

Keratocystic Odontogenic Tumor

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ABSTRACT

In 2005, the World Health Organization renamed the lesion, previously known as an odontogenic keratocyst, as the keratocystic odontogenic tumor (KOT or KCOT). The term odontogenic keratocyst (OKC) was first used by Philipson in 1963 and its clinical and histologic features were confirmed by Browne in 1970 and 1971. In this case report, a young patient with a histology report as an orthokeratinized variety of KCOT and it was a primary lesion with amycystic lining that was thick may be due to chronic irritation because of which it could be removed *in toto*. Resection was not advocated as it causes morbidity, peripheral ostectomy could not be performed as the buccal and lingual cortical plates were already thinned out with areas of perforation. Thus, enucleation with Carnoy's solution was considered ideal for this case. Also, this patient has been on regular follow-up for around 8 months showed good healing with no signs of recurrence.

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INTRODUCTION

In 2005, the World Health Organization renamed the lesion, previously known as an odontogenic keratocyst, as the keratocystic odontogenic tumor (KOT or KCOT). The term odontogenic keratocyst (OKC) was first used by Philipson in 1963 and its clinical and histologic features were confirmed by Browne in 1970 and 1971. At that time, it was believed to be a benign, but potentially aggressive and recurrent, odontogenic cyst, and probably represented the lesion previously termed a primordial cyst. Although most of these cysts were lined by parakeratinized epithelium, a few were orthokeratinized. Over the years, it has generally been agreed that the orthokeratinized versions have a lower incidence of recurrence than the parakeratinized version. Here, we are presenting an orthokeratinized version of OKC treated in a conservative manner.

CASE REPORT

A 24-year-old male patient reported to MGM Dental College with alleged history of pain and swelling over right side of the lower jaw. History revealed that he had first noticed a small and nontender swelling on the right side of the lower jaw in the body region 4 years back which had gradually

increased in size and was presently crossing the midline and involving the left body of the mandible (Fig. 1).

Extraoral examination revealed a swelling involving the right body and symphysis, measuring approximately 10 × 6 cm in size, anteroposteriorly extending from ramus of mandible till midsymphysis region and superoinferiorly from the ala tragus line till the inferior border of mandible on right side. There was a mild fullness on the left body region of the mandible. Mouth opening was around 30 mm. Skin over the swelling was erythematous on the right side and showed two discrete nodules. On palpation, the temperature of the overlying skin was not elevated, borders were diffuse and there was tenderness over the nodules which were fluctuant (Figs 2 and 3). Rest of the swelling was bony hard in consistency and nontender. Paresthesia was noted over the right lower lip.

Intraorally, the lower buccal vestibule was obliterated on both right and left side by a swelling which was extending from the right angle of the mandible to the left body. On palpation, swelling was bony hard in consistency, with areas of perforation close to the lower border of the mandible. Surface was lobulated and mucosa over the swelling was normal and showed no secondary changes. There was no expansion of the lingual cortical plate. Tooth 45 was missing and there was severe mesial tilting of 35 and distal tilting of 44.

OPG revealed a well defined, multilocular, radiolucent lesion with well corticated border extending from the lower third of the ramus on the right side up to the first molar on the left side. Impacted 45 was enveloped by the radiolucent



Fig. 1: Frontal view of the patient showing the extraoral swelling

lesion. Lower border of the mandible was thinned out and showed an area of discontinuity at the right angle. Inferior alveolar canal could not be traced completely (Fig. 4).

Axial CT scan revealed a hypodense lesion involving the left and right body of the mandible, with expansion of buccal and lingual cortical plates which were thinned out with areas of perforation. There were multiple septae extending into the lesion (Fig. 5).

Aspiration yielded a whitish, cheesy, foul smelling material, suggestive of an odontogenic keratocyst.

A preoperative incisional biopsy revealed a thin orthokeratinized lining 4-5 cell layered thick with flattened basal cell layer and prominent layer of stratum granulosum. In some areas, epithelial hyperplasia with rete ridge formation was also seen. Overall histopathological features were suggestive of orthokeratinized OKC.

Considering the fact that it was an orthokeratinized variety of OKC, it was treated conservatively. Under general anesthesia, patient was scrubbed, painted and draped. Crevicular incision was given from 47 to 36 along with two

distal releasing incisions. Buccal mucoperiosteal flap was raised. The overlying thinned out buccal bone was removed with rongeur to increase the size of the bony window. The cystic lesion was enucleated *in toto*. All the involved teeth were extracted. Sharp bony spicules were trimmed. Freshly prepared Carnoy's solution with chloroform, glacial acetic acid, absolute alcohol and ferric chloride was meticulously



Fig. 4: OPG showing well defined multilocular radiolucent lesion in the mandible



Fig. 2: Right side lateral view of the patient showing the swelling and the discrete nodules

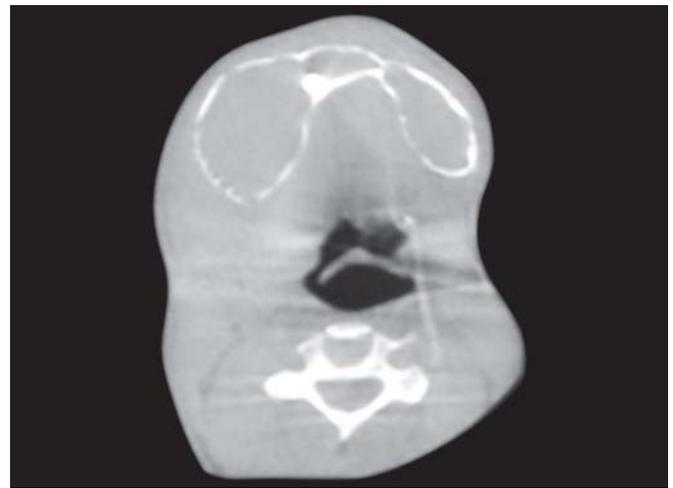


Fig. 5: Axial CT scan showing hypodense lesion involving bilateral body of mandible



Fig. 3: Intraoral view of the patient



Fig. 6: Intraoperative view

applied with cotton pledgets for 3 minutes over the bony walls, protecting the neurovascular bundle. This was followed by normal saline irrigation. As there was a huge dead space, a corrugated rubber drain was placed and primary closure was done (Fig. 6).

Postoperative follow-up showed satisfactory wound healing in all areas, except in the anterior region where there was a wound break down. This was allowed to heal by secondary intention. Follow-up after 3 to 4 weeks showed satisfactory healing with granulation and good wound fill up in the area of secondary healing as well (Fig. 7).

Excisional biopsy report confirmed the preoperative diagnosis.

DISCUSSION

A cyst is a pathological cavity that may be filled with fluid, semifluid or gaseous contents and which is not created by accumulation of pus (Krammer 1974). It is frequently, but not always lined by epithelium.



Fig. 7: Second postoperative day, frontal view of the patient

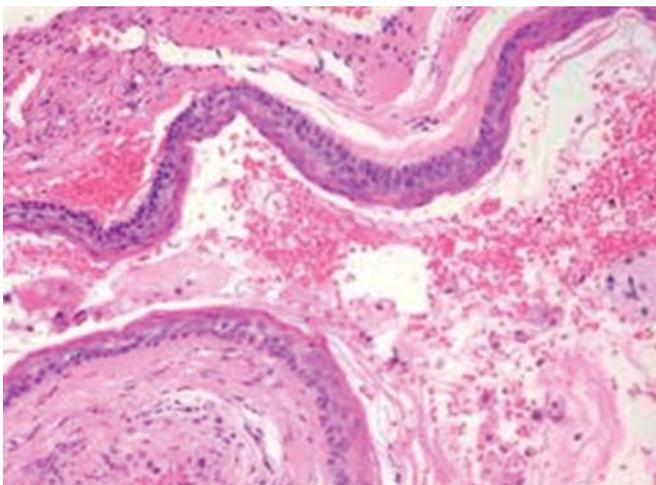


Fig. 8: Histologic features of OKC

OKC is defined as a cyst derived from remnants of dental lamina, with biologic behavior similar to benign neoplasm, with a distinctive lining of 6 to 10 cells in thickness, and that exhibits a basal cell layer of palisaded cells and a surface of corrugated parakeratin (Fig. 8).

Two main sources of epithelium in the pathogenesis of the cyst are dental lamina and remnants or extensions of basal cells from overlying oral epithelium. Enlargement theories are that they extend along the cancellous component of the bone. Forsell¹ (1980) found the rate of growth of keratocyst varies from 2 to 14 mm a year. Toller¹ (1970) suggested that osmolality played a role in cyst enlargement as the mean osmolality of keratocyst was 296 ± 15.6 mOsmol compared with serum osmolality of 282 ± 14.75 mOsmol. Main,¹ however, opined that mitotic value of keratocyst linings ranged from 0 to 19 and hence felt that mural growth in the form of epithelial proliferation was responsible for the growth. High recurrence potential may be attributed to the occurrence of satellite cysts which are retained during enucleation, very thin and fragile lining as a result of which portions of it may be left behind, and a tendency for multiplicity as in nevoid basal cell carcinoma. It has been suggested by Toller et al¹ that epithelium of keratocyst has intrinsic growth potential, a weak connective tissue—epithelial interface due to collagenolytic enzymes.

Radiographically, keratocysts may be classified as replacement, envelopmental, extraneous and collateral varieties.

Aggressive treatment has been advocated for the lesion because of its high recurrence rate of 5 to 62.5%. Most recurrent OKCs seem to show up within first 5 years after surgery. There is a striking difference in recurrence rates of parakeratinized variant (47.8%) and orthokeratinized cysts (2.2%).² Cysts that display keratinization often show the orthokeratinized type, and it is still unclear whether this is caused by metaplasia in response to irritation or they are orthokeratinized from the start. As our case was orthokeratinized, conservative treatment could be done.

Although it was designated as an odontogenic cyst, the lesion behaves more like a tumor. The reasons for this belief include its clinical behavior, with a high recurrence rate after enucleation and, more recently, the presence of tumor markers within the cyst. These markers consist of specifically proliferating cell nuclear antigen (PCNA), ki-67, BCE2 sequencing of the enzyme dihydrolipoyl acetyltransferase, matrix metalloproteinase (MMP) 2 and 9, and p53. This led to the 2005 WHO reclassification of OKC as a keratocystic odontogenic tumor.

Various treatment modalities that have been suggested are as follows:

- *Marsupialization or decompression:* It consists of opening up the cyst to the oral cavity and suturing the cyst lining to the oral mucosa, creating a permanent opening into the cyst. The cyst is therefore decompressed and decreases in size as new bone is laid down around it. It is noted on biopsy of the lining as the cyst decreases in size, that the lining transforms from the thin parakeratinized lining to a thicker lining more resembling the oral mucosa.² It is not known whether this process happens by metaplasia in the KCOT lining or by overgrowth of normal epithelium.
- *Enucleation with peripheral ostectomy:* As high recurrence rate after simple enucleation is believed to be caused by the presence of retained fragments of lining plus daughter cyst that are left behind, it may be that removal of 1 to 2 mm of bone beyond the visible margin of the lesion may be adequate to improve the cure rate. However, it is difficult to estimate how much bone to remove with a drill. This process is made easier by the use of a vital staining technique. Methylene blue or crystal violet can be painted on the walls of enucleated cyst and allowed to penetrate *in toto* the bone. The cavity is then washed out and bone retaining the stain is removed with a drill. This process usually removes around 2 mm of bone in the marrow and 1 mm of cortical bone.²
- *Enucleation with chemical treatment with Carnoy's solution:* It contains absolute alcohol 6 ml, chloroform 3 ml, ferric chloride 1gm and glacial acetic acid 1 ml. Average depth of bone penetration depends on duration of application (1.54 mm after 5 mins).³ Carnoy's solution is a caustic tissue fixative. It is neurotoxic and chemically fixes the inferior alveolar nerve or lingual nerve if it comes in contact with them for up to 2 minutes. The nerve should be therefore protected.² Some investigators state that it should be mixed fresh and used within 2 days, whereas others state that it can be left for several months.
- *Physical treatment with cryotherapy using liquid nitrogen:* Cryotherapy devitalizes organic tissue beyond the visible margin of the lesion, but leaves the inorganic bony framework intact. It devitalizes an area between 1 to 2 mm. A temperature of -20°C is required to devitalize the tissues and only liquid nitrogen can deliver this temperature. Cryotherapy kills cells by means of direct damage to intracellular and extracellular surfaces because of formation of ice crystals that affect the osmotic and electrolytic balance.⁴
- *Resection:* It is an aggressive procedure and results in considerable morbidity.

According to Johnson et al,⁵ simple enucleation is no longer endorsed due to high recurrence rate. A small KCOT where margins may be accessed may be enucleated with adjunctive measures like Carnoy's solution. A large expanding KCOT is best treated by a two-stage approach. Marsupialization first, followed by enucleation and adjunctive measures. Marginal or segmental resection is not advocated as primary treatment due to its morbidity. It may be considered in cases of recurrence.

According to Zhou,⁶ enucleation with subsequent open packing of iodoform gauze was shown to be comfortable for patients and appears to be an effective choice for management of large KCOTs of mandible.

According to Kaczmarzyk,⁷ 100 cases were analyzed for recurrence and results showed 0% for resection, 0% for enucleation with peripheral ostectomy and Carnoy's solution, 18.18% for enucleation with peripheral ostectomy, 26.09% for enucleation alone, 40% for marsupialization and 50% for enucleation with Carnoy's solution.

Leonardo et al⁴ found no recurrence with enucleation and liquid nitrogen cryotherapy in eight patients.

Pogrel⁸ found marsupialization as a definitive treatment for KCOT, in 10 patients. Marsupialization requires a co-operative patient who will irrigate the cavity and keep it open.

Thus, there appears no gold standard treatment modality for the keratocystic odontogenic tumor as it borders more toward a tumor than a cyst. The factors that have to be taken into account while deciding upon the treatment include the age of the patient, the clinical and radiological features, the site of the lesion, the extent of involvement, the histology of the lining epithelium and if it is a primary or recurrent lesion.

In our case, it was a young patient with a histology report as an orthokeratinized variety of KCOT and it was a primary lesion with cystic lining that was thick may be due to chronic irritation because of which it could be removed *in toto*. Resection was not advocated as it causes morbidity, peripheral ostectomy could not be performed as the buccal and lingual cortical plates were already thinned out with areas of perforation. Thus enucleation with Carnoy's solution was considered ideal for this case. Also, this patient has been on regular follow-up for around 8 months and showed good healing with no signs of recurrence.

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