

# ORTHOKERATINIZED ODONTOGENIC CYST: A CASE REPORT

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DOI: <https://dx.doi.org/10.4314/gdj.v20i1.9>

## ABSTRACT

Orthokeratinized odontogenic cysts (OOC) are rare developmental odontogenic cysts characterized by an ortho-keratinized stratified squamous epithelial lining. They were originally believed to be part of the spectrum of Odontogenic Keratocyst but are now considered a distinct entity.

They comprise approximately 1% of all odontogenic cysts and about 10% of cases previously coded as odontogenic keratocysts. They usually occur as a single radiolucent lesion.

In this paper, we present a case of OOC and compare them to previous case reports and a literature review.

**Keywords:** Orthokeratinized, odontogenic cysts, Odontogenic keratocyst.

## INTRODUCTION

Orthokeratinized Odontogenic Cyst (OOC) is a developmental odontogenic cyst characterized by an orthokeratinized stratified squamous epithelium<sup>1</sup>(epithelial) lining.

OOC was first described as a dermoid cyst as far back as 1927 by Schultz<sup>2</sup>. However, it was not until 1981 that Wright identified OOC as an orthokeratinized variant of Odontogenic Keratocyst owing to its different histopathology and reduced likelihood of recur<sup>3</sup>. They were originally thought to be part of the spectrum of Odontogenic Keratocyst (OKC)<sup>4</sup>.

In 2005, the World Health Organisation (WHO) re-classified the OKC as Keratocystic Odontogenic Tumour (KCOT) due to its high recurrence rate and aggressive biologic behavior. However, it stated that the orthokeratinized form was not part of the spectrum of KCOT<sup>5</sup>.

As odontogenic cysts were removed from the WHO 2005 classification, the OOC had no accepted designation. In 2017, however, the WHO re-introduced cysts into the classification and also determined insufficient evidence to call the OKC a benign neoplasm and reverted to OKC as the preferred terminology<sup>6</sup>. The OOC was included in the re-introduced section on odontogenic cysts, clearly separating it as a distinct entity from the OKC<sup>1</sup>.

OOC has a recurrence rate of less than 2%<sup>1</sup>, in comparison to a recurrence rate as high as 28% for OKCs<sup>6</sup>. OOC accounts for 10% of OKCs and therefore makes up approximately 1% of all odontogenic cysts<sup>1</sup>. Histologically, there is a definite distinction between the OOC and the OKC. The OOC is lined by uniform 4 to 6-cell thick orthokeratinized epithelium with a well-developed stratum granulosum and a flat luminal surface, unlike the OKC which is uniform 6 to 8-cell thick parakeratinized stratified epithelium. The luminal surface show corrugation. The basal cells of the OOC are low cuboidal cells compared to the tall columnar basal cells of the OKC which are palisaded<sup>1</sup>.

Whereas it is well established that about 5% of OKC may be associated with the Naevoid Basal Cell Carcinoma Syndrome (NBCCS) and may present with multiple lesions<sup>6</sup> no such association has been reported for OOC.

In this paper, we describe a case of mandibular OOC, followed by a review of previous literature

## CASE DESCRIPTION

A 43-year-old woman who was seen at the Oral and Maxillofacial Unit of the Komfo Anokye Teaching Hospital, Kumasi-Ghana, presented with a 2-year history of slow-growing painless swelling of the right mandible. There was no associated toothache or mobile teeth.

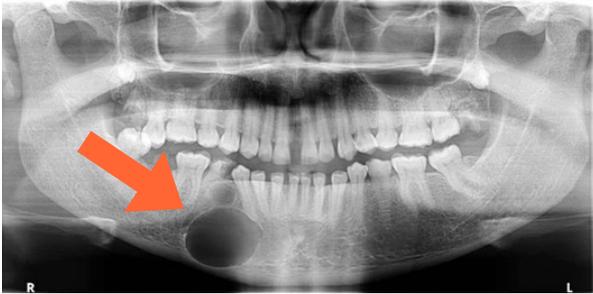
On examination, there was facial asymmetry with a right lower jaw swelling. On palpation, the mass was firm in consistency, well-circumscribed, smooth surfaced and measured about 3cm in length and 2cm in width. The swelling is continuous with the underlying bone, with no attachment to the overlying structures and no differential warmth.

Intraorally (Fig. 1), there was a well-demarcated right buccal mandibular swelling, spanning the region, from the distal of 43 to the mesial of 46. The swelling had resulted in a buccal vestibular expansion of the mandible at the affected region and was covered by normal mucosa. Sensation was intact on the right lower lip.



**Fig. 1** shows an intraoral picture of a right lower jaw swelling in the pre-molar region extending to the molar regions (indicated with the black arrow)

The panoramic radiograph (Fig. 2) showed a well-defined bilocular radiolucency, which was well-corticated. There was a mesial displacement of the roots of 44 and a distal displacement of 85, respectively, with no resorption. 85 had mesial caries. There were retained roots of 18,28,47.



**Fig. 2 Panoramic radiograph showing a well-defined bilocular radiolucency (indicated with the orange arrow) at the right lower jaw**

Thus, a provisional diagnosis of ameloblastoma was made, with odontogenic keratocyst and radicular cyst as differential diagnoses.

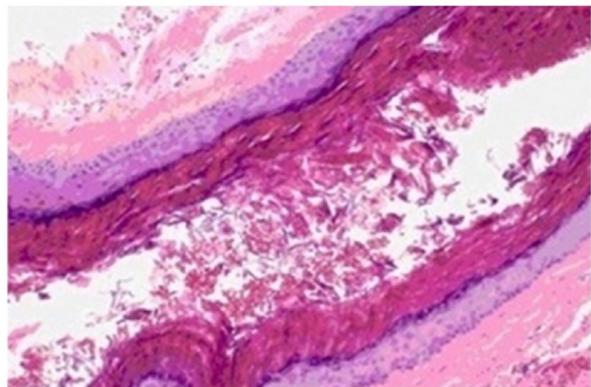
A firm cystic mass measuring 2.5cmx2cmx1cm was enucleated under General anesthesia. Gross pathology revealed a firm cystic mass measuring 2.5cm x 2cm x 1cm. (Figs. 3.1&3.2) with light tan content when cut open. Microscopic examination (Figs. 4&5) showed a non-inflamed fibrous cyst wall lined by a thin uniform ortho keratinized stratified squamous epithelium with a prominent granular cell layer. The Cystic cavity was filled with lamellated keratin flakes. No surface corrugation or basal layer cell palisading was seen. No signs of malignancy were seen.



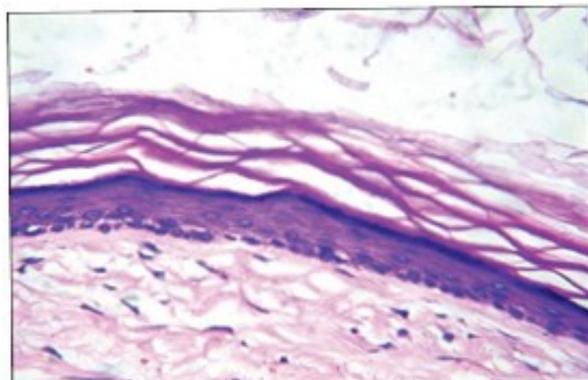
**Fig. 3.1 showing the lesion after enucleation**



**Fig.3.2 lesion cut opened after enucleation.**



**Fig. 4, shows a cyst with fibro collagenous wall and epithelial lined cavity filled with lamellated keratin flakes (Histology, H&E stain x 100)**



**Fig. 5, shows thin, uniform ortho keratinized stratified squamous epithelium with a prominent granular cell layer. No basal layer palisading or luminal corrugation is seen (Histology, H&E stain x 200)**

A diagnosis of Orthokeratinized Odontogenic Cyst was made.

The patient had no signs or symptoms to suggest NBCCS and did not meet the criteria for a diagnosis of NBCCS<sup>7</sup>.

## DISCUSSION

The case presented above highlights the diagnostic difficulty of OOCs, due to their clinical presentation being very close to several other odontogenic lesions. Based on the clinical and radiographic findings, OKC and ameloblastoma were documented as the differential diagnosis because OOC is not a common odontogenic lesion in this region.

Our patient was a female in her fourth decade. This is consistent with a systematic review by Dong et al.<sup>9</sup> which found that the largest proportion of OOC in western populations initially presented in the third to fourth.

OOC shows a male predominance<sup>1,8,9</sup>, with a study done by Dong et al.<sup>9</sup> showing a male-to-female ratio of 2.59:1 in 61 cases of OOC cases. This finding was inconsistent with our case which occurred in a female.

The mandibular presentation of OOC in our case is in agreement with studies done by Bresler et al.<sup>7</sup> who concluded that OOCs are seen in the mandible two and a half times more frequently than in the maxilla<sup>7</sup>.

Our patient reported to the hospital after a progressive enlargement of the swelling, this is consistent with cases of OOC in some literature<sup>9,10</sup>. The most common symptoms at presentation include swelling, pain or purulent discharge<sup>9,10</sup>. However, a systematic review in 2010 by MacDonald-Jankowski et al.<sup>8</sup> revealed that 48% of OOC presented as incidental findings, with a slightly lower percentage presenting with swelling (41%). As a considerable number of cases are incidental findings, a careful review of all routine radiographs, especially orthopantomographs, is indicated to assess for unexpected pathology.

The bilocular presentation of our case differs from the review by MacDonald-Jankowski et al.<sup>8</sup> showed that 93% of reported OOCs were unilocular<sup>8</sup>. In our case, the lesion showed marked expansion along with displaced, but not resorbed teeth roots. This has been reported in the literature<sup>8</sup>. This may be a helpful feature when trying to differentiate OOC from other common odontogenic tumours, such as ameloblastoma, which often show resorption of teeth

OOCs commonly present in association with an unerupted tooth<sup>4,8,10,13</sup>. The frequency with which OOCs occur with unerupted teeth has led some authors to postulate that an OOC represents a dentigerous cyst with orthokeratinization, arising from reduced enamel epithelium<sup>11</sup>. Whilst origin from dental lamina remnants is the most likely aetiology, its pathogenesis is still uncertain<sup>1</sup>.

Enucleation was the treatment of choice because the lesion was well-corticated. This is consistent with the reviews done by Nasir Uddin et al.<sup>12</sup>, Dong et al.<sup>9</sup> and case reports by Crane et al.<sup>10</sup>. Twenty months follow-up on our case have shown no recurrence.

In summary, OOC is a relatively new but rare entity, mimicking other jaw cysts including OKC and dentigerous cysts. A thorough clinical, radiographic and microscopic analysis is key in the diagnosis and treatment.

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