Orthokeratinized Odontogenic Cyst of the Maxilla: Report of a Case and Review of the Literature

Nazanin MAHDAVI¹, Nasim TAGHAVI²

Department of Oral Pathology, School of Dentistry, Tehran University of Medical Sciences, TEHRAN, IRAN, 2 Shahid Beheshti University of Medical Sciences, TEHRAN, IRAN

ABSTRACT

The orthokeratinized odontogenic cyst is a relatively rare developmental odontogenic cyst of the jaws that occurs predominantly in males between the third and fourth decades. According to the 2005 World Health Organization's classification, orthokeratinized odontogenic cyst is not a part of the keratocystic odontogenic tumor spectrum. We present a case of a 41-year-old male with a history of remarkable lingual expansion in the anterior part of the maxilla since last year with rapid growth in the last three months as a unilocular well–defined radiolucent lesion extending from maxillary left central incisor to the right second premolar. Orthokeratinized odontogenic cyst is a specific type of keratinized odontogenic cysts which is completely different from keratocystic odontogenic tumor from the viewpoint of the clinical and pathological features.

Key Words: Odontogenic tumor, Jaw cyst, Odontogenic cyst

INTRODUCTION

The orthokeratinized odontogenic cyst (OOC) is a relatively rare developmental odontogenic cyst of the jaws, identified as the orthokeratinized variant of the odontogenic keratocyst by Wright in 1981(1, 2). Due to the significant differences between these two keratinized cystic lesions of the jaws, the World Health Organization's (WHO) new classification of the head and neck tumors (2005) reclassified the parakeratinized type of the cystic lesion as keratocystic odontogenic tumor (KCOT) (3). Since then, OOC is not a part of the spectrum of KCOT and should be considered as a separate entity.

OOC occurs predominantly in the mandible and in males (4-6). These cysts are often asymptomatic and discovered as an incidental finding especially in the orthodontics' radiographs (6, 7). Here we report a case of a relatively large size OOC in the anterior region of the maxilla and a review of the previous literature.

CASE REPORT

A 41-year old male patient presented at a physician's private office with the chief complaint of the lingual expansion of the maxilla for one year and a rapid growth in the last three months. There was no tenderness on the palpation and the covering mucosa was intact. General physical status was normal and there was no problem in the past medical history.

Received: 25.04.2014 Accepted: 12.05.2014

Radiographically a unilocular radiolucent lesion with corticated borders extending from maxillary left central incisor to the right second premolar, crossing the midline, without associating any impacted tooth was seen (Figure 1). On the cone beam computed tomography (CBCT) images, lingual expansion of the mentioned area was remarkable (Figure 2). A clinical diagnosis of calcifying odontogenic cyst and KCOT was established.

The lesion was enucleated under general anesthesia and sent for the histopathologic examination. Gross examination showed a cystic lesion with elastic consistency and creamy brown colour, measuring $5.3 \times 3.7 \times 1.8$ cm. In the microscopic evaluation a cystic lesion lined by orthokeratinized stratified squamous epithelium with prominent granular layer subjacent to the orthokeratin layer was seen. The basal layer showed little tendency of palisading. In the underlying fibrotic connective tissue wall, scattered chronic inflammatory cells infiltration and foci of hemorrhage was evident (Figure 3A,B). According to these features the diagnosis of orthokeratinized odontogenic cyst was made. After the surgery the patient was followed up. No sign of recurrence was found during one year (Figure 4).

DISCUSSION

OOC is an uncommon developmental odontogenic cyst and constitutes about 5.2% to 16.8% of the cases that had been previously diagnosed as odontogenic keratocyst in different case series (5). The lesion makes up approximately

⁽Turk Patoloji Derg 2017, 33:81-85)

Correspondence: Nasim TAGHAVI

Shahid Beheshti University of Medical Sciences, Department of Oral Pathology, TEHRAN, IRAN

E-mail: nasim.taghavi@yahoo.com Phone: +98 912 181 53 87

11% of all the cystic lesions of the jaws(8). MacDonald et al. in a systematic review on different population and Li et al. showed that males are more frequently affected than females just like our case. This finding is in contrast to the Dong et al. findings (4-6).

Table I shows cases of OOC reported in the English case series. According to the previous studies OOC occurs

mostly in the third and fourth decades of life (4, 5); in line with these finding, in the present case, the patient's age was 41. Moreover, MacDonald et al. stated that there is preponderance for female in the second decade of life. Because of the coincidence of menarche in this decade, it might be related to the hormonal element to the occurrence of the lesion in the second decade of life in women (2).



Figure 1: Panoramic view of the lesion shows a unilocular radiolucent lesion with corticated borders extending from maxillary left central incisor to the right second premolar.



Figure 2: CBCT shows lingual expansion in the anterior segment of the Maxilla.



Figure 3: A) Orthokeratinized stratified squamous epithelial lining with prominent granular cell layer (H&E; x400), **B)** The underlying fibrous connective tissue with scattered chronic inflammatory cell infiltration (H&E; x40).



Figure 4: Panoramic follow up imaging shows no sign of recurrence.

The posterior segment of the mandible and the ramus region has been reported as the most common sites of involvement (4-6), but this case, was located in the anterior region of the maxilla with crossing the midline and extension to the other side. Dong et al. reported the occurrence rate of OOC in the anterior maxilla less than 5% (5).

OOCs are usually asymptomatic and a slow growing swelling is the most frequent presenting symptom and is occasionally accompanied by pain (6). The rapid growth observed in our case is not a common sign. A few case studies showed that OOCs affecting East Asian groups might be more aggressive and show different signs like paresthesia (6).

Radiographically, OOC usually appears as a well-defined solitary unilocular or multilocular radiolucent lesion (4-6).

46.7% to 68% of OCCs are associated with an unerupted tooth (5, 6) and are generally diagnosed as a dentigerous cyst. The maximum diameter of the cyst ranges from 20-70 mm with the mean size of 48 mm in various articles (4,5). In the present case, the maximum diameter of the lesion was 5.3 cm.

Kasat et al. reported a case of OOC with multiple supernumerary teeth (9). Rare cases of bilateral involvement of the mandible, dysplastic changes in the epithelial lining of the OOC, association of the OOC with the calcifying odontogenic cyst and complex odontoma have additionally been reported in the literature (10-12). Bolbaran et al. reported a case of OOC in a patient with Nevoid Basal Cell Carcinoma Syndrome (13). The recurrence rate was about 0-4% after complete enucleation during an average period

		Gender		Location	Pain	Incidental finding	Shape	Root resorption	Tooth displacement	Swelling		Unerupted tooth	Recurrence
Author/ year	Number of cases	Male: female	Mean age	Maxilla: Mandible	yes:no	yes:no	Unilocular: multilocular	yes:no	Yes:no	Yes:no	Mean size	Yes:no	Yes:no
MacDonald (2010) (6)	192	127:65	34.89	43:104	26:82	39:43	119:9	0:5	4:3	37:53	ING	50:22	7:158
Dong (2009) (5)	61	ING*	ING*	6:55	13:48	ING*	47:7	ING*	ING*	46:61	4.8	27	0:42
MacDonald (2010) (8)	ING*	2:3	34.20	2:4	2:3	0:5	3:2	0:5	4:1	ING*	ING	3:2	ING*
Gonzalez Galvan (2013)(2)	3	2:3	33.6	0:3	1:2	1:2	2:1	0:3	1:2	2:1	ING	2:1	ING*

Table I: Orthokeratinized odontogenic cysts reported in the case series

*ING: Information not given

of 6.4 to 7.8 years of follow-up (4, 6). This recurrence rate is completely different than the 30% or more recurrence rate associated with KCOT.

Histopathologically, OOC is a cystic lesion with an orthokeratinized stratified squamous epithelium lining and a prominent granular cell layer in varying thicknesses. KCOT and other keratinized odontogenic cysts such as gingival cyst of newborn and also dermoid cyst must be considered in the histopathological differential diagnosis. KCOT shows a regular epithelium with 5-10 layers thickness with corrugation in the surface; palisading in basal layer not present in OOC (2,14,15). Gingival cyst of the newborn shows a parakeratotic flattened epithelial lining. It occurs in the gingiva of the newborn and is not an intraosseous cyst. Dermoid cyst, however, is lined by orthokeratinized stratified squamous epithelium with a granular cell layer similar to OOC but shows skin appendages in the cyst wall and often occurs in the midline of the floor of the mouth.

Immunohistochemical studies comparing KCOT and OOC have shown a higher expression of Bcl-2, Ki-67, P53 and TGF- α in KCOT than OOC. These findings suggest reduced proliferative and anti-apoptotic activity in OOC (5, 16, 17). Besides, some differences in the keratin expression in KCOT and OOC support the difference in characteristics of these two lesions (18). Zhang et al. study implied a significant difference in stromal collagen fibers of KCOT and OOC (19). Roy et al. similarly reported increased activity of the stromal myofibroblasts in KCOT compared to OOC, which may explain the more aggressive behaviour of KCOT (20).

In conclusion, OOC exhibits distinctive clinical, histopathological and biological features that vary substantially from KCOT 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 KCOT must be considered in the differential diagnosis.

REFERENCES

- 1. Wright JM. The odontogenic keratocyst: Orthokeratinized variant. Oral Surg Oral Med Oral Pathol. 1981;51:609–18.
- González Galván Mdel C, García-García A, Anitua-Aldecoa E, Martinez-Conde Llamosas R, Aguirre-Urizar JM. Orthokeratinized odontogenic cyst: A report of three clinical cases. Case Rep Dent. 2013;2013:672383.
- Philipsen HP. Keratocystic odontogenic tumour. In: Barnes L, Eveson J, Reichart P, Sidransky D. editors. WHO Classification of Tumours. Pathology and genetics of tumours of the head and neck. Lyon: International Agency for Research on Cancer (IARC); 2005. 306–7.
- Li TJ, Kitano M, Chen XM, Itoh T, Kawashima K, Sugihara K, Nozoe E, Mimura T. Orthokeratinized odontogenic cyst: A clinicopathological and immunocytochemical study of 15 cases. Histopathology. 1998;32:242-51.
- Dong Q, Pan S, Sun LS, Li TJ. Orthokeratinized odontogenic cyst: A clinicopathologic study of 61 cases. Arch Pathol Lab Med. 2010;134:271-5.
- 6. Macdonald DS. Orthokeratinized odontogenic cyst: A systematic review. Dentomaxillofac Radiol. 2010;39:455-67.
- Siar CH, Ng KH. Orthokeratinised odontogenic keratocysts in Malaysians.Br J Oral Maxillofac Surg. 1988;26:215–20.

- Macdonald DS, Li TK. Orthokeratinized Odontogenic cyst in Hong Kong community: The clinical and Radiographic features. Dentomaxillofac Radiol. 2010;39:240-5.
- Kasat VO, Saluja H, Kalburge JV, Kini Y, Nikam A, Laddha R. Multiple bilateral supernumerary mandibular premolars in a nonsyndromic patient with associated orthokeratised odontogenic cyst- A case report and review of literature. Contemp Clin Dent. 2012;3:S248-52.
- de Fátima Bernardes V1, de Lacerda JC, de Aguiar MC, Gomez RS. Calcifying odontogenic cyst associated with an orthokeratinized odontogenic cyst. Head Neck Pathol. 2008;2:324-7.
- Bolbaran V, Martinez B, Rojas R. Odontogenic keratocysts: A retrospective study of 285 cases. (I. Clinical aspects). Med Oral. 2000;5:331–7.
- Yoshida H, Onizawa K, Yusa H. Squamous cell carcinoma arising in association with an orthokeratinized odontogenic keratocyst. Report of a case. J Oral Maxillofac Surg. 1996;54:647-51.
- Bolbaran V, Martinez B, Rojas R. Odontogenic keratocysts. A retrospective study of 285 cases. (II. Histopathological aspects). Med Oral. 2000;5:338–44.
- da Silva MJ, de Sousa SO, Corrêa L, Carvalhosa AA, De Araújo VC. Immunohistochemical study of the orthokeratinized odontogenic cyst: A comparison with the odontogenic keratocyst. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2002;94:732-7.

- 15. Shear M. Odontogenic keratocyst: Clinical features. Oral Maxillofacial Surg Clin North Am. 2003;4:335-45.
- Deyhimi P, Hashemzade Z. Comparative study of TGF-alpha and P53 markers expression in odontogenic keratocyst and orthokeratinized odontogenic cyst. Dent Res J. 2012;9:S39-44.
- 17. Rangiani A, Motahhary P. Evaluation of bax and bcl-2 expression in odontogenic keratocysts and orthokeratinized odontogenic cysts: A comparison of two cysts. Oral Oncol. 2009;45:41-4.
- Aragaki T, Michi Y, Katsube K, Uzawa N, Okada N, Akashi T, Amagasa T, Yamaguchi A, Sakamoto K. Comprehensive keratin profiling reveals different histopathogenesis of keratocystic odontogenic tumor and orthokeratinized odontogenic cyst. Hum Pathol. 2010;41:1718-25.
- Zhang JY1, Dong Q, Li TJ. Differences in collagen fibers in the capsule walls of parakeratinized and orthokeratinized odontogenic cysts. Int J Oral Maxillofac Surg. 2011;40:1296-300.
- 20. Roy S, Garg V. Evaluation of stromal myofibroblasts expression in keratocystic odontogenic tumor and orthokeratinized odontogenic cysts: A comparative study. J Oral Maxillofac Pathol. 2013;17:207-11.