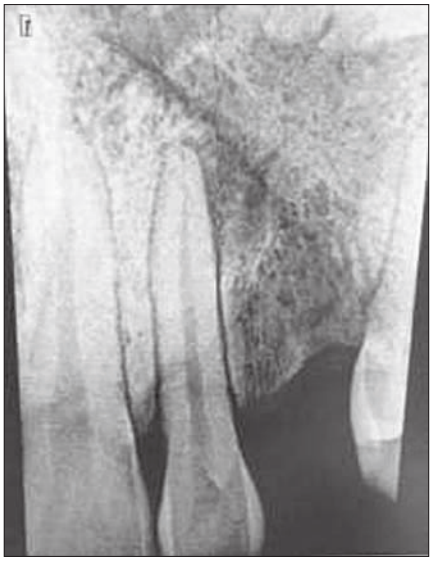

Fig. 9: Follow up intraoral radiograph revealing significant bone formation in the enucleated region with no signs of recurrence.

DISCUSSION

OOC represents 7-17% of all keratinizing cyst of the jaw (13). The prevalence of orthokeratinized variant ranges from 3.3% to 12.2% (5). However, the exact incidence of OOC cannot not be enumerated, as majority of the previously reported cases were considered to be a variant of OKC. However, the incidence of OKC is higher than that of OOC globally as well as in the Indian population (14), (15).

Li et al stated that males in their third and fourth decades of life are frequently involved with the occurrence of OOC (6), which was very similar to our patient. The age is crucial when deciding the choice of surgical treatment since employing aggressive surgical techniques in children can cause disturbances in the growth and development of jaws and teeth.

Swelling is the main clinical feature of OOC with mandible more frequently involved. This was contrary to our case, where the swelling was seen involving the anterior maxilla which is a rare occurrence and very few cases have been reported in the literature (16 - 18). Most of the swellings were asymptomatic in cases of OOC (19). OOCs are significantly associated with swelling, incidental discovery, well-defined margins and impacted teeth than OKCs. 48% of OOC were discovered as incidental findings with 41% first presented with swellings and 24% first presented with unerupted tooth (20).

Histologically, the OOC shows a cystic cavity lined by a regular thin stratified squamous epithelium, about 4- to 9-cell layers thick. This epithelium presents a well-defined basal layer that exhibits cuboidal or flat cells, prominent granular cell layer with nuclear hyperchromatism, and a thick superficial layer of orthokeratin. This entity must be differentiated from the OKC that shows a regular epithelium of 5- to 10-cell layers thick with the basal cells lined with an elongated nucleus and the presence of a characteristic superficial corrugated layer of parakeratin (21). Ultrastructurally, OOC shows loose attachment between superficial shreds of orthokeratin and a compact layer of underlying keratin. The histopathologic differential diagnosis should also include intraosseous epidermoid cyst. However, the absence of skin appendages in OOC is the differentiating feature. Satellite or daughter cyst which is common in OKC is not seen in OOC, this could potentially be a reason for its low recurrence (21).

In contrast to OKC, OOC has less proliferative activity and biological activity as evident by the immunohistochemical studies done which showed a reduced expression of Ki67 and p63 protein (19), (23), (24). Only 4% of OOCs showed recurrence and due to its rarity, more studies are needed to better understand the etiopathogenesis and clinical-radiographic feature of this lesion. The differential diagnosis of the OOC includes other radiolucent lesions of the jaws, mainly odontogenic lesions such as AOT, dentigerous cyst, paradental cyst, OKC and ameloblastoma. OOC presents similar radiographic characteristics with the ameloblastoma and OKC, such as its tendency to involve the mandibular angle or to appear as a multilocular radiolucency. Unlike these entities, the OOC does not cause aggressive growth and root resorption (25). In our case, considering the clinical, radiographic history, it was provisionally diagnosed as an AOT. However, upon surgical excision and histopathological evaluation it was diagnosed as OOC.

For our treatment, taking into consideration the age of the patient, we decided to surgically enucleate the cystic lesion as its remains the standard choice of treatment for OOC followed by peripheral ostectomy, as this would not only completely remove the cyst but also remove any remaining daughter cells.

Prognosis following enucleation is excellent and recurrence has been reported in less than 2% cases compared to OKC which presents a recurrence rate between 8 and 25% after enucleation. Larger lesions require surgical resection followed by chemical cauterization with carnoy's solution. Thus it is important to differentiate between the two entities. OOC should be considered always in the differential diagnosis of all the radiolucent lesions involving impacted teeth. This is a classic example of odontogenic lesion having clinicopathologic and radiographic features similar to AOT but on histopathological diagnosis was an OOC.

CONCLUSION

OOC exhibits distinctive clinical, histopathological and biological features that vary substantially from OKC with a better prognosis and lower recurrence rate. It should be mentioned that other radiolucent lesions of the jaws such as dentigerous cyst, ameloblastoma and OKC must be considered in the differential diagnosis of OOC. The patient age, extent of the lesion and histopathological picture should be taken into consideration when designing an effective treatment plan.

Abbreviations

OOC - orthokeratinized odontogenic cyst
KCOT - keratocystic odontogenic tumors
OKC - odontogenic keratocyst
CBCT - cone beam computed tomography scan
AOT - adenamatoid odontogenic tumor

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