CASE REPORT

Conservative management of keratocystic odontogenic tumour in a young child with decompression and an intraoral appliance: 5-year follow-up

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SUMMARY

Keratocystic odontogenic tumour (KCOT) is considered one of the most aggressive odontogenic lesions presenting high recurrence rate which varies according to treatment modalities employed for management. The treatment rendered should have a lowest possible risk of recurrence and least morbidity while still eradicating the lesion. Although the radical treatment options like enucleation and en bloc resection are associated with lesser recurrences, these can lead to greater morbidity, especially in children with developing teeth and jaw bones, thus, emphasising need to consider more conservative treatment options like decompression and marsupialisation. The purpose of this article was to report the case of an 11-year-old male child with KCOT in the mandibular region associated with impacted premolar treated successfully with decompression and intraoral appliance. After 24 months of follow-up, the spontaneous eruption of premolar was noted with complete resolution of periapical radiolucency. No recurrence was noted even after 5-year follow-up.

BACKGROUND

The keratocystic odontogenic tumour (KCOT) previously known as odontogenic keratocyst, was first described in 1956 by Philipsen as a developmental cyst of odontogenic origin.¹ Later, the WHO in 2005 referred odontogenic keratocyst to the benign odontogenic tumours category and defined it as 'a noncancerous uni- or multi-cystic, intraosseous tumour of odontogenic origin, having a typical inner lining of parakeratinized stratified squamous epithelium as well as the likelihood of a potent, destructive behavior' and termed it as 'keratocystic odontogenic tumour'.²

KCOT primarily involves the mandibular ramus and angle region. Radiographically, KCOTs appear as a well-defined unilocular or multilocular lesion with scalloped borders and are difficult to differentiate from other cyst and tumours of jaws.³ KCOTs are well known for their high recurrence rate. The rate of recurrence is variable and varies according to different treatment modalities ranging from as low as 0% to the maximum of 62%. In the majority of cases, recurrence occurs within the first 5 years after the treatment.^{4 5}

The treatment modalities vary from aggressive management in terms of peripheral ostectomy with/without application of Carnoy's solution, liquid nitrogen cryotherapy and options like marsupialisation and decompression followed by secondary enucleation.⁶⁻⁸ The choice of treatment should take into account various factors, including patient's age, size and location of the cyst, soft tissue involvement, history of treatment and the histological variant of the lesion.⁹ The goal is to choose the treatment modality that will completely eradicate the lesion with the lowest possible risk of recurrence and the least morbidity. The purpose of the present case report is to describe the conservative management of KCOT in a young child with decompression only and its outcome over a 5-year period.

CASE PRESENTATION

An 11-year-old male child reported to the Unit of Pedodontics and Preventive Dentistry, with the chief complaint of swelling in the lower right posterior region of the jaw for the last 2 months. As per the history, the swelling gradually increased to the present size and was not associated with any trauma, pain and fever. Medical history was non-significant with no history of any systemic illness or long-term medication. On examination, a diffuse swelling extending from corner of mouth to the angle of mandible on the right side was noticed. On palpation, the swelling was firm and tender with normal skin colour and temperature. Intra-orally, a diffuse swelling extending buccally as well as lingually from mesial of mandibular right second premolar to distal of permanent mandibular first molar obliterating the buccal vestibule was noticed (figure 1). The overlying mucosa was normal in colour and texture. On palpation, the swelling was firm and tender with egg shell cracking sign present over the most prominent area. The right lower deciduous canine and second molar, as well as left lower deciduous canine, were over-retained without any mobility.

INVESTIGATIONS

Panoramic radiograph showed a unilocular radiolucency with irregular borders associated with impacted second premolar and extending from right lower deciduous canine to the distal of

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Figure 1 Diffuse swelling extending from mesial of primary mandibular right second molar to distal of right first permanent molar.

the permanent first molar. There was mesial displacement of impacted permanent canine with thinning of the lower border of the mandible. Also, though not involving the cystic cavity, a horizontally impacted 33 was evident on radiograph (figure 2). Fine needle aspiration yielded a thick yellow liquid.

DIFFERENTIAL DIAGNOSIS

Keratocystic odontogenic tumour, dentigerous cyst and ameloblastoma were considered in differential diagnosis.

TREATMENT

Considering the age of the child and the extent of the lesion and in an attempt to decrease the morbidity and trauma by preserving tooth and bone, it was planned to initially manage the lesion conservatively by decompression. After informing parents of the child about the treatment plan and obtaining their consent, 85 was extracted under local anaesthesia to establish a surgical drainage and a biopsy tissue sample was obtained from the access cavity. Iodoform gauze dressing was inserted into the cavity. Histopathological examination of the biopsy sample revealed a large area of degenerated keratin and few strips of stratified squamous epithelium along with mild inflammatory infiltrate and the findings were suggestive of a keratocystic odontogenic tumour (figure 3). The iodoform dressing was changed every alternate day for 2 weeks and later a surgical stent made up of self-cure acrylic resin with acrylic

Figure 2 Unilocular radiolucency with irregular borders extending from distal of primary mandibular right canine to the distal of right first permanent molar leading to the inferior displacement of the primary mandibular right second molar with thinning of the lower border of the mandible.

Figure 3 Histopathology of cystic lining showing large area degenerated keratin and few strips of stratified squamous epithelium along with mild inflammatory infiltrate.

extension into cystic cavity was placed over the extraction socket to maintain its patency (figure 4). Home care instructions regarding the cleaning of the appliance and irrigation of the cystic cavity with 0.12% chlorhexidine solution using plastic syringe were explained to the child and parents. The child was recalled biweekly for 6 months following surgery and bimonthly thereafter. Panoramic radiographs were obtained regularly at 6-month interval.

OUTCOME AND FOLLOW-UP

One-year follow-up radiograph revealed a decrease in radiolucency and erupting second premolar showed continuous root formation (figure 5). The acrylic appliance was discontinued. At 24 months, the second premolar was seen to be spontaneously erupting in the oral cavity and the radiograph revealed complete resolution of the radiolucency. At that time, horizontally impacted left mandibular canine was also surgically extracted. After 5-year follow-up, the patient continued to be asymptomatic and impacted second premolar had completely erupted into oral cavity (figure 6). The radiographic examination revealed complete healing of surgical site and root completion with 45

Figure 4 Surgical stent made up of self-cure acrylic resin in place.

Figure 5 Orthopantomogram showing bone regeneration and development of root of permanent mandibular right second premolar at 1-year follow-up.

DISCUSSION

The KCOT is one of the most aggressive lesions of odontogenic origin with high recurrence rate and tendency to invade adjacent tissues. The recurrence rate may depend on lesion size, the treatment modality employed for the eradication of lesion and invasion of daughter cells into the surrounding tissues.¹⁰ The resorption of bone by the KCOTs is mediated by activation of osteoclast like cells and collagenases. Expression of interleukin-1, bcl-2 protein and survivin, an inhibitor of apoptosis, may contribute to aggressive behaviour of the lesion and also differentiate it from other non-neoplastic jaw cysts. An aggressive pathological behaviour, the molecular alterations in growth factors (p53, Proliferating cell nuclear antigen and bcl-2) and mutations in the *PTCH* tumour suppressor gene had strengthened the concept of considering the lesion as neoplasm.¹¹ However, there is no consensus regarding the benign yet neoplastic character of KCOT as some studies could support the neoplastic origin of the lesion, but others showed that these lesions are developmental cysts with some neoplastic properties because of the high intrinsic growth potential.^{12 13} WHO classification on cysts and tumours of oral cavity in 2017 suggested that the term KCOT should be reverted back to odontogenic keratocyst as *PTCH* gene mutation was also reported in many non-neoplastic lesions.¹⁴

Treatment of KCOTs has always been a debatable subject. The most advocated treatment option in literature is the complete removal of the lesion and its margins with the associated impacted tooth. However, in young children possibility to preserve and promote eruption of impacted teeth should not

Figure 6 Intraoral view at 5-year follow-up showing erupted premolar.

Figure 7 Orthopantomogram at 5-year follow-up showing complete resolution of the lesion and no sign of any recurrence.

be ignored and more conservative treatment options should be considered. Conservative treatment options of marsupialisation and decompression for the treatment of KCOTs were criticised in the past, considering these techniques do not allow complete removal of the lining and epithelial remnants can continue to proliferate which can lead to recurrence.¹⁵ However, recently marsupialisation or decompression and secondary enucleation have been discussed by some authors as the first line of treatment option for KCOT.¹⁶ It has been stated that marsupialisation decompression leads to significant reduction in size and thickening of the lining of the lesion after which a secondary enucleation can be done easily with decreased morbidity, thus preserving associated vital structures such as the inferior alveolar nerve and developing teeth, especially in young children where teeth and bone are still developing. August *et al*¹⁷ in their study have demonstrated epithelial dedifferentiation and loss of keratinisation with decompression and irrigation. However, there are few limitations of decompression as it requires patient compliance, may fail to completely eradicate lesion or there may be some malignant changes in the cavity which may go undiagnosed, and this technique may require secondary enucleation.¹⁸ The resolution of KCOTs after marsupialisation is also considered by some to be non-compatible with a neoplastic process and one of the reasons of reclassifying the lesion to the cystic category in 2017.¹⁴

KCOTs present most frequently in second and third decades of life with the male predilection.³ In the present case, the child affected was male and of 11 years of age. In the present case, considering the age of the child and to preserve the permanent tooth, conservative management of the lesion with decompression only was opted for. Copious irrigation and packing of the cavity with iodoform gauze reduced the risk of infection and recurrence. A surgical stent made up of self-cure acrylic resin with acrylic extension inside the cystic lumen was used to maintain the patency of surgical opening to allow drainage of cystic fluid. It also prevented the foreign bodies from entering the

Learning points

- ▶ Treatment of keratocystic odontogenic tumours (KCOTs) should be based on the age of the patient, size and location of the lesion, history of any recurrence and histological variant.
- ▶ Decompression as the conservative management of KCOTs should be considered as it reduces morbidity and preserves the vital tissues, especially in young children.
- ▶ Patient compliance is the key factor in determining conservative treatment option.

cystic lumen. The patient was trained to irrigate through this surgical opening using syringe and needle two times per day as a part of his oral hygiene. Regular recall visits are important. The successful management of present case using decompression and intraoral appliance without any recurrence after 5-year follow-up emphasises the need to stress on the conservative treatment options for the management of KCOTs, especially in children.

Contributors MR: Involved in writing of the case report and patient care. SKB involved in writing of the report and intervention/performing the surgery. AG involved in writing of the report and planning the treatment. PG involved in patient care.

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REFERENCES

- Phillipsen H. On keratocysts in the jaws. *Tandeleageblad* 1956;60:963–81.
- Phillipsen HP. Keratocystic Odontogenic Tumour. In: Barnes L, Eveson JW, Reichart P, Sidransky D, eds. *Pathology and genetics of head and neck tumours*. Lyon: World Health Organisation classification of tumours, IARC. 2005.
- Stoelinga PJ. Long-term follow-up on keratocysts treated according to a defined protocol. *Int J Oral Maxillofac Surg* 2001;30:14–25.
- Bataineh AB, al Qudah M. Treatment of mandibular odontogenic keratocysts. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1998;86:42–7.
- Zecha JA, Mendes RA, Lindeboom VB, et al. Recurrence rate of keratocystic odontogenic tumor after conservative surgical treatment without adjunctive therapies - A 35-year single institution experience. *Oral Oncol* 2010;46:740–2.
- Irvine GH, Bowerman JE. Mandibular keratocysts: surgical management. *Br J Oral Maxillofac Surg* 1985;23:204–9.
- Maurette PE, Jorge J, de Moraes M. Conservative treatment protocol of Odontogenic Keratocyst: a Preliminary Study. *J Oral Maxillofac Surg* 2006;64:379–83.
- Morgan TA, Burton CC, Qian F. A retrospective review of treatment of the odontogenic keratocyst. *J Oral Maxillofac Surg* 2005;63:635–9.
- Kolokythas A, Fernandes RP, Pazoki A, et al. Odontogenic keratocyst: to decompress or not to decompress? A comparative study of decompression and enucleation versus resection/peripheral ostectomy. *J Oral Maxillofac Surg* 2007;65:640–4.
- Brannon RB. The odontogenic keratocyst. A clinicopathologic study of 312 cases. Part II. Histologic features. *Oral Surg Oral Med Oral Pathol* 1977;43:233–55.
- Mendes RA, Carvalho JF, van der Waal I. Biological pathways involved in the aggressive behavior of the keratocystic odontogenic tumor and possible implications for molecular oriented treatment - an overview. *Oral Oncol* 2010;46:19–24.
- de Vicente JC, Torre-Iturraspe A, Gutiérrez AM, et al. Immunohistochemical comparative study of the odontogenic keratocysts and other odontogenic lesions. *Med Oral Patol Oral Cir Bucal* 2010;15:e709–e715.
- Yildirim G, Ataoglu H, Kalayci A, et al. Conservative treatment protocol for Keratocystic Odontogenic Tumour: a follow-up Study of 3 Cases. *J Oral Maxillofac Res* 2010;1:e7.
- Soluk-Tekkeşin M, Wright JM. The World Health Organization classification of Odontogenic Lesions: a summary of the changes of the 2017 (4th) Edition. *Türk Patoloji Derg* 2017;33:1–18.
- Meningaud JP, Oprean N, Pitak-Arnop P, et al. Odontogenic cysts: a clinical study of 695 cases. *J Oral Sci* 2006;48:59–62.
- Pogrel MA, Jordan RC. Marsupialization as a definitive treatment for the odontogenic keratocyst. *J Oral Maxillofac Surg* 2004;62:651–5.
- August M, Faquin WC, Troulis MJ, et al. Dedifferentiation of odontogenic keratocyst epithelium after cyst decompression. *J Oral Maxillofac Surg* 2003;61:678–83.
- Marker P, Brøndum N, Clausen PP, et al. Treatment of large odontogenic keratocysts by decompression and later cystectomy: a long-term follow-up and a histologic study of 23 cases. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1996;82:122.

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