REVIEW

Treatment of a patient with large keratocystic odontogenic tumour in the mandible: case report with literature review

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Abstract

Presented heretofore is the case of a 26-year-old male suffering from a keratocystic odontogenic tumour that exhibits progressive development in the mandible. During our evaluation, the patient underwent numerous diagnostic and therapeutic procedures to restore his normal appearance and function.

The keratocystic odontogenic tumour (KCOT), formerly classified as a parakeratinised type of cystic lesion named odontogenic keratocyst (OKC), was subsequently reclassified in 2005 by the World Health Organisation (WHO) Working Group as a neoplastic lesion. Currently, the KCOT is classified as a benign unicystic or multicystic intraosseous tumour of odontogenic origin, possessing a characteristic lining consisting of parakeratinised stratified squamous epithelium with the potential for aggressive, infiltrative behaviour¹.

Several factors influencing the aforementioned reclassification include the following: the potential for locally destructive behaviour; a high recurrence rate and abundant mitotic activity in the cystic epithelium²; the potential for epithelial budding of the basal layer; the presence of satellite cysts attached to the side walls of the pericystic cavity³; the predilection to proliferate; chromosomal aberrations⁴; and lastly, primary pertinence to mutation of the PATCHED gene (PTCH) in its aetiology⁵.

Case report

An 18-year-old patient was referred to our department by a dentist after a painful swelling developed in the submental region. Following the facial swelling aforementioned, the patient detected a progressive displacement of lower teeth coupled with mandibular body distension. Clinical examination revealed painful swelling of the submental region, enlargement of the submental lymph nodes, and lower teeth experiencing displacement and distension of the mandibular body, ranging from the area of the second left premolar to the second right molar (Fig. 1). The patient exhibited negligent oral hygiene and required multiple tooth extractions. The utilisation of pantomographic radiography indicated a large multicystic lesion exhibiting sharply defined borders distending the mandibular body. The range of the multicystic lesion spanned from second left premolar to second right molar (Fig. 2). No root resorption occurred alongside the significant displacement of lower incisors. All teeth remained vital.

Following administration of local anaesthesia, a fine-needle aspiration biopsy upon the lesion was performed and revealed abundant purulent fluid. Bacteriological assessment indicated abundant growth of oral streptococci. Cytological assessment revealed a concentrated protein liquid with leukocytes. The laboratory test results of patient’s level of calcium,
phosphorus, and alkaline phosphatase were within normal range. The aggravated symptoms of progressive lesion development required the patient to undergo surgical treatment, with the main objective being the elimination of major intraoperative trauma in the impaired bone structure. The surgical procedure was performed in two stages. During the initial surgical stage, the lesion was decompressed under local anaesthesia with sample tissue biopsied for histopathological assessment and the affected bone cavity packed with iodoform gauze (Fig. 3). After receiving post-operative histopathological assessment which revealed a para-keratinised type of OKC (2003; Fig. 4), gauze packs were removed, and an acrylic obturator-type appliance was constructed and inserted into the bone cavity (Fig. 5). Systematic and long-term post-first stage surgical follow-ups focused on rebuilding bone structure evaluated with imaging studies with the obturator, being regularly corrected as the bone rebuilding progressed. During this phase of treatment, the patient’s oral hygiene had experienced further improvement with the administration of caries treatment.

Nineteen months following decompression, the lesion had decreased immensely, thus its range spanned from right canine to second left incisor (Figs 6 and 7). An obturator was eliminated, and orthodontic treatment was started so as to produce future satisfactory occlusion. An orthodontist applied metal orthodontic braces on the mandibular dental arch. Sixty-two months proceeding lesion decompression, shallowness of the bone cavity in the medial region of the mandible required the second stage of surgical

Figure 1 Preoperative intraoral picture showing lower teeth experiencing displacement and distension of the mandibular body, ranging from the area of the second left premolar to the second right molar.

Figure 2 Preoperative orthopantomogram showing large multicystic lesion distending the mandibular body, spanned from second left premolar to second right molar.

Figure 3 Intraoperative picture showing decompression with sample tissue biopsied for histopathological assessment.

Figure 4 Histopathology. Original material: a cyst lined with seemingly non-keratinising epithelium is visible. Focal parakeratosis is present, however (inset). H + E, lens magnification 40x.

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treatment, consisting of enucleating the residual mass of the lesion under local anaesthesia (Figs 8 and 9). Subsequent histopathological assessment revealed the presence of focal areas of residual parakeratosis (2005). The patient, very cooperative and satisfied with the results of the treatment, still required systematic post-surgical check-ups and long-term orthodontic treatment. His clinical status and proper functioning were satisfactory, and self-esteem was dramatically improved.

Seven years after the lesion decompression and 2 years following enucleation, the regular follow-up visit revealed an asymptomatic recurrence. The pantomographic radiography indicated a small unicystic lesion with sharply defined borders distending the mandibular body area spanned from right canine to second right premolar (Fig. 10). Preoperative computed tomography (CT) scans revealed the lesion located mainly in the proximity of the right first premolar with its lingual expansion as well as the mandibular body distention in buccal direction (Fig. 11). The buccal cortical bone remained intact. The recurrence of the lesion required the patient to undergo surgical treatment. Following administration of local anaesthesia, the lower right first premolar was extracted in order to facilitate the radical removal of the lesion. The lesion was enucleated with peripheral bone curettage with rotary instruments. Post-operative histopathological assessment confirmed the presence of a KCOT (2010; Fig. 12).

Currently, the patient is very satisfied with the results of treatment. He is presently restored to an improved appearance combined with pre-KCOT functioning.
The KCOT (formerly parakeratinised type of OKC) received its revised designation from WHO to better illustrate its neoplastic nature characterised by local aggressiveness and the expression of its highly recurring infiltration pattern into surrounding tissues. KCOTs have a slight predilection towards male patients, commonly occur in the second or third decade of life, and typically located in the posterior region of the mandible. Maxillary tumours are prone to infections even when immature and are more likely to be diagnosed at an earlier stage of development. Larger KCOTs are particularly common occurrences at the angle of the mandible and ascending ramus, and can be significant in children.

Therapeutic approaches in different studies have varied from marsupialisation and enucleation, which may be combined with adjuvant therapy, such as cryotherapy and Carnoy’s solution, to marginal or radical resection. Decompression, followed by delayed enucleation of the residual mass of the tumour, hinders a complete removal of the entire epithelial lining during the first stage of treatment. This may consequently lead to a continuous proliferation of the epithelium, potentially facilitating recurrence or malignisation. The patient’s required compliance for a prolonged period may prove to be disadvantageous. Some authors consider the use of decompression as a delay in proper diagnosis of either ameloblastomatous or malignant transformation. On the other hand, decompression leads to histological changes of the KCOT lining, eventually resulting in the lining being replaced by oral epithelium. Studies of Piattelli et al., based on investigation of the immunohistochemical expression of the
B-cell lymphoma 2 bcl-2 protein prove that the immunoprofile of KCOT varies from odontogenic cysts. The results of the conducted study indicate that KCOT, juxtaposed with odontogenic cysts, demonstrate bcl-2 positivity in the basal cell layer15. A literature review suggests that bcl-2 is rarely expressed by some basal keratinocytes of normal epithelium14. Therefore, bcl-2 protein could be useful to differentiate KCOT lining from normal epithelium. Pogrel and Jordan reported bcl-2 negativity in normal oral mucosa and in the KCOT lining, present after decompression that supports the notion of its transformation into normal epithelium during this treatment15. Presence of inflammation after decompression perhaps changes the biological behaviour of the KCOT into a less aggressive form, evincing transformation of epithelial lining into non-keratinised epithelium16–18. August et al. evaluated the epithelium of KCOTs after decompression and reported differentiation of the epithelium, with 64% of the patient presenting loss of cytokeratin-10 expression in analysed epithelium, what lower the risk of recurrence16,17,19. After reviewing the results of the conducted in vitro studies, it could be seen that interleukin-1 (IL-1) stimulates epithelial cell proliferation directly and/or indirectly by inducing the secretion of keratinocyte growth factor from interacting fibroblasts20,21. The proliferating activity of the epithelial cells is related to the expansion of KCOT22. Therefore, IL-1A is considered to be an important factor in regulating the KCOT growth. Ninomiya et al. reported that the epithelial cells of KCOT express IL-1A mRNA and protein, and decompression by marsupialisation immensely reduce the expression proportionally with the epithelial cell proliferation23. As a consequence of these benefits, decompression offers significant advantages in decreasing the size of the tumour.

Based on the available literature, recurrence rates vary from 0% to 100%5. The majority of research in this area indicates that most recurrences appear within the first 5–7 years; however, relapse may occur 9 or more years after the initial treatment24. Established results have shown different recurrence rates for specific treatment modalities. Despite many studies indicative of radical methods advantages over conservative treatment, no conclusive proof has been established convincing that conservative methods do not increase the success rate in treatment of KCOT. Brøndum and Jensen reported no recurrences during a follow-up period of 7–17 years in 12 patients with large KCOTs treated by decompression and irrigation25. Pogrel and Jordan reported no relapse during a follow-up period of 1.8–4.8 years in 10 patients with large KCOTs treated by marsupialisation and decompression15. Marker et al. reported a recurrence rate of 8.7% after decompression with subsequent enucleation of the tumour12,19.

Despite the fact of recurrence in the presented case and considering the complications of radical surgery, decompression, followed by delayed enucleation, offers increased quality of life especially in young cooperative patients suffering from a large KCOT.

It is commonly accepted that conservative treatment by decompression offers significant advantages in eliminating major surgical procedures. Despite the fact that decompression requires longer time when compared with other methods of treatment, it reduces the chance of a pathological fracture, loss of teeth vitality, or bony discontinuity with definitive treatment.

It needs to be emphasised that systematic and long-term follow-ups are considered to be a key element for successful results.

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**References**


