



A Review of the Odontogenic Keratocyst and Report of a Case

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Authors' contributions

This work was carried out in collaboration among all authors. Authors MCS, DAT, MC, GC and MS studied the case and evaluated. Authors MCS and DAT designed the case study and wrote the first draft of the manuscript. All authors managed the literature searches. All authors read and approved the final manuscript.

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Case Study

ABSTRACT

The Odontogenic keratocyst (OKC) is a developmental, non – inflammatory chronic cystic lesion, on radiograph it may be unilocular or multi locular OKC is a cyst of odontogenic origin, usually asymptomatic with an aggressive clinical behavior including a high recurrence rate and tendency to invade bone and adjacent soft tissues. Diagnosis is based on the clinical history, clinical appearance, and radiographs and histology. A case of odontogenic keratocyst involving the ramus of the mandible is presented in this article emphasizing on the characteristics and various features of OKC.

Keywords: *Odontogenic keratocyst; keratocyst odontogenic tumor.*

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1. INTRODUCTION

Odontogenic keratocyst is a distinctive form of developmental odontogenic cyst that deserves a special consideration because of its specific clinical behavior and histopathologic features. The term odontogenic keratocyst was first given by philipsen in 1956 [1]. OKC's most commonly occur in the second and third decades of life and show a slight predilection for males (males to female ratio 1.3:1). The recent WHO classification categorizes OKC as a developmental non-inflammatory odontogenic cyst that arises from the cell rests of dental lamina [2]. Majority of the OKC's occur in the mandible, most commonly in the angle-ascending ramus region.

The clinical and radiographic features of OKC are indefinite; while some may be associated with pain, swelling or drainage, most of them are asymptomatic. OKC's commonly occur in the tooth bearing areas (82%) and some of the cases show an association with atleast one impacted tooth (27% in mandibular third molar) [3]. Here we report a case of odontogenic keratocyst associated with an unerupted mandibular third molar.

2. CASE REPORT

The patient was a 50-year-old female who presented to our department with a chief-complaint of a slowly enlarging swelling, purulent discharge, and bad taste since 4 months duration from the left posterior region of the mandible. She was also having difficulty while swallowing. Her past medical history was negative for major systemic diseases. She has been blind for the past 15 years.

Clinical examination revealed a slight facial asymmetry due to a swelling over the region of

the left mandibular angle. Swelling dimensions were approximately 4 x 3.5 cm antero-posteriorly and 3.5 x 3 cm supero-inferiorly (Fig. 1). The swelling was non-tender and firm in consistency. Skin over the swelling was normal.

On intra oral examination there was obliteration of buccal vestibule of the molar teeth (Fig. 2). Overlying surface of the swelling was of same colour as that of surrounding mucosa. On palpation it was firm in consistency and non tender, on application of pressure a white creamy exudate oozed out of the area distal to 37. Aspiration of the swelling yielded a cream coloured hazy fluid (Fig. 3).

Based on the clinical findings and aspiration of the lesion (FNAC), a provisional diagnosis of odontogenic keratocyst was made, with a differential diagnosis of dentigerous cyst and ameloblastoma.

A panoramic radiograph revealed a well-defined radiolucency on the left ramus of the mandible which was approximately 4x2x1 cm in size, oval in shape, extending anteriorly from distal aspect of 38 to posteriorly to 0.5 cm below the condyle; radiopaque scalloping margins with uniform radiolucency, expansion of inferior border of the mandible at the left angle region, and the inferior alveolar nerve canal was displaced inferiorly (Fig. 4). The bone was intact with no evidence of fracture. Computed tomography revealed a cyst-like radiolucency with scalloped borders and cortical perforations were seen (Fig. 5a & 5b).

Radiographic findings were suggestive of odontogenic keratocyst with a differential diagnosis of unicystic ameloblastoma, and odontogenic myxoma.



Fig. 1. Extra oral clinical photograph showing swelling in the left lower half of the face

An incisional biopsy was done under local anesthesia. Histopathological features of the lesion revealed stratified squamous cell epithelium with parakeratosis and prominent basal layer without rete ridges. Sub-epithelium showed fibro collagenous stroma with islands of squamous epithelium (daughter cysts) (Fig. 6a and 6b). A final diagnosis of odontogenic keratocyst involving the left angle-ramus area was made.

Bramley PA et al. in 1971 proposed a rationale for surgical management of odontogenic keratocyst [4]. He has put forward the treatment options based on the nature of the lesion.

Small single cysts with regular spherical outline: If the access is good it should be enucleated from an intra oral approach.

Unilocular lesions with scalloped or loculated periphery and small multilocular lesions: This type of lesions should be treated by marginal excision; resection of the containing of the block of bone, while maintaining the continuity of posterior and inferior borders as in the ascending ramus, angle and body of the mandible. An extra oral exposure may be needed if obtaining intra oral exposure is difficult.

Large multilocular lesions with or without cortical perforation: This may require resection of the involved bone followed by primary or secondary reconstruction using reconstruction plates of choice like stainless steel, vitallium or titanium.



Fig. 2. Intraoral clinical picture showing a diffuse swelling in relation to 36, 37

In our case considering the size of the lesion as it was huge and involving ascending part of the ramus, coronoid and condylar process. With evidence of thinning of buccal and lingual cortical plates and perforations as seen on computed tomograph. We have proceeded with surgical

resection of the involved bone followed by reconstruction with titanium condylar plates. As the keratocysts involving the ascending ramus were more prone to reoccur, marsupialization couldn't be carried out because of the poor accessibility and aggressive nature of the lesion.



Fig. 3. Showing dense creamy exudate on aspiration

3. DISCUSSION

The history of odontogenic keratocyst dates back to 1826 when Mickuliz first described it as a part of familial condition affecting the jaws. In 1926 it was referred to as "cholesteatoma."- meaning a cystic or "open" mass of keratin squames with a living "matrix". Later in 1945 Robinson mentioned this cyst as primordial cyst as they arose from remnants of the dental lamina or the enamel organs before enamel formation has had taken place. However it was not until 1956 the cyst was named odontogenic keratocyst by philipsen 1. Since then the terminology has been a matter of dispute due to the distinct clinical, radiological, histopathologic features of OKC. Some investigators classify OKC as a benign tumor but the aggressive nature of OKC has put the dilemma as to whether it is a cyst or neoplasm. In 1967 TOLLER suggested that OKC is to be named as benign neoplasm [5]. Shear used the term "keratocystoma" citing the aggressive nature of the odontogenic keratocyst and finally labeled it as a benign cystic neoplasm OKC was reclassified and renamed as keratocystic odontogenic tumor (KCOT) in the WHO classification of head and neck tumors in 2005 [6]. Redesignation of the OKC as the KCOT is based on the well known aggressive behavior of the lesion, histology and new information regarding its genetics.



Fig. 4. Panoramic radiograph showed a well defined radiolucency in relation to left molar ramus area

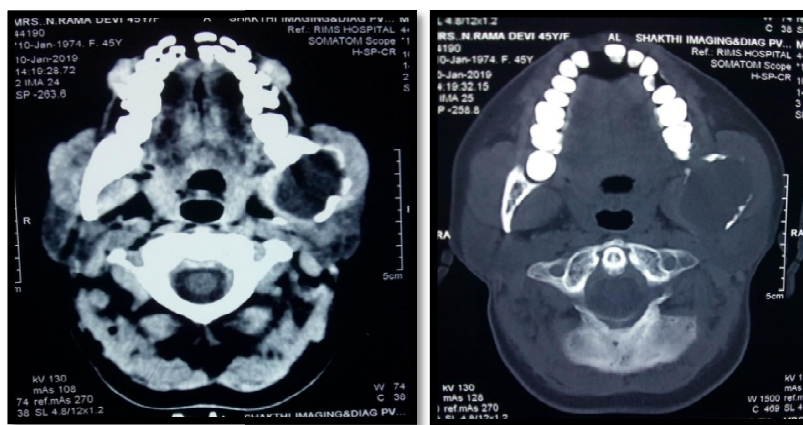


Fig. 5a and 5b. Computed tomography revealed expansile corticated and scalloped cystic lesion

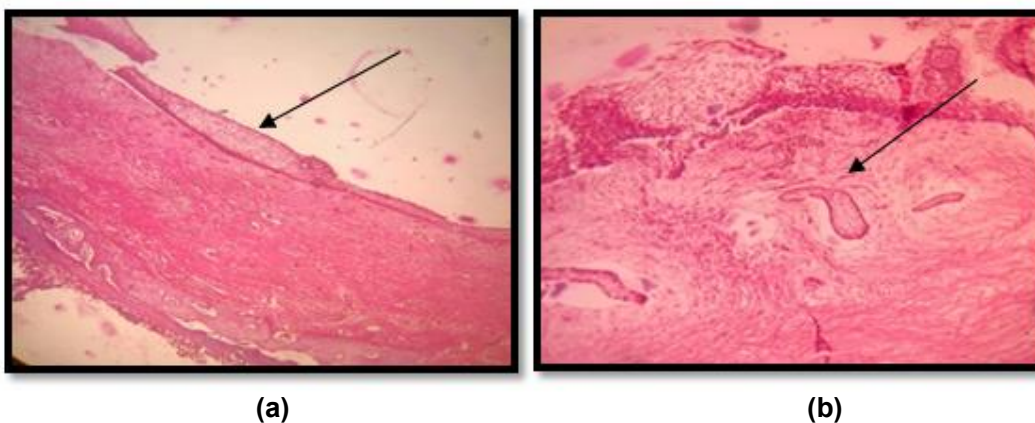


Fig. 6a. Histopathologic section shows stratified squamous epithelium showing parakeratosis with prominent basal layer without rete ridges

Fig. 6b. Histopathologic section shows sub-epithelium islands of squamous epithelium (daughter cysts)

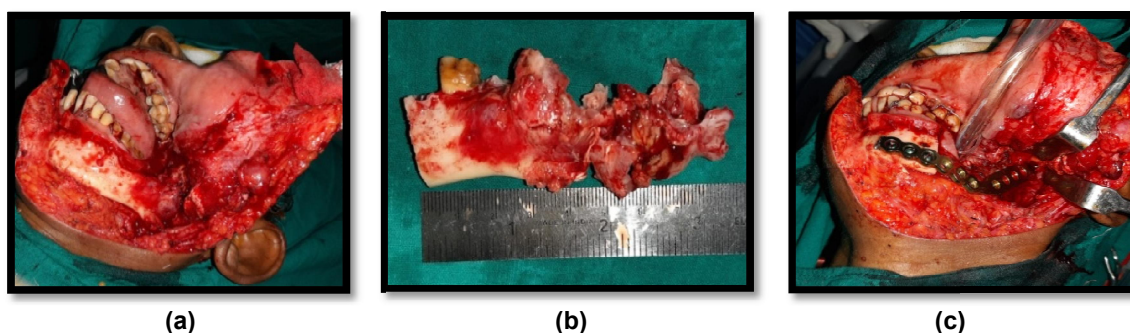


Fig. 7a. Showing the exposed cyst involving the angle- ramus up to the condyle of the mandible

Fig. 7b. Showing the resected mandible along with the condyle

Fig. 7c. Showing reconstruction of the defect with titanium reconstruction plates

The PTCH (Patched gene), a tumor suppressor gene involved in both nevoid basal cell carcinoma syndrome and sporadic KCOTS, commonly occur in chromosome 9q22.3-q31.36-40 [7]. PTCH forms a receptor complex with the oncogene SMO (smoothed) for the SHH (Sonic Hedge Hog) ligand. The growth-signal-transduction is inhibited by PTCH binding to SMO. This inhibition is released by SHH binding to PTCH. When the normal functioning of PTCH is lost, the proliferation stimulating effects of SMO are permitted to predominate [8]. But there was substantial evidence that PTCH gene mutation can also occur in non neoplastic lesions like dentigerous cysts [9]. Moreover, many researchers challenged the neoplastic process of OKC as marsupialization causes resolution of the cyst [10]. Hence the 2017 WHO classification reverted back to the well accepted terminology of odontogenic keratocyst-OKC [11].

Most of the OKC'S arise from the cell rests of dental lamina or from the basal cells of oral epithelium and are thus primordial-origin odontogenic keratocysts [12]. The remaining 40% arise from the reduced enamel epithelium of the dental follicle and are thus dentigerous-odontogenic keratocysts as in our case. The clinical identification is crucial in determining the treatment as the recurrences are more frequently seen after treatment of primordial type of OKC. OKCs may occur at any age but the highest incidence is generally in the second and third decades of life. There is a slight male predilection. Approximately 20-45% of OKC'S are associated with unerupted tooth and about 70% of the cases involve the mandible especially the molar, angle and ramus region. Here in our case all the three distinctive characteristics can be appreciated.

OKC'S tend to grow in antero posterior direction within the medullary cavity of the bone and may cause an obvious bone expansion [13]. A hazy radiolucent lumen can be seen on a conventional radiograph which is suggestive of a dense proteinacious material such as keratin. Resorption of roots of the erupted teeth is rare with displacement of teeth adjacent to the cyst occurring more frequently than resorption.

The histopathologic features of OKC are more specific. Diagnostic features include a uniform cyst lining, hyperchromatic and palisaded basal cells, wavy parakeratin production and a flat interference between the epithelium and connective tissue wall. One of the most peculiar characteristic features of OKC is the appearance of satellite cysts/ islands of odontogenic epithelium [14] which was seen in our case. High recurrence rate can be attributed to the satellite cysts that are retained during enucleation; thin, fragile cystic walls can be left intact.

Unicyclic ameloblastoma and orthokeratinized odontogenic cyst (OOC) though present with the same clinical and radiographic features, Histologically Unicyclic ameloblastoma has ameloblastic epithelial lining which is pathognomic of this cyst. The suprabasilar areas often loosen up giving the appearance like a stellate reticulum [15]. OKC shares similar characteristics with Orthokeratinized odontogenic cyst with respect to age of occurrence and site, yet these two lesions differ in their biological activity [16]. OKC can be differentiated from Orthokeratinized odontogenic cyst on various features like older age group, more antero-posterior extension without expansion, characteristic histopathological features different from Orthokeratinized odontogenic cyst,

parakeratinized layer, high recurrence rate, association with basal cell nervous syndrome.

The treatment options for OKC'S range from simple conservative treatment like enucleation, (with or without curettage), or marsupialization. Marsupialization is a technique relying on incomplete removal of the cyst lining. Opening a window into the cyst forms an invagination of the oral cavity or the maxillary antrum. It relies on the principle that decompression halts expansion of the cyst and appositional growth of bone occurs, and the former cyst lumen becomes smaller with time. Many modification of the procedure have been taken place over the time like usage of decompression tubes, marsupialization catheter. One such method has been demonstrated by COSTA F.W.G et al where he used a segment of polyethylene suction tube, prepared according to the radiographic size of the lesion. Using a disposable needle, a hole is drilled near the extremity, large enough to allow the passage of a 0.8-mm orthodontic stainless steel wire. With the aid of a needle holder, one end of the wire is shaped into a loop and the other end is inserted through the hole in the tube, pulled back, and twisted. The tooth crown is etched with acid and the loop is attached to the dental surface with composite resin. Advantage of this technique over other methods was that it provides greater stability and minimizes the need for additional surgical interventions, when compared to traditional methods where decompression devices were attached to the surrounding structures with sutures. This provides insufficient stability in case of surgical wound dehiscence. Poor adjustment increases the likelihood of device-related complications [17].

Though conservative treatment preserves the anatomic structures they have a high risk for recurrence. The aggressive treatment includes peripheral ostectomy, chemical curettage with Carnoy's solution, or *en bloc* resection. Carnoy's solution is composed of 3 ml of chloroform, 6 ml of absolute ethanol, 1 ml of glacial acetic acid, and 1 g of ferric chloride, is often used as a complementary treatment of lesions with high recurrence rates such as odontogenic keratocyst. The action of this solution is given by chemical cauterization, promoting a superficial necrosis of about 1.5 mm of depth after 5 minutes of bone cavities exposure. In a study conducted by Albuquerque et al on surgical treatment with or without Carnoy's solution in aggressive tumors of odontogenic origin the authors found a beneficial effect of the Carnoy's solution in reducing the

recurrence rate in several cases of jaw aggressive odontogenic tumors. This emphasizes the importance of Carnoy's solution when used in conjunction with conservative procedures like enucleation [18]. The problem of using Carnoy's solution in bone cavity is its effect on the inferior alveolar nerve (if denuded or is free in the cavity after procedure). While the neurotoxic effects of Carnoy's solution on inferior alveolar nerve is still being debated [19], the chloroform used in the Carnoy's solution has carcinogenic potential. Hence, Carnoy's solution must be used with caution in order to avoid the deleterious effects on the nerve and surrounding tissues.

In the present case the surgical intervention included hemimandibulectomy (Fig. 7a, 7b) followed by reconstruction with titanium condylar plates (Fig. 7c).

4. CONCLUSION

Odontogenic keratocyst is a unique entity among odontogenic cysts, due to its varied clinical, radiological and histopathologic features. Hence the correlation of histopathologic findings with clinical and radiographic features is of paramount importance to achieve a definitive diagnosis, as most of these lesions have a prognostically different biologic behaviours and the final diagnosis helps to proceed with the appropriate treatment procedure.

CONSENT

As per international standard or university standard, patient's consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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