A radiolucent lesion crossing the midline in maxilla: a rare presentation of odontogenic keratocyst in young patient

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Abstract Odontogenic Keratocyst (OKC) is a developmental non-inflammatory odontogenic cyst which is proposed to be arising from cell rests of dental lamina. Among the jaw cysts OKCs account for third most common following radicular and dentigerous cyst. Most of the studies have stated that posterior part of the mandible is the most common site, but there are inconsistencies regarding the prominent location of OKCs in the maxilla. Very few studies and cases are reported with OKCs crossing maxillary midline. If do occur they are in older individuals. According to WHO reclassification, this cyst is considered as Keratocystic Odontogenic Tumour (KCOT) because of its neoplastic nature. This article describes a rare site of occurrence of this lesion especially in young patient.

Keywords OKC · Maxillary midline · Young patient

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Introduction

The OKC is a distinctive histopathologic type of developmental odontogenic cyst. It was first described by Philipsen in 1956 [1]. The first case of OKC was presented by Mikulicz as 'dermoid cyst' [2]. OKCs are known for their peculiar behavior and varied origin, debated development, unique tendency to recur and disputed treatment modalities. These cysts are generally thought to be derived from either the epithelial remnants of the tooth germ or the basal cell layer of the surface epithelium [2]. The OKC is now designated by the WHO as KCOT and is defined as 'a benign uni- or multicystic, intraosseous tumour of odontogenic origin, with a characteristic lining of parakeratinized stratified squamous epithelium and potential for aggressive, infiltrative behaviour.' WHO recommends the term KCOT as it better reflects its neoplastic nature. Other synonyms include odontogenic keratocystoma and primordial cyst [3,4].

It is estimated that the OKC makes up 10% to 12% of all developmental odontogenic cysts. Recognition of its

characteristic microscopic features is important because of its high recurrence rate, which has been estimated at around 30% [1]. In addition, the OKC is the specific type of odontogenic cyst that may be associated with the Nevoid Basal Cell Carcinoma Syndrome (NBCCS) [1,3,4].

When the age of occurrence is considered, the majority of patients are in second and third decade of life, but cases ranging from 5 to 80 years have been reported with a mean age of 70 years [1,2]. Approximately 65% of OKCs occur in mandible with a predilection for the molar/ramus region [1]. But there are inconsistencies regarding the prominent location of OKCs in the maxilla [5]. Very few studies and cases are reported with OKCs crossing maxillary midline [6–8]. If do occur they are in older individuals [6]. Frequently, the lesion is associated with an impacted tooth and clinically it mimics a dentigerous cyst. However, OKC can occur anywhere within the jaws and can resemble other lesions [1,9]. Malignant changes in the lining of OKCs have been reported but it has been suggested that it occurs more frequently in lesions with keratinizing linings [10–12].

In this report we describe an unusual case of odontogenic keratocyst with impacted supernumerary tooth in the maxillary anterior segment crossing midline in a patient aged around 30 years.

Case report

A 32-year-old male patient reported to the Department of Oral and Maxillofacial Surgery with the complaint of pain and swelling on upper left side of the face since one and half months. Patient gave the history of mild continuous nonradiating pain in the upper anterior tooth region for past one year. Pain was aggravated by chewing and relieved by medications. On systematic clinical examination a diffused swelling was evident extending from left ala of nose to left zygomatic prominence mediolaterally and from the upper border of ala to upper lip obliterating the nasolabial fold (Fig. 1). A thorough clinical examination revealed an intra-oral draining sinus in relation to maxillary left second premolar region (Fig. 2). On palpation the swelling was soft in consistency indicating

buccal cortical plate resorption. Radiological examination depicted an unilocular radiolucency extending from root apex of upper right canine to mesial root of upper left first molar. There was an inverted conical supernumerary tooth between left central and lateral incisors. Root of left canine was deflected distally (Fig. 3). A tentative diagnosis of infected dentigerous cyst was made.

The surgical enucleation of the lesion was planned under local anesthesia. But considering the fragile cystic lining and cheesy contents of the cystic cavity, prophylactic chemical cauterization was done using Carnoy's solution. A medicated gauze pack was kept inside the bony cavity with one end out through left nostril. This gauze pack was removed after 2 days. Enucleated tissue was sent for histopathological examination. Histopathological report showed stratified squamous epithelial lining, typical 'tomb stone' pattern of basal epithelium, numerous hyaline bodies surrounded by lymphocytes, plasma cells and macrophages which were densely dispersed in stroma along with satellite cysts and hemorrhagic areas (Fig. 4). These findings were suggestive of infected odontogenic keratocyst.

The healing was satisfactory and the patient was followed up for one year with no evidence of recurrence. The latest radiographic examination revealed normal bony pattern.

Discussion

Jaw cysts are very common due to the presence of odontogenic epithelial remnants. Among the jaw cysts OKC is the third most common [1,5]. The WHO working group recommends the term KCOT because of its aggressive and infiltrative nature [3,4]. Multiple OKCs are frequently associated with NBCCS [8]. In addition to multiple OKCs, NBCCS is also characterized by multiple basal cell carcinomas, bifid ribs, calcifications of falx cerebri, frontal bossing, multiple epidermoid cysts, medulloblastoma etc [1,4]. It has been suggested that daughter cysts are more likely to be seen in OKCs associated with NBCCS. Daughter cysts, thin friable cystic lining and various other explanations are given for high recurrence of OKCs [8,12].

The mean age of occurrence of OKC is around 37 year 9 months with the peak prevalence in the second and third decades

Fig. 1 Extra-oral appearance of patient

Fig. 3 Radiographical appearance of the lesion

Fig. 2 Intra-oral draining sinus in relation to 25

Fig. 4 Histological appearance of the lesion

of life. OKC also manifests with bimodal distribution, with an initial peak from 15 to 45 years and second smaller peak from 55 to 65 years [1]. Radiographically OKC appears as well defined radiolucency which may be unilocular or multilocular. Most of the studies have shown that mandible is more commonly involved than maxilla. In addition the posterior part of the mandible is the commonest location [1,8]. But there are inconsistencies regarding the predominant location of the OKCs in the maxilla [5].

Neville et al. after studying 18 cases of OKCs of maxillary midline concluded that it is important to include OKC in the differential diagnosis of anterior midline maxillary radiolucency, especially when they occur in older individuals [1]. Since this case being an OKC occurring in the maxillary midline of the younger patient, the young age cannot be excluded from the same. Its presentation with impacted supernumerary tooth, location and draining sinus made us provisionally diagnose the case as infected dentigerous cyst. Hence it is important for the clinician to consider OKC in the differential diagnosis of lesions crossing the maxillary midline, when they occur in a younger patient also. WHO's reclassification of this lesion from cyst to tumour underscores its aggressive nature and should motivate the clinician to manage the disease in a correspondingly aggressive manner.

Conclusion

Although OKC is the third most common cyst of jaw bones, there are very few reports with the involvement of maxillary midline. The OKC occurs commonly in the mandibular posterior region, minimally in maxilla and rarely crossing maxillary midline. In spite of its large age range of incidence, the lesions crossing maxillary midline have been reported specially in older individuals. Exact diagnosis is necessary to carryout proper treatment procedure to prevent recurrence. Hence one should also consider OKC in the differential diagnosis of a radiolucent lesion crossing maxillary midline in a younger patient.

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