



**CASE REPORT**

**MULTIPLE ODONTOGENIC KERATOCYSTS- A RARE CASE REPORT**

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**ARTICLE INFO**

**Article History:**

Received 17<sup>th</sup> November, 2017

Received in revised form 27<sup>th</sup>

December, 2017

Accepted 4<sup>th</sup> January, 2018

Published online 28<sup>th</sup> February, 2018

**Key words:**

Odontogenic Keratocyst, OKC, Cyst

**ABSTRACT**

Odontogenic keratocysts/primordial cyst is a prominent developmental odontogenic cyst arising from the dental lamina. Attributing to its aggressive growth and development into tumoral lesions, there are several debates and opinions about the varied origin and development of this cyst to the modalities of treatment. They have high rates of recurrence. This present study represents a case of odontogenic keratocysts associated with all the impacted 3rd molars and maxillary canine in a 13 year old female, who presented with a symptomatic swelling on the involved areas. The case report has aimed at ruling out any underlying syndromic signs and a programmed treatment of the cysts in order to achieve a complete healing of the lesions. The recurrence was assessed at equal intervals of time.

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**INTRODUCTION**

Phillipsen in 1956 brought the Odontogenic Keratocyst into the picture.<sup>[1]</sup> In 1962, the histologic criteria necessary to diagnose OKC was put forward by Pindborg and Hansen.<sup>[2]</sup> As the origin of the lesion was thought to be the tooth primordium, the initial terminology for an odontogenic keratocyst (OKC) was "primordial cyst." The World Health Organization (WHO) histologic typing of odontogenic tumours listed "odontogenic keratocyst" (OKC) as the preferred terminology for such cysts with a keratinized lining in 1992.<sup>[3]</sup> It has locally destructive behaviour (extension more than expansion), high recurrence rate and association with nevoid basal cell carcinoma syndrome or "Gorlin syndrome"<sup>[4, 5]</sup>. Odontogenic keratocysts accounts for approximately 3-11% of all cysts in the jaws.<sup>[8]</sup> The occurrence can be in all ages with a peaking incidence in the second and fourth decades of life.<sup>[9]</sup> The youngest patient reported was of 5 years.<sup>[10]</sup> The mean age of patients with multiple odontogenic keratocysts (OKCs), with or without Nevoid basal cell carcinoma syndrome, is less than those with single non recurrent OKCs. Odontogenic keratocysts (OKCs) are generally thought to be derived from remnants of the dental lamina (rests of Serre), traumatic implantation or down growth of the basal cell layer of the surface epithelium,<sup>[11]</sup> or reduced enamel epithelium of the dental follicle.<sup>[12]</sup> Studies have suggested a genetic cause, specifically a *PTCH* gene aberration, in the etiology of these cysts.

A para keratinized variant, an ortho keratinized variant, and combination of the two were recognized initially as the histologic variants. The ortho keratinized variant was ultimately considered as "orthokeratinized odontogenic cyst" (OOC), as it shows less aggressive clinical behaviour and recurrence pattern.<sup>[6]</sup> The histologic features of OKCs include a thin epithelial lining, usually consisting of fewer than six cell layers in a corrugated tissue composed of thin, irregular bundles of collagen, and often contain islands of epithelium that may represent daughter cysts. There will be a tendency for the epithelium to separate from the underlying cyst wall in some cases.<sup>[7]</sup> Histopathologically, they typically show a thin, friable wall, which is often difficult to enucleate from the bone in one piece, and have small satellite cysts within fibrous wall. This accounts to the high recurrence rates of OKC<sup>[11]</sup>

Cyst Contents of OKC is a dirty white, viscid suspension of keratin, which has an appearance of pus, but without an offensive smell. The smear should be stained and examined for keratin cells. It has low protein content and is mostly albumin. Total protein is found to be below 4 g/100 ml.

**CASE REPORT**

A 13 year old female patient came to the outpatient Department of Oral & Maxillofacial Surgery, Yenepoya Dental College Mangalore on 16th August 2016 with the chief complaint of swelling and pain on the right cheek. Pain was severe, continuous and throbbing in nature. The swelling was slowly enlarging in size since 3 months. The patient was well nourished and properly oriented. Extra oral Examination revealed diffused swelling over the left and right cheeks. (Figure 1) In intraoral examination, swellings over the buccal and palatal aspects in the maxillary third molar region were seen bilaterally.

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Figure 1 Extra oral Photographs

In the mandible, the buccal and lingual aspects of the third molar region were swollen bilaterally. The maxillary and mandibular third molars were missing on both sides. The left maxillary canine was also found missing. The findings were confirmed by palpation. Palpable swelling was present in the maxilla on the buccal and palatal aspects extending distal to the second molar till the maxillary tuberosity region bilaterally. In relation to the mandible, expansion of the buccal and lingual cortical plates extending from distal aspect of second molar till the retromolar trigone region bilaterally was palpated. Palpable swelling was present on the buccal aspect between the maxillary left lateral incisor and first premolar. (Figure 2)



Figure 2 Intra oral Photographs

Orthopantomograph and Computed Tomography Scans revealed radiolucency with well-defined margins with embedded teeth in the maxilla and mandible on both the sides in relation to the third molar region and left maxillary canine region. A provisional differential diagnosis of dentigerous cyst, Odontogenic Keratocyst or Ameloblastoma was made. Surgical management was planned to perform the enucleation of the cyst along with the removal of the involved tooth and application of Carnoy's Solution. (Figure 3)



Figure 3 Orthopantomograph

The patient was operated under General Anaesthesia in aseptic conditions. Cysts in relation to maxillary and mandible third molars on both sides and left maxillary canine were enucleated. The teeth associated were also removed along with lining of the cysts. (Figure 4) Chemical cauterisation was done using Carnoy's Solution. The contents of the enucleated cystic lining were sent for histopathological examination. (Figure 5)

Hemostasis was achieved and the soft tissues were closed using (3-0) Vicryl Sutures.



Figure 4 Intra Operative Photographs

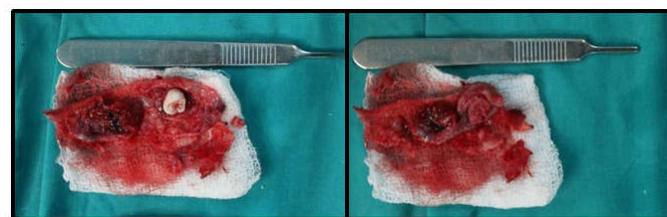


Figure 5 Specimen sent for biopsy Photographs

The Histopathological Examination of the specimen sent confirmed the lesion to be odontogenic keratocyst. The histologic evaluation revealed cystic lining comprising of corrugated Para- keratinized stratified squamous epithelium of 6 to 8 cells thickness. The epithelial cells showed palisaded layer of tall columnar basal cells exhibiting tomb stone appearance. Detachment of a portion of the cystic lining from fibrous wall was evident in few areas.

(Figure 6) These histological features of the cystic lining were characteristic of odontogenic keratocyst. Follow ups were done at intervals of 3 months, 6 months, 1 year and 1 year 4 months. No recurrence was noted.

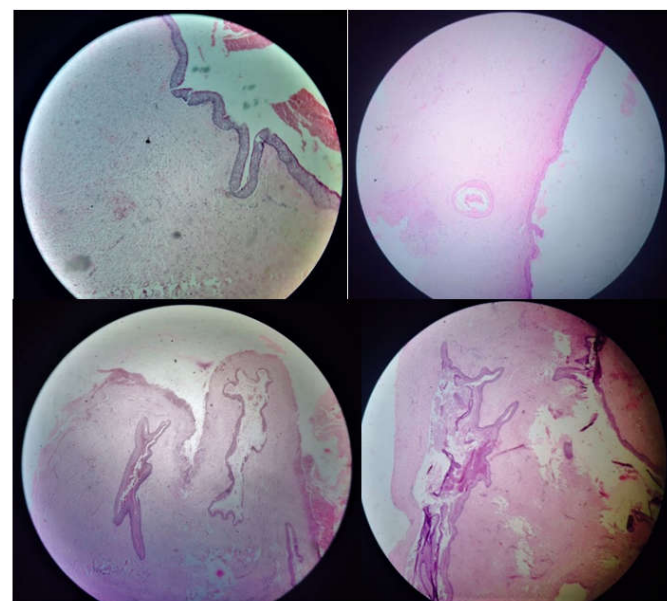


Figure 6 Histopathological Images

## DISCUSSION

Cyst formation in the jaws requires three elements: a source of epithelium, a stimulus for epithelial proliferation and the capacity for bone resorption and cyst growth. These processes are quite well understood and have been reviewed extensively elsewhere for inflammatory cysts. The OKC presents as a well demarcated corticated radiolucency in radiographs. They have scalloped margins. About 25% of OKCs are multilocular. Most of them cause tooth displacement. They often present very late and are frequently identified during the routine radiographic examination because these cysts typically expand in the antero-posterior direction.

The OKCs mostly occur in the mandible and overall about half are located at the angle and ascending ramus. Many of these may be associated with an unerupted third molar (wisdom) tooth and occasionally envelop the crown of the tooth in a dentigerous relationship.<sup>[13]</sup> Less than 1% of all cases of OKC occur in the maxilla with sinus involvement. One interesting aspect in our presented cases is that the OKCs were involvement of multiple areas in the maxilla and mandible with associated unerupted teeth.

The histological diagnosis of OKC is rarely a problem as the histology of OKC is distinctive. OKCs usually occur as solitary lesions but multiple cysts do occur and are usually associated with nevoid basal cell carcinoma syndrome (NBCCS, Gorlin Goltz syndrome). It is an autosomal dominant condition resulting in multiple odontogenic keratocysts in combination with multiple nevoid basal cell carcinomas and skeletal abnormalities. Many authors argue that the high recurrence rate, permeative growth pattern, the presence of satellite cysts and budding, all indicate "aggressive" behavior and are consistent with an eoplasm.<sup>[14]</sup> OKC originates from dental lamina remnants, as suggested by many authors. Some authors say that it should be considered a benign cystic neoplasm due to its growth capacity and development characteristics related to the mutation of a suppressor gene, PTCH, found in sporadic and in association to basal cell nevus syndrome keratocysts.<sup>[15, 16]</sup> Intracystic fluid pressure was found to be involved in OKC growth recently.<sup>[17]</sup>

Recurrence is usual in the first 5 years after the surgery, but may occur in much longer time intervals. Depending on the kind of treatment management and follow-up period, recurrence rate was found to vary from 0% to about 62%.<sup>[18]</sup> The recurrence of OKC is thought to be based on great mitotic activity and growth potential found in epithelium. Other sources of recurrences are remnants of dental lamina and epithelial islands. These findings lead to the recommendation of surgery to eradicate epithelium components of these cysts and excision of the mucosa where OKC adheres.<sup>[19]</sup>

In Surgical Management, various treatment alternatives have been suggested, such as marsupialization, enucleation, and enucleation with Carnoy's solution, enucleation with cryotherapy, curettage and resection. Simple enucleation was associated to a higher recurrence rate, while resection and enucleation with bone curettage presented lower rates.<sup>[20]</sup> Due to higher rates of recurrence found in OKC associated with teeth, special attention should be given to the dentate area if enucleation is chosen as treatment.<sup>[8]</sup> In our exhibited case

treated with enucleation and curettage followed by chemical cauterization, no recurrence was found after 1 year and 4 months of follow-up.

OKC has clinical diagnostic difficulties due to relative lack of specific clinical and radiographic characteristics. As OKC has features similar to dentigerous cyst in the radiographs and a meloblastic main incidence and appearance, these are the most common provisional diagnoses for OKC. Ideally, histopathological examination and accurate clinical, radiographic, trans-surgical observation are essential to determine the most effective treatment in order to avoid recurrence.

### Ethical Approval

All procedures performed in the case report involving human participant were in accordance with the ethical standards of the Institutional Ethics Committee of Yenepoya University. Approval has been taken for following ethical standards.

Informed consent was obtained from participant included in the study in her vernacular language after explaining the procedure.

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**How to cite this article:**

Gunachandra Rai B et al (2018) 'Multiple Odontogenic Keratocysts- A Rare Case Report', *International Journal of Current Advanced Research*, 07(2), pp. 10365-10368. DOI: <http://dx.doi.org/10.24327/ijcar.2018.10368.1753>

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