Keratocystic Odontogenic Tumour of the Mandible: a case report of decompression with a customised removable tube and review of literature.

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ABSTRACT
Keratocystic odontogenic tumour (KOT or KCOT) is defined by the WHO to be ‘a benign uni- or multicystic, intraosseous tumour of odontogenic origin’. In 2005, the World Health Organization renamed the lesion; previously known as an odontogenic keratocyst (OKC) as the KCOT. WHO recommends the term KCOT as it reflects its neoplastic nature. In this case report, a 21-year-old female with a histologically proven large parakeratinised KCOT of the right mandible underwent treatment that involved a 14-month period of decompression, followed by enucleation (with Carnoy’s application) of the residual cyst. During the period of decompression, a custom made removable mandibular chrome-cobalt appliance was used to hold the decompression tube in the oral cavity.

INTRODUCTION
Keratocystic odontogenic tumour (KOT or KCOT) is defined by the WHO to be ‘a benign uni- or multicystic, intraosseous tumour of odontogenic origin’. In 2005, the World Health Organization renamed the lesion; previously known as an odontogenic keratocyst (OKC) as the KCOT. WHO recommends the term KCOT as it reflects its neoplastic nature (Johnson and Batstone, 2013; Jattan et al., 2013).

The term OKC was first used by Philipson in 1963 and its clinical and histological 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 parakeratinised stratified squamous epithelium, a few were orthokeratinised. It is now generally accepted that the presence of orthokeratinised histology has a lower incidence of recurrence (Ramawami et al., 2013).

BACKGROUND
KCOT is a unique lesion because of its locally aggressive behaviour, high recurrence rate, and characteristic histological appearance. Their management remains controversial owing to multiple different treatment protocols with varying recurrence rates. Often the first surgical approach in KCOT management is enucleation with adjunctive treatment such as application of Carnoy’s solution, liquid nitrogen cryotherapy and peripheral ostectomy. Some advocate surgical resection of the associated region of jaw to minimise recurrence, although resection generally has been reserved for patients who have undergone several surgical procedures to remove the same recurring KCOT. Patients also tend to require long follow-up because of the nature of KCOT and its significant propensity to recur (Deboni et al., 2011). Decompression and marsupialisation have been well described and are usually reserved for very large cysts.

Treating KCOTs via either marsupialisation or a period of decompression has been shown to considerably reduce the size of the cystic lesion, enabling the lesion to shrink away from important structures such as the inferior alveolar nerve and tooth roots. Definitive treatment of the reduced lesion can be carried out as a second-stage procedure. Previous literature has shown that during decompression, the cyst wall can undergo metaplastic change. It has been hypothesised that this metaplasia of the cyst wall may be a contributing factor in the reduction of recurrence rate (Ramawami et al., 2013).

Securing the decompression tubes to the oral cavity has usually been described using wire, sutures, or other similar materials. The use of a removable appliance offers advantages such as the at-home removal of the appliance on a regular basis to clean both the decompression tube and the supporting dentition. It also provides the opportunity to exchange the decompression tube if required.

Here we present a parakeratinised KCOT treated in a conservative manner via decompression over a 14-month period, followed by enucleation of the remaining cyst using a removable appliance not previously described in the literature.

CASE-REPORT
A 21-year-old female was referred to the Department of Maxillofacial, Oral Surgery, and Dental at Waikato Hospital, Hamilton, New Zealand. She had been referred to the service from a local Oral and Maxillofacial Surgeon (OMS), presenting with a twelve week history of recurrent infection of the lower right third molar. The OMS had supplied a conebeam CT radiograph (Figures 1, 2) which showed an extensive multiloculated radiolucency related to the unerupted and impacted tooth 48. The patient had no evidence of paraesthesia or mandibular asymmetry. Her medical history was unremarkable except for allergies to penicillin and erythromycin. She was a life-long non-smoker.

Intraorally the patient was in fixed orthodontic retention with sound dentition. Pus was clearly draining from behind the distal aspect of tooth 47, which was tipped distally and slightly tender to percussion. Tooth 47 tested positive to both hot and cold pulp testing. All other teeth of the lower right mandible were clinically asymptomatic.

CT scan showed a unilocular cystic lesion involving the entire ascending ramus of the mandible up to the level of the condylar neck, involving the sigmoid notch, and coronoid process of the right mandible. Scalloping of the cortical outline was particularly evident at the posterior and inferior borders extending to cause fenestrations in the lingual plate on the inferior aspect (Figures 1, 2).

The clinical and radiographic findings were consistent with a diagnosis of KCOT and was deemed to be amenable
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Upper and lower alginate impressions and a wax occlusal record were taken in order to fabricate a custom-made-mandibular chrome-cobalt appliance which was designed to securely hold the decompression tube (Figure 5). The patient was commenced on antibiotic therapy and decision was made not to perform an incisional biopsy until the infection was resolved.

Under general anaesthetic teeth 18, 28, and 38 were surgically removed conventionally. On the 48 site the buccal and superior bone of the ramus was intact. A bony window was made on the superior surface of bone directly over the 48 tooth. This allowed the underlying cystic lining to be clearly visible and a representative sample of both the bone and the cystic lining were excised and sent for histopathological analysis. The cyst contents consisted of thick yellow coloured keratinaceous material. The deeply impacted 48 tooth was surgically removed and the chrome-cobalt appliance with the decompression tubing was fitted. Unfortunately one of the cast clasps fractured necessitating the framework to be remade. An interim decompression tube was therefore placed into the cystic cavity and wired to the mandibular dentition for stability. Patency of the decompression tube was confirmed with irrigation. Post-operatively the patient was instructed to irrigate down the decompression tube using 0.2% chlorhexidine gluconate (healthE Jaychem Mouthwash) three times a day. Postoperative OPG confirmed correct placement of the tube. Histology confirmed a parakeratinised KCOT.

The recast chrome cobalt frame was then fitted in outpatient clinic and the decompression tube was placed in the frame. Over the following months, the patient was able to remove the decompression tube and framework in order to clean the appliance as well as her mouth and then replace it herself. At three, six, twelve, and fourteen months post-op, the decompression tube was gradually shortened and OPG radiographs were taken to view bony changes of the cyst within the mandible and to verify the position of the tube. Throughout the postoperative period, the patient maintained immaculate oral hygiene, aided by the fact she was able to remove the appliance daily to clean both the appliance and her teeth.

After 14 months, an interval CT scan showed the cyst had significantly reduced in size; sufficient to undertake definitive enucleation of the residual KCOT (Figures 3, 4). Under a general anaesthetic the remaining cyst was enucleated with excision of the overlying mucosa. Bony curettage was completed with a burr: peripheral ostectomy. The inferior alveolar nerve was not visualised or encountered. Modified Carnoy’s solution (Chloroform 6mL, Ferric Sulfate 1g, Glacial acetic acid 1mL, ethanol 6mL) was applied to the cavity for three 3-minute cycles with saline washout between.

Histopathological analysis of the residual cyst wall found this lesion to be a parakeratinised variant of a Keratocystic Odontogenic Tumour. At 20 months follow up there is no evidence of recurrence and no paraesthesia of the IDN. There is no pocketing around the 47 tooth and the surgical site is well healed.

DISCUSSION
Surgical treatment methods for KCOTs have been diverse, ranging from simple enucleation to enucleation with adjunctive treatment such as the application of Carnoy’s solution and cryotherapy. Marsupilation or decompression can be carried out as the first of a two stage treatment plan. Enbloc resection
is the most aggressive treatment option and addresses the ‘neoplastic nature’ of the KCOT (Ramaswami et al., 2013).

**Simple enucleation:** Enucleation refers to the surgical removal of a mass without cutting or dissecting it. Enucleation does not ensure complete removal because remnants of a cyst’s epithelial components may easily be overlooked. Epithelial remnants or residual tissue are ostensibly primed potentiators of recurrence (August et al., 2003). For this reason, KCOT’s treated with enucleation are shown to have a significantly higher recurrent rate than those treated with other methods (Madras and Lapointe, 2008).

**Enucleation with adjunctive treatment:** Post-enucleation use of techniques such as chemical cautery, cryotherapy modalities, Carnoy’s solution, aggressive curettage of bony walls, or peripheral ostectomy with a bone bur have been advocated as means of lowering recurrence by removing remaining epithelium (Ramaswami et al., 2013).

Higher recurrence rates after simple enucleation are believed to be caused by the presence of retained lining fragments and daughter cysts that are left behind. It is thought that the removal of 1 to 2mm of bone beyond the visible margin of the lesion may be adequate to reduce the rate of recurrence. However, it is difficult to estimate how much bone to remove with a drill. This process is made easier by the use of vital staining technique. Methylene blue or crystal violet can be painted on the walls of enucleated cyst and allowed to penetrate into the bone. The cavity is then washed out and bone retaining the stain is removed with a drill. This process usually removes around 2mm of bone in the marrow and 1mm of cortical bone (Ramaswami et al., 2013).

Carnoy’s solution contains alcohol, chloroform, ferric chloride and glacial acetic acid. The average depth of bone penetration depends on duration of application (1.54mm after 5 minutes). 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 longer than 2 minutes. The nerve should therefore be protected. Some investigators state that Carnoy’s solution should be mixed fresh and used within 2 days, whereas others state it can be left for several months (Ramaswami et al., 2013). Due to the fact that medical grade ferric chloride is no longer available, current recipes for Carnoy’s solution are not the same as their traditional predecessors, hence the term ‘modified Carnoy’s’ is now considered more appropriate.

Cryotherapy (using liquid nitrogen) devitalises organic tissue beyond the visible margin of the lesion, but leaves the inorganic bony framework intact. It devitalises an area between 1 to 2mm. A temperature of -20 degrees Celsius is required to devitalise the tissue. 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. Cryotherapy weakens the bone and can result in complications such as iatrogenic fracture (Ramaswami et al., 2013) (Schmidt and Pogrel, 2001).

**Surgical resection:** En-bloc resection is an aggressive procedure that results in considerable morbidity. It involves removal of the entire segment of jaw containing the cystic lesion, and is commonly associated with sacrifice of the inferior alveolar nerve (Ramaswami et al., 2013).

Both Decompression and Marsupialization involve opening up the cyst to the oral cavity via removal of overlying mucosa and bone, creating a persistent communication between the cystic cavity and external environment. The cyst is therefore decompressed and decreases in size, being replaced by new bony infill. Unlike decompression, Marsupialization involves the suturing of the oral mucosa to the cystic membrane to maintain a persistent communication from the oral cavity into the cyst lumen space. With decompression a tube is inserted into the cystic cavity, which maintains a communication from the oral cavity into the cyst lumen providing a portal for irrigation. Sole responsibility is placed on the patient to irrigate the cavity on a daily basis for an extended period of time, often between three and twelve months (Madras and Lapointe, 2008). Histological analysis of the cyst lining reveals that as the cyst decreases in size, the lining can transform. It has been hypothesised that this process may occur by either metaplasia or overgrowth of normal oral epithelium. Either way, the resultant cyst lining is seen to be histologically less like KCOT and more representative of normal oral mucosa (Ramaswami et al., 2013).

KCOTs have been found to have a variable and high recurrence rate (5 to 62.5%) (Ramaswami et al., 2013). In 1976, Brannon proposed 3 mechanisms for KCOT recurrence: 1) incomplete removal of the cyst lining, 2) growth of a new KCOT from satellite cysts (or odontogenic rests left behind after surgery) and 3) development of a new KCOT in an adjacent area that is interpreted as a recurrence. The wide range in reported recurrence rates has been attributed to the variation in follow-up times used by examiners, the surgical technique used and the number of cases incorporated into the studies. Most recurrences take place within 5–7 years after treatment, although some have been reported more than 10 years following initial intervention. These findings emphasise the importance of long-term follow-up as an essential aspect of the KCOT treatment plan (Madras and Lapointe, 2008).

A review of the literature suggests that recurrence rate varies with different treatment protocols. According to Johnson et al., (2013), simple enucleation is no longer endorsed due to a high recurrence rate. A small KCOT where margins may be accessed should be enucleated with an adjunctive measure. A large expanding KCOT is best treated by a two-stage approach; Marsupialization or decompression as the initial treatment, followed by enucleation and adjunctive measures. Marginal or segmental resection is not advocated as primary treatment due its morbidity but may be considered in cases of recurrence.

For simple enucleation, a recurrence rate between 17% and 56% has been reported. With the additional application of Carnoy’s solution, a recurrence has been found to decrease to the region of 1% to 8.7%. Secondly, the use of cryotherapy with enucleation appears to reduce the recurrence rate compared with enucleation alone, however complications such as difficulty applying the freezing agent and iatrogenic fracture have been reported. Third, marsupialization or decompression as a sole definitive treatment is associated with a significantly higher recurrence rate than when the KCOT is subsequently enucleated and treated with Carnoy’s solution. Finally, resection, while having a recurrence rate of 0%, is associated with significant morbidity to the patient including paraesthesia, anaesthesia, loss of dentition, and aesthetic disfigurement. Many authors find this morbidity unacceptable, especially given that KCOT is a benign disease. It is now widely accepted that enucleation with an adjunctive measure such as application of Carnoy’s solution, peripheral ostectomy or cryotherapy can minimise both morbidity of treatment and recurrence of the cyst. Alternatively, marsupialization and decompression tube treatment followed by cystectomy is likewise effective and useful for large cysts, as this treatment does not result in a significantly higher rate of recurrence than enucleation plus...
Carnoy’s solution, and provides advantages in reducing risk of damaging important structures such as nerves and tooth roots. However, as the latter option requires a protracted course of treatment, patient compliance must be considered; lesions treated in this manner require several months of at-home irrigation by the patient as well as clinical observation before enucleation (August et al., 2003).

There is a striking difference in recurrence rates of parakeratinised variant and orthokeratinised cysts, with reported recurrence rates of 47.8% and 2.2% respectively. Cysts that display keratinisation often show the orthokeratinised type. It is still unclear whether this is caused by a metaplastic response to irritation or if they are orthokeratinised from the start (August et al., 2003).

The literature describing decompression tube treatment for KCOT have previously described fixing the decompression tube to the dentition. This creates the need of some form of fixation of the decompression tube into the oral cavity, preventing it from regularly falling out of the cystic cavity (via the epithelial lined tract). Previous authors have suggested using arch wire to fix the decompression tube to the associated teeth or suturing the tube to the soft tissue. This technique has been successful, however it does not allow the patient to easily remove and insert the tube, risks the tube being displaced and does not allow for easy oral hygiene.

Creating a removable rather than fixed appliance offers several advantages. Regular removal and reinsertion of the appliance allows the patient to practice optimal oral hygiene. Minimal patient education is required to teach the patient how to clip the appliance onto the dentition while inserting the tube portion into the cystic cavity. In addition, the appliance maintains a state of constant orthodontic position, whereas wiring can predispose the patient to tooth movement. Furthermore, the length of the tube can be easily altered as the cyst shrinks and the cavity fills in with bone.

This method did however come with its complications. Unfortunately the appliance was unable to be fitted in the operating theatre during the first procedure (extraction of all wisdom teeth and formation of a window into the cystic cavity). This was because the appliance (which had been made three weeks in advance) did not fit to the dentition. It appeared that tooth 47 had further tilted in the time between obtaining the initial alginate impressions and the first operation. As a result the decompression tube was wired into place as a temporary measure.

In order to resolve this early stage complication, new alginate impressions were taken at a post-operative clinic appointment for the fabrication of a new framework (Figure 3). Fortunately, this framework obtained a perfect fit and was successful through the entire course of treatment. The patient was able to remove the appliance when brushing, and was advised to leave it in place at all other times.

To avoid this issue happening in future, the mandibular-chrome-cobalt appliance should be made as close to the date of surgery as possible. In saying this, consideration of the time required by technicians to make such appliances must also be taken into account.

It is crucial to confirm histological diagnosis of KCOT before embarking on a protracted conservative treatment such as decompression therapy. Misdiagnosis can result in inappropriate decompression of other pathologies such as ameloblastoma. Under normal circumstances and with no evidence of acute infection, the usual practice of Waikato Hospital’s Maxillofacial department is to perform an incisional biopsy, often under local anaesthesia, to confirm the diagnosis before commencing decompression therapy. In this case, we elected to treat the patient’s infection with antibiotics, then in a single operation under general anaesthetic remove the third molars, perform an incisional biopsy of the cyst wall and insert the decompression tube.

Finally, optimal oral hygiene and patient compliance are the forefront to determine success of decompression tube treatment. Edentulous or partial edentulous patients may not be suitable due to the obvious lack of sufficient dentition.

REFERENCES

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