

## CASE REPORT

# Orthokeratinizing odontogenic cyst of maxilla with complex odontoma

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**ABSTRACT**

The orthokeratinized odontogenic cyst (OOC) and odontoma are the odontogenic cyst and tumor respectively that are minimally invasive neoplasms of head and neck region. OOC is a rare variant of odontogenic cyst characterized by the presence of excessive orthokeratin covering the cystic lining. Odontoma is a benign neoplasm/hamartoma often discovered accidentally on panoramic radiographs. We came across a case of a 26-year-old male with swelling on his face along with difficulty in breathing. On the basis of radiographic and histopathological findings the final diagnosis of OOC associated with odontoma was given. However, there is no report in the English literature of the simultaneous occurrence of these two lesions and hence this case is very rare. It is unclear whether the two lesions were just coincidental or were actually related to each other.

**Key words:** Odontogenic cyst, odontoma, orthokeratinized odontogenic cyst

**INTRODUCTION**

The orthokeratinized odontogenic cyst (OOC) was recognized as a variant of odontogenic keratocyst (OKC) based on the presence of abundant orthokeratin in the cystic lining.<sup>[1,2]</sup> OKC has been renamed as “keratocystic odontogenic tumor” (KCOT), because it more appropriately reflects its local destructive and aggressive behavior.<sup>[3]</sup> Odontomas are hamartomas composed of various dental tissues like enamel, dentin, cementum, and sometimes pulp. They are slow growing benign tumors showing nonaggressive behavior.<sup>[4]</sup> This is the first case report of OOC of maxilla associated with odontoma reported in the English literature.

**CASE REPORT**

A 26-year-old male reported to the Dental Department, with the chief complaint of swelling and difficulty in breathing for about 5 months. The extraoral clinical examination revealed a diffuse swelling on the left side of the face obliterating the nasolabial fold. The swelling was associated with slight pain and continuous headache. Patient also complained

of continuous tear discharge from the left eye. Computed tomography scan revealed a soft tissue lesion completely obliterating the left maxillary sinus and extending up to the nasal cavity; right maxillary sinus was normal. The swelling extended anteriorly up to external nasal orifices and posteriorly to the nasopharynx. The nasal septum showed deviation to the right side. Left ethmoid sinus was also found to be involved. The (orthopantomogram) showed similar radiolucency of the lesion which was interrupted by some flecks of radiopacities [Figures 1 and 2]. Also, both the maxillary canines were impacted with the left maxillary permanent canine involved with the lesion and were closely associated with the small flecks of radioopacity.

The presence of keratin and cholesterol crystals in the fine needle aspiration cytology report directed toward the cystic lesion having keratin in the lumen. A provisional diagnosis of OKC was arrived upon. Complete enucleation of the lesion was carried out under general anesthesia, which was followed by extraction of all the left maxillary teeth. Along with the lesion frontoethmoid sinus was also excised as the lesion had involved the same. After completion of the surgical procedure the entire cavity was completely cleaned and was packed with Gel foam. Prosthesis (obturator) was made from the presurgical records and was delivered to the patient after surgery. Following the surgery, the healing of the surgical wound was uneventful and the patient was kept under observation to check for recurrence.

The histopathological examination revealed the cystic space which was lined by a continuous layer of stratified epithelium

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DOI:  
10.4103/0973-029X.125230

of 6-8 cell layer thickness with prominent basal cell layer and the absence of rete ridges. The stratified epithelium was orthokeratinized in nature with prominent stratum granulosum layer. Abundance of orthokeratin was present in the cystic lumen. The connective tissue showed minimal or no inflammatory response. One portion of the lesion was hard and solid which was processed by decalcification. The decalcified section showed presence of dentine and pulp like tissue arranged in an irregular pattern. Enamel spaces were seen in some areas. On the basis of thorough histopathological examination, the final diagnosis of orthokeratinizing odontogenic cyst associated with complex odontoma was given [Figures 3 and 4].

**DISCUSSION**

The prevalence of orthokeratinizing odontogenic cyst is very low and so the information regarding this variant is still limited. OOCs are difficult to diagnose due to lack of specific clinical and radiographic characters.<sup>[2]</sup> A total of 48% of OOCs were discovered as incidental findings, 41% first presented with swellings and 24% first presented with pain.<sup>[1]</sup> A predilection for the mandible, particularly the posterior sextants (68%) has

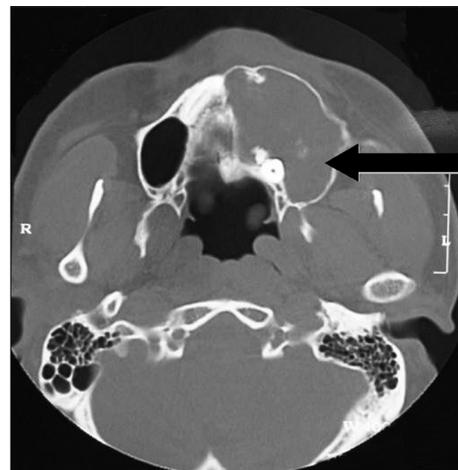
been reported in the literature. The association with unerupted teeth suggests that many OOCs may have first developed during adolescence, when the third molars were developing, and were only noticed later either owing to the development of symptoms or as an incidental discovery during investigation of another dental problem.<sup>[1]</sup>

In the present case, the radiographic and clinical presentation of the patient suggested an invasive and aggressive lesion. Final diagnosis of OOC with complex odontoma was given on the basis of histopathological examination. Therefore, the enucleation of the lesion followed by debridement of the surgical defect was considered to be sufficient.

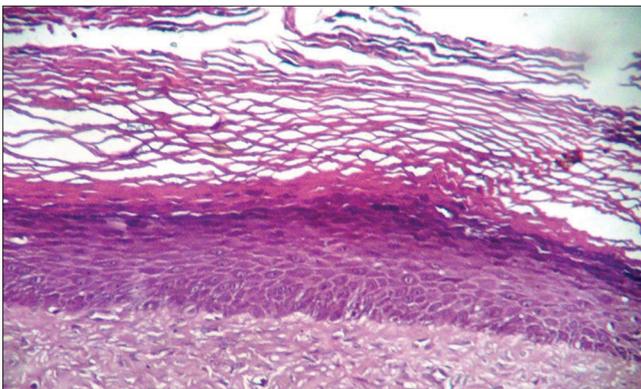
The term OKC was first introduced by Philipsen.<sup>[5]</sup> OKC is an aggressive cystic lesion that has a tendency to recur if not adequately removed. The recurrence rates have been documented as a wide range varying from 3% to 60% (Shear, 1992). While the study by Crowley, Kaugars and Gunsolle suggested that recurrence rates of the OKC and OOC were 42.6% and 2.2%, respectively.<sup>[6]</sup>



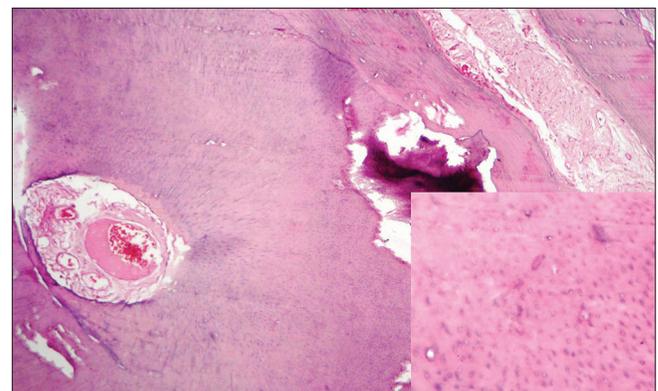
**Figure 1:** The orthopantomogram showing thinning of the maxillary sinus base of the left side, the extension of the lesion is till alveolar bone and teeth of the left side. Right maxillary sinus seems to be normal. Flecks of radiopacities are also seen



**Figure 2:** Computed tomography scan shows soft tissue tumor completely obliterating the left maxillary sinus and extending up to the nasal cavity



**Figure 3:** The photomicrograph shows cystic lumen lined by a continuous layer of orthokeratinized stratified squamous epithelium of 6-8 layer thickness with the absence of rete ridges. The lumen is extensively filled with orthokeratin (H&E stain, ×100)



**Figure 4:** The photomicrograph shows dentine and pulp like tissue arranged in a haphazard and irregular pattern (H&E stain, ×100). Inset: Dental tubules (H&E stain, ×400)

Histological examination demonstrates several striking differences between the epithelial lining of orthokeratinized and parakeratinized cysts. The typical KCOT exhibits a highly cellular parakeratinized epithelial lining with surface corrugations and a palisaded layer of basal cells while the OOC lacks these features. Instead, the thin, uniform, orthokeratinized lining epithelium is characterized by onion-skin-like luminal surface keratinization, prominent stratum granulosum and low cuboidal or flattened basal cell layer with little tendency of nuclear palisading.<sup>[5,7]</sup>

Immunocytochemical results demonstrate significantly fewer Ki-67-positive proliferating cells in the epithelial linings of OOC against the cystic lining of KCOT. The high, predominantly suprabasal proliferative activity of the KCOT lining is usually not seen in OOC. This finding suggests that there is a difference in the proliferation and maturation of the two types of epithelia. Epithelial lining of OOC shows a higher degree of squamous differentiation and exhibits low degree of cellular activity than KCOT.<sup>[5,8]</sup>

The *p63*, a member of *p53* tumor suppressor gene family, plays a major role in the maintenance of epithelial stem cells, as well as in their terminal differentiation. In the absence of *p63*, stem cells and their progenies die by apoptosis; and the crippled stem cells are unable to bolster cell proliferation and self-renewal. Some studies have demonstrated that *p63* expression in OOCs was significantly less intensive in comparison with KCOTs, indicating epithelial cells in OOCs may possess a lower proliferative and self-renewal potential. These findings thus appear to reflect the variations in epithelial cell maturation and proliferation. OOC seems to assume a different cell differentiation and exhibits a lower cellular activity than KCOT.<sup>[7]</sup>

One of the consistent findings with KCOT/OKC is the positive expression of Epithelial Membrane Antigen (EMA) and Carcino Embryonic Antigen (CEA) which is in contrast with OOC lining where it is completely absent. The 36 kDa cell surface glycoprotein, considered to be a marker of the basal cell carcinoma, is occasionally found in the KCOT but is completely absent OOC.<sup>[5]</sup>

Therefore, OOC exhibits a number of distinctive clinical, pathologic and behavioral features which vary substantially from KCOTs. It appears to represent an uncommon but consistent group of odontogenic developmental cysts that cannot be classified as other established cyst types and should therefore constitute a separate clinical entity.<sup>[2]</sup>

Odontogenic tumors originate from the tissues associated with tooth formation and reproduce to a major or minor extent, the inductive relationship between the various components of tooth germ. Odontogenic tumors constitute a diverse group of lesions because of different degrees of intertissue interaction and various growth patterns.<sup>[9]</sup>

The most common odontogenic tumors are odontomas. In 1946, Thoma and Goldman classified odontomas as follows:<sup>[10]</sup>

- Geminated composite odontomas: Two or more, more or less well-developed teeth fused together
- Compound composite odontomas: Made up of more or less rudimentary teeth
- Complex composite odontomas: Calcified structure bearing no great resemblance to the normal anatomical arrangement of dental tissues
- Dilated odontomas: The crown or root part of tooth shows marked enlargement
- Cystic odontomas: An odontome that is normally encapsulated by fibrous connective tissue in a cyst or in the wall of a cyst.

According to World Health Organization classification, odontomas can be divided into three groups.<sup>[10]</sup>

- Complex odontoma: When the calcified dental tissues are simply arranged in an irregular mass bearing no morphologic similarity to rudimentary teeth
- Compound odontoma: Composed of all odontogenic tissues in an orderly pattern, which result in many teeth-like structures, but without morphologic resemblance to normal teeth
- Ameloblastic fibroodontoma: Consists of varying amounts of calcified dental tissue and dental papilla-like tissue, the latter component resembling an ameloblastic fibroma. The ameloblastic fibroodontoma is considered as an immature precursor of complex odontoma.

A new type known as hybrid odontoma is also reported by some authors.<sup>[11]</sup>

Odontomas are classified as early as 1937 by Worth<sup>[12]</sup> which is as follows:

- Epithelial odontomas arising from dental epithelium: Dental cyst, dentigerous cyst and multilocular cyst (adamantinoma)
- Composite odontomas arising from the dental epithelium and dental mesoblastic tissues: Complex, compound, geminated and dilated

Odontomas are reported to be associated with calcifying odontogenic cyst<sup>[13,14]</sup> and dentigerous cyst.<sup>[4,15,16]</sup> The development of OOC has been postulated to be from the remnants of dental lamina, whereas odontomas are considered to be arising from hyperactive dental lamina.<sup>[11,17]</sup> So the simultaneous occurrence of these lesions can be correlated with the abnormal activity of the remnants of dental lamina. But further studies are required to establish this association.

## CONCLUSION

Extensive lesions in the jaws need to be thoroughly investigated to make a distinction between their benign and malignant

nature so as to choose an appropriate surgical approach for their treatment. Due to the benign nature of both OOC and odontoma, total excision was chosen as the treatment of choice in our case. Healing was uneventful and showed excellent bone regeneration in the surgical defect. The patient has been followed up for over 3 years and has so far showed no signs of recurrence. This is a very rare case of an extensive OOC associated with an odontoma.

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**How to cite this article:** Kulkarni M, Kheur S, Agrawal T, Ingle Y. Orthokeratinizing odontogenic cyst of maxilla with complex odontoma. *J Oral Maxillofac Pathol* 2013;17:480.

**Source of Support:** Nil. **Conflict of Interest:** None declared.