CASE REPORT

Odontogenic Keratocyst in the Anterior Maxilla - A Case Report

Mary Oshin, Keerthi Sai, G. Deepthi, D. Shyam Prasad Reddy
Department of Oral and Maxillofacial Pathology, Kamineni Institute of Dental Sciences, Narketpally, Telangana, India

Email for correspondence: oshin.mary19@gmail.com

ABSTRACT

Odontogenic keratocysts (OKCs), previously known as keratocystic odontogenic tumors, are benign cystic neoplasms involving the mandible or maxilla and believed to arise from the dental lamina, the primordium of developing tooth germ, or from the basal cell layer of the oral epithelium. They are locally aggressive and have a high recurrence potential rate due to its infiltrative behavior. OKCs usually are present in the second to third decades of life, most commonly occurring in the posterior body and ascending ramus of the mandible. Syndromes associated with multiple OKCs are nevoid basal cell carcinoma syndrome, Gorlin–Goltz syndrome, Marfan syndrome, Ehlers–Danlos syndrome, etc. Hereby, in this case report, a rare case of OKC, located in the anterior maxillary region, crossing the midline is presented.

Key words: Anterior maxilla, keratocystic odontogenic tumor, odontogenic keratocyst

INTRODUCTION

Odontogenic keratocyst (OKC) is a common developmental odontogenic cyst which is considered to be one of the most aggressive odontogenic cysts of the oral cavity. It is known for its rapid growth and its tendency to invade the bone.[1] The OKC is a controversial odontogenic developmental cyst that has undergone conceptual and terminological changes in recent decades. The term OKC was first coined by Philipsen in 1956.[2] Pindborg and Hansen in 1963 described the essential features of this type of cyst. It is named keratocyst because the cyst epithelium produces so much keratin that it fills the cyst lumen. Furthermore, flattening of the basement membrane and palisading of the basal epithelial cells, reminiscent of odontogenic epithelium, are characteristics of OKC.[3] In 1967, Toller suggested that the OKC may best be regarded as a benign neoplasm rather than a conventional cyst based on its clinical behavior.

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In 2005, OKC was reclassified based on several factors, such as locally destructive behavior and high recurrence rate, proliferation and budding of the basal epithelial layer into the underlying connective tissue in the form of daughter cysts, and the presence of mitotic figures found in the suprabasal layers of the lesional epithelium and also mutation of PTCH (patched) gene involved in both syndrome-associated and sporadic KOTs, which occurs on chromosome 9q22.3-q31.[2] This evidence resulted in the separation of the subtypes of OKC into two distinct diseases and the abandonment of the term "OKC" by the WHO in 2005. The parakeratinized subtype became the "keratocystic odontogenic tumor" (KCOT), which integrated the group of odontogenic epithelial tumors into its classification, while the orthokeratinized subtype continued in the group of odontogenic developmental cysts as "orthokeratinized odontogenic cyst." [3]

Recently, in 2017, the KCOT returned to the WHO classification of odontogenic developmental cysts, retaking the original terminology "OKC" as in their study concluded that solitary KCOT is more likely to be less biologically aggressive and should be classified as a cyst rather than a tumor. This means that more than half of KCOTs manifest themselves as ordinary cysts.[4]

OKC usually arises from the cell rests of the dental lamina and epidemiologically OKC accounts

for approximately 7.8% of all cysts of the jaw and incidence varies from 4% to 16.5%. It occurs at all ages with peak incidence in the second and fourth decades of life. It predominantly occurs in caucasian population with a male: female ratio of 1.6:1. Location wise, it is most commonly seen twice in the mandible as compared to the maxilla. In the mandible, it occurs usually in the angle-ascending ramus region (69–83%). The mandibular cyst crosses the midline and the maxillary cyst may involve the sinus and nasal floor, premaxilla, and maxillary third molar region.

OKC is mostly an intraosseous lesion though a peripheral counterpart also has been reported in the buccal gingival in the canine region of the mandible. Peripheral OKCs have female predominance with a male: female ratio of 2.2:1. Syndromes associated with multiple OKCs are nevoid basal cell carcinoma syndrome, Gorlin–Goltz syndrome, Marfan syndrome, Ehlers–Danlos syndrome, Noonan syndrome, orofacial digital syndrome, and Simpson–Golabi–Behmel syndrome.[5] The present case of OKC is located in the anterior maxillary region, crossing the midline which is rare and uncommon, making the clinical and radiological diagnosis difficult.

CASE REPORT

A 23-year-old male patient reported to the Department of Oral Surgery at Kamineni Institute of Dental Sciences, with a chief complaint of swelling and pus discharge in his upper left front tooth region for 1½ years. The patient was apparently asymptomatic 1½ years back and then noticed painless swelling over the upper front tooth region [Figure 1] which subsided on itself. The patient noticed pus discharge from the same region after 3 months for which he used medication and got subsided. There was no extraoral swelling. The pus discharge was continuous for 6 months.

On intraoral examination, there was a soft fluctuant lesion with a sinus tract in the maxillary vestibular region of the canine. Margins were diffused with a soft consistency. The lesion was not tender on palpation. There was no response in relation to 21, 22, 23, and 24 to heat test and electric pulp testing. Panoramic radiograph revealed a unilocular radiolucency extending from 21 to 24 with a well-demarcated border [Figure 2].

Incisional biopsy was performed and was sent for histopathological examination. The histopathological findings revealed a cystic lining of 6–8 cell thick, exhibiting corrugated parakeratinized epithelium with palisading basal cells showing hyperchromatic nuclei and reversal of polarity along with underlying connective tissue. The histopathological features were suggestive of OKC [Figures 3 and 4].

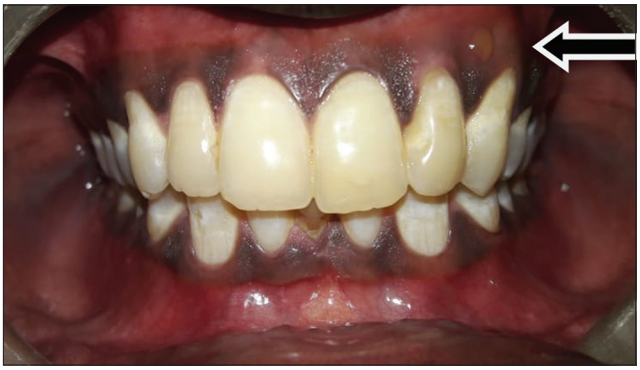

Figure 1: Clinical picture showing a soft fluctuant swelling in relation to 22 and 23

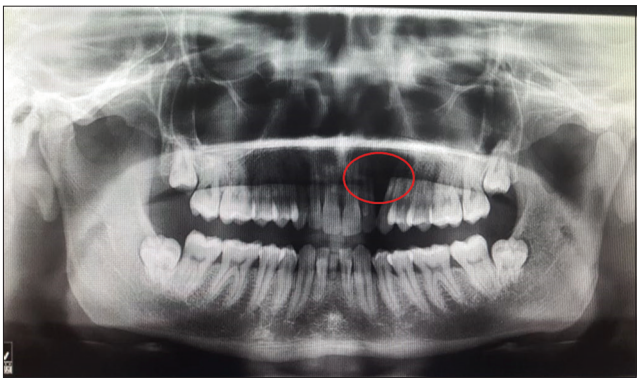

Figure 2: Orthopantomogram showing diffuse radiolucency in relation to 22 and 24

DISCUSSION

OKC is a developmental odontogenic cyst, with a biological behavior similar to a benign neoplasm. OKC was named as primordial cyst by Robinson in 1945 and as OKC by Philisen in 1956. The characteristic marks of OKC were first represented by Pindborg and Hansen in 1963. The term OKC has been reclassified in the 2005 edition of the WHO Classification of Head and Neck Tumors, from cystic to neoplastic lesions, and named KCOT because of its aggressive behavior, and PTCH1 gene mutation or inactivation. In 2017, the WHO Classification of Head and Neck Pathology reclassified OKC back into the cystic category.[3,5]

OKC may be derived from the dental lamina or its remnants, the primordium of developing

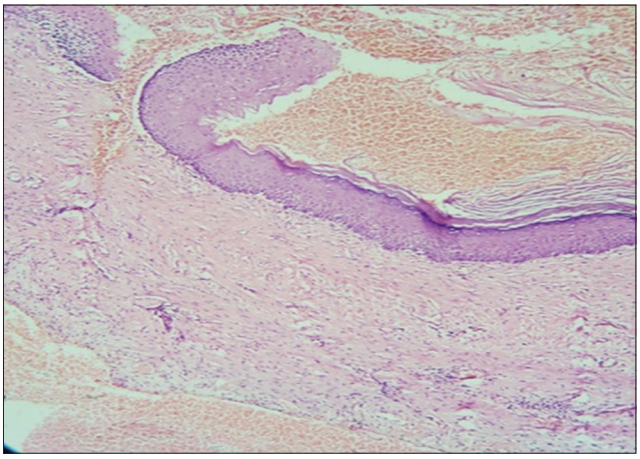

Figure 3: H- and E-stained section showing the cystic lining epithelium of 6–8-layer thickness and underlying connective tissue stroma (40)

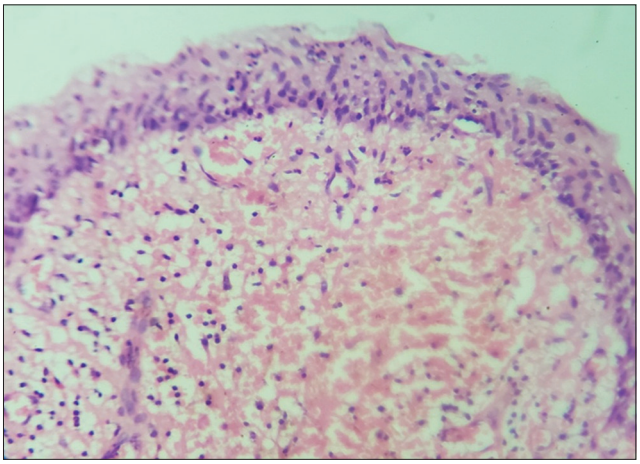

Figure 4: H- and E-stained section showing the palisading basal cells with hyperchromatic nuclei and surface corrugations in the cystic lining epithelium

tooth germ, or from the basal cell layer of the oral epithelium. It is mostly believed that OKC develops due to cystic degeneration of the stellate reticulum in developing tooth. Etiopathogenesis of OKC involves a high proliferation rate, which shows a significantly greater expression of proliferating cell nuclear antigen, Ki-67, overexpression of (the antiapoptotic protein) Bcl-2, MMP-2, and MMP-9. OKC is believed to grow due to active cellular proliferation and has a destructive growth manner through bone trabeculae in anteroposterior orientation. OKC always expands through cancellous bone, so noticeable bone expansion is seen only in later stages; this distinguishes OKCs from other jaw cysts The rate of growth of keratocysts varies 2–14 mm a year, with an average of 7 mm.[5-8]

OKCs constitute about 7.8% of whole jawbone cysts, with incidence varying between 4% and 16.5%. It involves a wide range of age groups, with a peak occurrence in the second and fourth decades of life. The white population are commonly involved with a male: female ratio of 1.6:1. OKCs are found twice in the lower jaw than the upper jaw. These cysts usually occur in the angle-ascending ramus area (69%–83%). While the upper jaw OKCs may extend to the maxillary sinus, nasal floor, premaxilla, and maxillary third molar area.[9]

OKCs of children are usually associated with basal cell nevoid syndrome (BCNS) inherited as an autosomal dominant trait and manifests as multiple cysts. BCNS can result from mutations in the PTCH1 gene on chromosome 9q22, the PTCH2 gene on 1p32, or the SUFU gene on 10q24-q25. All of these mutations affect the Sonic Hh pathway. BCNS is characterized by multiple basal cell carcinomas of the skin, jaw cysts, rib anomalies, vertebral anomalies, and intracranial calcifications.[9]

OKCs are commonly asymptomatic and discovered during routine imaging procedures (orthopantomography) as they grow slowly in an anteroposterior pattern because of which they are large in size without any noticeable bony swelling. Small OKCs are usually asymptomatic, but larger ones may show clinical manifestations such as pain, swelling, or drainage.[10]

Radiologically, OKCs demonstrate a well-defined radiolucent area with smooth and often corticated margins and may be unilocular or multilocular. Tooth displacement is commonly seen rather than root resorption. Most lesions, however, are unilocular, and 40% appear in association with a crown of an impacted tooth "like a dentigerous cyst." About 30% of upper and 50% of lower OKCs induce buccal cortical plate enlargement. Mandibular lingual expansion is present sometimes. The radiographic findings, although often highly suggestive, are not diagnostic.[11]

OKCs are usually an intra-bony disorder; however, peripheral (extra-bony) cysts have been found in the mandibular canine area, with a male: female ratio of 2.2:1.[5] Peripheral OKC shows saucerization of the underlying bone.[11]

The cystic lumen may contain clear liquid, similar to transudate of serum, or it may contain a cheesy material that, on microscopic examination, consists of keratinaceous debris. Cytologically,

OKC's aspiration is mostly cellular, mainly composed of clusters of keratinized cells with no nuclei. The parakeratinized cells had small pyknotic nuclei and showed less tendency to form groups in a background that contained granular debris.[9]

The diagnosis of OKC is based on the histopathologic features. Microscopically, OKCs exhibit thin epithelium (6–10 cell layers), corrugated (rippled) parakeratotic lining on its luminal surface, and palisading columnar/cuboidal basilar cells with hyperchromatic nuclei with reversal of polarity. Commonly, they exhibit focal separation of the epithelial lining from the adjacent connective tissue and keratin flakes might be present in the cystic cavity. OKC also exhibits characteristic features like microcyst or "daughter cysts" formation , epithelial budding at the basal cell layer and remnants of the dental lamina (odontogenic rests).[9]

The present case manifests itself in the anterior maxillary region, crossing the midline which is uncommon. It was asymptomatic for 1½ years, slowly growing in the anteroposterior direction, and was diagnosed with radiographic and histologic findings. The occurrence in rare location makes this lesion exceptional. Through examination of clinical, radiographic, and histopathological features is mandatory for accurate diagnosis of OKC.

Therapeutic interventions of OKC include marsupialization and enucleation, combined with adjuvant cryotherapy with Carnoy's solution, and marginal or radical resection. OKCs are characterized by high tendency to post-operative recurrence (30–60%). Causes of high recurrence rates include incomplete removal, remnants of the dental lamina, and the presence of daughter/satellite cysts within the cyst wall. Because recurrence may be long delayed in this lesion, follow-up of any case of OKC with annual radiographs is essential for at least 5 years after the surgery.[12]

CONCLUSION

OKC is an aggressive lesion with high recurrence rate. There have always been controversies regarding the cystic or the lesion's neoplastic behavior. The use of fine-needle aspiration and incisional biopsy may help in the early diagnosis of OKCs and perform more conservative treatment for those lesions without teeth involvement and cortical bone perforation and prevent aggressive surgical plan for OKCs.[9] OKCs located in unusual locations need to be carefully investigated radiographically and histopathologically to prevent misdiagnosis.

REFERENCES

1. Brannon RB. The odontogenic keratocyst. A clinicopathologic study of 312 cases. Part I. Clinical features. Oral Surg Oral Med Oral Path 1976;42:54-72.
2. Sivapathasundaram B. Shafer's Textbook of Oral Pathology. 8th ed. Elsevier Health Sciences. 2016
3. Ribeiro-Junior O, Borba AM, Alves CA, De Gouveia MM, Deboni MC, Naclério-Homem MD. Reclassification and treatment of odontogenic keratocysts: A cohort study. Braz Oral Res 2017;31:e98.
4. Singh HP. Need to reclassify keratocystic odontogenic tumor into cyst and neoplasm. Natl J Maxillofacial Surg 2016;7:111.
5. Passi D, Singhal D, Singh M, Mishra V, Panwar Y, Sahni A. Odontogenic keratocyst (OKC) or keratocystic odontogenic tumor (KCOT). Journey of OKC from cyst to tumor to cyst Again: Comprehensive review with recent updates on who classification. Int J Curr Res 2017;9:54080-6.
6. Almache ME, Calle MF, Gavilanes MP, Yaguana DV, Campoverde JD. Large dimension odontogenic keratocysts: Case report. Res Soc Dev 2021;9:43091211426.
7. Regezi J, Sciubba J, Jordan R. Oral Pathology: Clinical Pathology Correlations. 7th ed. USA: Elsevier Saunder; 2017. p. 254-7.
8. Shear M, Speight PM. Cysts of the Oral and Maxillofacial Regions. 4th ed. United States: Wiley; 2008.
9. Hamied MA, Al-Shaikhani SM, Ali ZD. Odontogenic keratocyst (a literature review). Al-Kindy Coll Med J 2021;17:52-61.
10. Vallejo-Rosero KA, Camolesi GV, De Sá PL, Bernaola Paredes WE. Conservative management of odontogenic keratocyst with long-term 5-year follow-up: Case report and literature review. Int J Surg Case Rep 2020;66:8-15.
11. Essaket S, Benjelloun L, Chbicheb S. Odontogenic keratocyst mimicking a radicular cyst. Integr J Med Sci 2021;8:1-4.
12. Woo SB, Eisenbud L, Kleiman M, Assael N. Odontogenic keratocysts in the anterior maxilla: Report of two cases, one simulating a nasopalatine cyst. Oral Surg Oral Med Oral Path 1987;64:463-5.