

CASE REPORT

## Orthokeratinized Odontogenic Cyst

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### Abstract:

Hypohidrotic ectodermal dysplasia is an uncommon disorder of tissues derived from ectoderm, characterized by the triad of hypohidrosis and hypodontia, which form the essential features of the syndrome 1. A case of a 7 year old child with hypohidrotic ectodermal dysplasia with oligodontia and marked resorption of alveolar ridges is presented. Prosthetic rehabilitation with removable acrylic prosthesis done on achieving excellent esthetics, functionality and adaptation.

**Key words:** Orthokeratinized odontogenic cyst, Odontogenic keratocyst, Keratocystic odontogenic tumour

### INTRODUCTION:

The Orthokeratinized Odontogenic Cyst (OOC) was first described by Schultz in 1927 and in 1945 Philipsen considered it to be a type of odontogenic keratocyst.<sup>1</sup> In 1981, it was Wright who specified its clinicopathological aspects and suggested that it be called odontogenic keratocyst- Orthokeratinized variant.<sup>2</sup> With the new World Health Organization classification considering the odontogenic keratocyst as a neoplasm and renaming it as keratocystic odontogenic tumour (KCOT) it becomes imperative that both clinicians and pathologists alike possess a thorough knowledge of the differences between the more aggressive KCOT and the less aggressive OOC so that patients receive the most appropriate treatment. Here, we report a case of OOC arising in the mandible of a patient.

### CASE REPORT:

A 48 year old male patient presented with a complaint of a gradually growing swelling in the

lower jaw since the past four months. Extra oral examination revealed a diffuse, nontender, firm swelling of the lower third of the face. Expansion of both buccal and lingual cortical plates was evident. All laboratory findings were found to be normal. Radiographic survey of the area revealed a well defined radiolucency in the body of the mandible extending from 37 to 45 location and extending up to the inferior border of the mandible (Fig 1). Impacted teeth were seen bilaterally in the premolar regions. Computed tomography showed an expansile osteolytic lesion and a provisional diagnosis of odontogenic keratocyst was suggested (Fig 2). Incisional biopsy of the lesion was done and microscopic examination showed a cyst lined by orthokeratinized stratified squamous epithelium which was 6-10 layers thick with a distinct granular cell layer and a cuboidal basal layer which lacked any palisading. The stratum corneum was very prominent with numerous keratin flakes seen in the lumen (Fig 3 and 4). A diagnosis of orthokeratinized odontogenic

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cyst was given. The cyst was surgically enucleated along with removal of the impacted teeth. Gross examination of the excised specimen revealed a thin cystic sac with a smooth luminal surface. The lumen also contained white cheesy material. Microscopic examination of the specimen revealed an epithelial lining similar to that seen in the incisional biopsy. The connective tissue was fibrovascular. Loss of epithelial lining was seen in areas with chronic inflammatory infiltrate. Numerous cholesterol clefts with associated multinucleated foreign body giant cells were also seen (Fig 5). Even in multiple samples, characteristic features of odontogenic keratocyst could not be found. The lesion was finally diagnosed as an orthokeratinized odontogenic cyst with inflammatory changes.

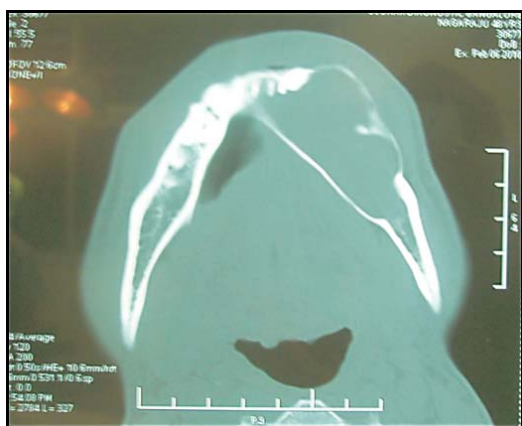


Fig 1.

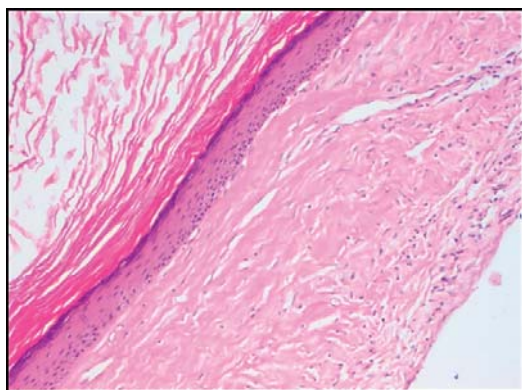


Fig 2.

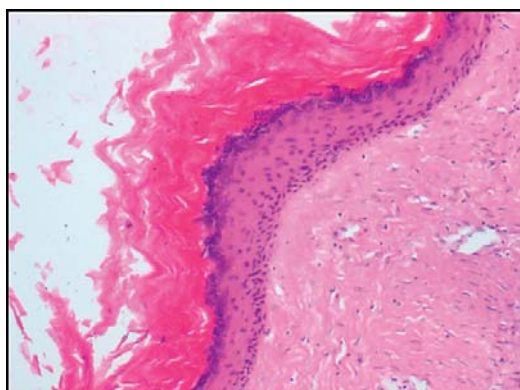


Fig 3.

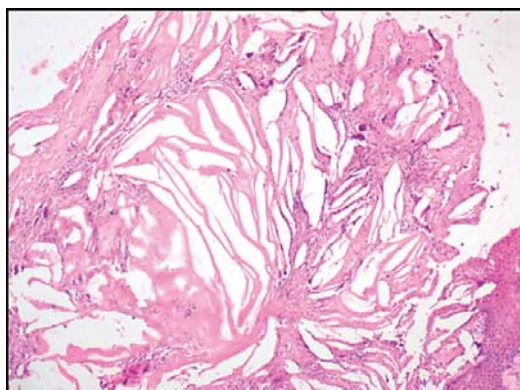


Fig 4.



Fig 5.

- Fig. 1-** Orthopentamogram showing a unilocular radiolucency and impacted supernumerary teeth.
- Fig. 2-** CT scan exhibiting expansile osteolytic lesion in the mandible
- Fig. 3-** Orthokeratinized odontogenic cyst exhibiting a uniform orthokeratinized stratified squamous epithelial lining.
- Fig. 4-** Prominent granular cell layer ,a thick keratin layer and cuboidal basal cells seen.
- Fig. 5-** Numerous cholesterol clefts along with foreign body giant cells.

## DISCUSSION:

Until recently orthokeratinized odontogenic cyst has been considered to be a type of odontogenic keratocyst. But considering the fact that this cyst is less aggressive in behaviour, has a low recurrence rate as well as possesses different histopathological features, it is now considered a separate entity.

Over the years numerous studies have been carried out to prove that these two entities are distinct from each other both clinically and histologically.

The exact incidence of OOC is not clear either because of the improper identification or classification of this entity in literature. Clinicopathological studies on odontogenic keratocysts have reported the orthokeratinized variant as ranging from 3.3% to 12.2%.<sup>3,4,5</sup>

Clinically the two entities are quite similar yet different. OOCs are generally solitary asymptomatic lesions, occurring in the third to fourth decade and with a male predilection. They occur more commonly in the mandible with an affinity for the posterior portion.<sup>6,7,8</sup> They do not occur in patients with Naevoid Basal Cell Carcinoma Syndrome (NBCCS).<sup>2</sup> KCOTs also exhibit similar findings regarding age, sex and site of occurrence but they are associated with NBCCS patients and thus tend to exhibit multiple lesions.

Radiographically OOCs tend to be unilocular lesions and are more often associated with impacted teeth as compared to KCOTs.<sup>5</sup> In our case impacted teeth were seen bilaterally.

Differences in the staining pattern of numerous immunohistochemical markers have also proven that they are not aggressive cysts. Decreased expression of Ki-67 and p63 in OOCs as compared to KCOTs indicates the low proliferative activity.<sup>6</sup> Antiapoptotic marker bcl-2 was found to be negative in the basal cell layer as against a positive expression in KCOT.<sup>9</sup> Immunoprofiling of the epithelial lining and the capsule using cytokeratins and extracellular matrix proteins revealed that the OOC was a well formed and more organized cyst as compared to OKC.<sup>1</sup> Many of such findings explain the fact why OKC is now considered a neoplasm.

Histologically the OOC is characterized by a thin, uniform epithelial lining, 4 to 8 cell layers thick and composed of orthokeratinized stratified squamous epithelium with a prominent granular cell layer as was seen in the present case.<sup>2</sup> The basal cells are usually cuboidal or flattened and do not exhibit any palisading or polarization. The KCOT however exhibits a parakeratinized epithelium with a well demarcated basal layer formed by columnar or cuboidal cells whose nuclei are polarized and palisaded. da Silva suggested that the expression of fibronectin in OKCs may explain the polarization of the basal cell nuclei and also the more aggressive behaviour.<sup>1</sup> This extracellular matrix protein was however only scantily scattered in cases of OOCs. The parakeratin squames are known to be very sparse in OKCs as compared to the more abundant leafy keratin flakes seen in OOC.<sup>10</sup>

In addition to these findings the present case also showed inflammatory changes like loss of epithelial lining, cholesterol clefts and foreign body giant cells.

The histogenesis of this cyst is still unclear. Vuhahule et al suggested that these could be dentigerous cysts with orthokeratinization or even represent central dermoid or epidermoid cyst due to similar histological features.<sup>11</sup> Zhu suggested that while the OKC may arise from the dental lamina with the presence of the dental papilla required for its development, the OOC may arise from oral epithelium under the influence of dental papilla or only the oral epithelium.<sup>12</sup>

Series of authors have reported the very low rate of recurrence of this entity and the lack of aggressiveness thus suggesting that enucleation be the mode of treatment.<sup>8</sup> But thorough sampling of the specimen is required as presence of areas of parakeratinization and polarization should lead to the lesion being diagnosed as a KCOT.

This review thus throws light on the pertinent differences between OOC and KCOT and how a thorough pathological examination of a keratinizing cyst is very important as there may be a tendency of clinical misdiagnosis which in turn could affect the prognosis of the patient.

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