



## Case Report

# Orthokeratinized Odontogenic Cyst of The Mandible: A rare case report

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**Abstract:** Orthokeratinized odontogenic cyst (OOC) is a relatively rare developmental odontogenic cyst that occurs in the jaw bones. It was initially defined by Wright in 1981 as Orthokeratinized variant of Odontogenic Keratocyst (OKC) with distinct clinicopathological features. After undergoing a few revisions, OOC was finally considered its own entity as another type of odontogenic cyst in 2010. The OOC may appear as a multilocular radiolucency at the posterior mandible that may be suggestive of ameloblastoma or OKC and can be mistakenly diagnosed clinically as a Dentigerous Cyst due to involvement of an impacted tooth. As the recurrence rate of OOC is low, the choice mode of treatment is enucleation. The purpose of this article is to present a case of OOC arising in the right angle of mandible which is associated with an impacted lower right wisdom tooth.

**Key words:** Odontogenic keratocyst, wisdom tooth, Orthokeratinized odontogenic cyst, odontogenic cyst, enucleation.

## Introduction

Orthokeratinized odontogenic cyst (OOC) is a relatively uncommon developmental odontogenic cyst of the jaw bones<sup>1,2</sup>. Wright in 1981 has identified it as the orthokeratinized variant of odontogenic keratocyst (OKC)<sup>1</sup>. However due to significant differences between these ortho and para-keratinized cystic lesions of the jaws, the World Health Organization (WHO) in 2005 classified the parakeratinized type of the cystic lesion as keratocystic odontogenic tumour (KCOT)<sup>2,3</sup>. The OOC meanwhile is considered as an independent clinical entity<sup>4</sup>.

In the recent WHO Classification of Head and Neck Tumours<sup>5</sup> Keratocystic Odontogenic Tumour (KCOT) has been reclassified as an Odontogenic Keratocyst and OOC has been recognized as an odontogenic cyst. Orthokeratinized odontogenic cyst is a distinct entity from OKC, and should not be referred as orthokeratinized variant of OKC<sup>5</sup>. OOC occurs predominantly in males and frequently affect the molar and posterior region of the mandible<sup>4,6-8</sup>. These cysts are often asymptomatic until becoming secondarily infected<sup>13</sup>. It also can grow large in size leading to cortical expansion and presents as a swelling, along with pain, although in the majority of cases, it can be incidentally detected during radiographic examination<sup>6,7</sup>. Radiographically, OOC usually appears as a well-defined unilocular or multilocular radiolucency and commonly associated with an unerupted tooth without causing root resorption<sup>8-12</sup>.

The histopathology of OCC shows a cystic cavity lined by thin (5-8 cell layers thick) and uniform stratified squamous epithelium with prominent granular cell layer and orthokeratin<sup>1,2</sup>. In comparison to odontogenic keratocyst (OKC), the keratin surface is thick and lamellated rather than corrugated. The basal cells are usually cuboidal with no palisading of nuclei. Occasionally, the epithelial lining may be non-keratinised or parakeratinised due to inflammation. Due to its less aggressive behavior and low recurrence rate compared with OKC, surgical enucleation of the lesion with the removal of involved teeth is the mainstay of the treatment<sup>6-9</sup>. Here we report a case of Orthokeratinized odontogenic cyst, occurring in the right posterior region of the mandible of a young gentleman.

## Case Report

A 26 year old man was seen in the Oral and Maxillofacial Surgery Clinic, Hospital Tuanku Fauziah, Perlis for examination and evaluation of a tender swelling in the region of right cheek for five days duration. The swelling was progressively increasing in size, associated with severe pain with restricted mouth opening. Generally, vital signs were stable. No evidence of dysphagia and shortness of breath noted. His medical history showed no associated systemic disease.

Extraoral physical examination revealed a diffused swelling over the posterior region of right cheek extending to the right submandibular region. It was firm, warm and tender on palpation however with no evidence of cutaneous fistula. No paraesthesia on the right side of lower lip was noted. Intraoral examination showed limited mouth opening and purulent discharge from the fistula located at the mesial gingiva of lower right second molar tooth. Tooth 47 was mobile and found to be non-vital. The lower right wisdom tooth was clinically missing, with the right retromolar region swollen. In addition, buccal and lingual bony expansion was present at the lower right molar region. No paraesthesia of gingiva at lower right mandibular teeth region was noted.

Panoramic radiography revealed a unilocular radiolucent lesion at the right mandibular molar region which was associated with impacted lower right third molar tooth and periradicular region of lower right second molar tooth

(Figure 1). Computed tomography scan of the mandible showed a large unilocular expanding cystic lesion measuring about 5 cm by 2.5 cm which caused thinning and perforation of right mandibular cortical plate. Impacted lower right third molar tooth was within the cystic lesion. Multiple air pockets were identified within the lesion which indicates an infected cystic lesion (Figure 2).

Based on the clinical and radiological evidence, initially patient was diagnosed as having right buccal abscess secondary to infected odontogenic cyst. Intraoral drainage of abscess was done. He was prescribed with oral antibiotics and analgesics for 5 days duration. During subsequent follow up, incisional biopsy was performed. The histopathological examination revealed fibrous tissue lined by thin (5-8 cell layers thick) orthokeratinized stratified squamous epithelium with prominent granular cell layer. (Figure 3 and 4). The basal cells are cuboidal with no palisading of nuclei with thick lamellated



Figure 1. OPG shows a unilocular lesion on the right angle of mandible, associated with impacted lower right wisdom tooth (48). No sign of root resorption of lower right second molar tooth (47).



Figure 2. Axial view of CECT scan showing the cystic lesion causing thinning and perforation of buccal and lingual cortical plates on the right side of mandible.

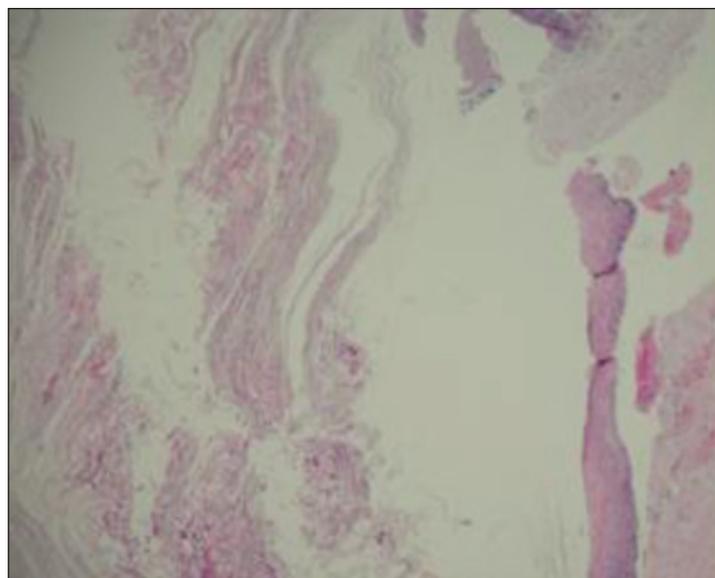


Figure 3. Histopathologic features showing thin stratified squamous epithelium lining fibrous tissue with adjacent thick lamellated keratin (Hematoxylin and eosin x20).

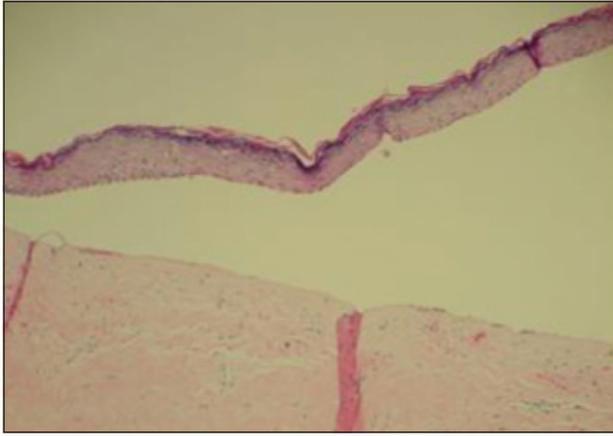


Figure 4. Histopathologic features showing thin orthokeratinized stratified squamous epithelium with prominent granular cell layer (Hematoxylin and eosin x20).



Figure 5. Surgical exposure of cystic lesion at right side of mandible.

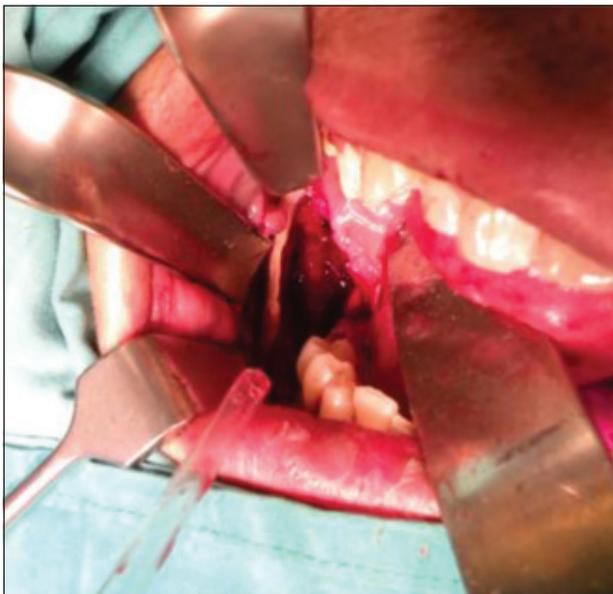


Figure 6. Post enucleation of the cyst along with removal of impacted lower right wisdom tooth (48) and lower right second molar tooth (47), showing an exposed Inferior Alveolar Nerve and thin buccal and lingual plates.

keratin seen. The histopathological interpretation was orthokeratinized odontogenic cyst of right body extending to angle of mandible.

A complete enucleation and curettage was done under general anaesthesia along with the removal of involved lower right impacted third molar tooth (48) and lower right second molar tooth (47). (Figure 5 and 6). In view of very thin buccal and lingual cortex of right angle of mandible, extra precaution was taken to prevent pathological fracture while performing the surgery. During the procedure, the lesion was found attached to the inferior alveolar nerve. As shown in Figure 6, the right inferior alveolar nerve was exposed due to extensive bucco-lingual expansion of the lesion. Once the whole lesion was removed, the cavity was thoroughly irrigated with povidone iodine 10% solution and rinsed with normal saline 0.9%. Then the cavity was packed with whitehead's varnish dressing following complete removal of the cystic lesion (Figure 7). The dressing was replaced during subsequent follow up until the cystic cavity reduced in size.

Post operatively, the patient did not have any numbness involving the right part of lower lip and chin. Mandible was firm with good dental occlusion. The specimen obtained from the enucleation was analyzed again for its histopathological figures. Based on clinical, radiological and histological features, a diagnosis of orthokeratinized odontogenic cyst (OOC) of right angle of mandible was made. Currently after more than one year follow up, no signs of recurrence was detected, as also shown in the OPG (Figure 8).

## Discussion

Historically Orthokeratinized Odontogenic Cyst (OOC) has been diagnosed as Odontogenic Keratocyst (OKC)<sup>1</sup>. However WHO has designated OOC as a distinct clinicopathological entity as it demonstrates different clinicoradiological and pathological aspects when compared to other developmental odontogenic cysts especially OKC<sup>2</sup>. OKC is highly aggressive and recurs at greater frequency than other types of odontogenic cysts. The recurrence rate ranges from 3% to 60%. Studies show that the OKC recurred in at least 42.6% of cases, compared with only 2.2% for the OOC<sup>3</sup>. Thus, it suggests the importance of distinguishing between OOC and OKC.

Orthokeratinized Odontogenic Cyst is a rare developmental cyst of the jaw bones that occurs most frequently between the third and fourth decades with a male gender predilection<sup>4,6</sup>. However it also can occur during the second decade<sup>1</sup> as reported in our case. The lesion is located mainly in the molar region and posterior mandible as reported in our case<sup>4,6-8</sup>. However, OOC can also affect other regions of the mandible and maxilla<sup>10,11</sup>. Swelling is the main clinical feature with or without pain, although in most cases described, the lesion was asymptomatic<sup>9-11</sup>. However in our case, the patient presented with painful swelling associated with secondary infection of the cystic lesion which was rarely reported<sup>13</sup>. Initially the abscess was drained intra orally and antibiotics were given before definitive treatment is performed.

As also seen in our case, the majority of OOC radiographically present as unilocular radiolucencies with 60% of cases associated with an impacted tooth along with perforation of buccal and lingual cortical plates<sup>11,12,14</sup>. Other radiolucent lesions of the jaw bones differentiated from the OOC are mainly the other odontogenic lesions namely dentigerous cyst, unicystic ameloblastoma and OKC<sup>6,9,15</sup>. The OOC may appear as a multilocular radiolucency at the posterior



Figure 7. Post-op OPG with whitehead's varnish dressing inside the cavity at right angle of mandible.



Figure 8. Post-operative OPG a year after operation showed no signs of recurrence at right angle of mandible.

mandible that may be suggestive of ameloblastoma and OKC, but as noted in our case, OOC shows no root resorption of the involved teeth, which is a common characteristic feature of ameloblastoma<sup>6,8</sup>.

Histologically, OOC shows a cystic cavity lined by a thin layer of orthokeratinized odontogenic epithelium, with prominent granular layer and low cuboidal basal cells to distinguish from OKC<sup>1</sup>. As reported in our histopathological finding, a mild mixed inflammatory infiltrate is present in the underlying fibrous wall suggestive of infected cyst. Thick keratin flecks were also present. These findings indicate OOC of right angle of mandible.

OOC involving the mandible should be treated promptly as with any other cystic lesions<sup>15</sup>. If left untreated it may lead to pathological fracture where treatment will become more complicated<sup>16</sup>. Open reduction and internal fixation with

plating might be indicated to fix the fracture site. In our case, there was no sign of mandible fracture prior to surgery. Nevertheless, extra caution was taken during the surgery to prevent mandibular fracture from happening.

Due to the less aggressive behavior of this lesion compared to OKC, enucleation and peripheral ostectomy along with the removal of involved teeth are still the mainstay of treatment<sup>7-12</sup>. In this case, the lesion was enucleated along with the removal of associated teeth. Peripheral ostectomy was also done. Before closing the soft tissue part, the operated site was packed with Whitehead's Varnish (WHV) dressing. Apart from minimizing post-operative complications such as bleeding and infection, WHV also acts to accelerate the bone healing<sup>17</sup>. In our case, the WHV dressing was changed three weekly until the cavity became smaller in size.

## Conclusion

Orthokeratinized odontogenic cyst (OOC) is a rare odontogenic cyst which shows independent clinical and pathological features. This lesion must be differentiated from the other radiolucent lesions of the jaws such as OKC and Ameloblastoma due to different prognosis and further surgical management. As the recurrence rate of OOC is much lower than OKC or other odontogenic cysts, enucleation and curettage along with removal of involved tooth are the mainstay of the treatment. Despite its low recurrence rate, regular follow up is still recommended.

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## Conflict of Interest

None declared.

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