Conservative management of multiple keratocystic odontogenic tumours in a child with Gorlin-Goltz syndrome: a case report

C. WILSON, M. MURPHY

Abstract: Background The recommendations regarding the management of keratocystic odontogenic tumour (KCOT) vary widely in the literature. The authors highlight that conservative surgical management should still be considered in some cases. Case report A young patient with Gorlin-Goltz Syndrome and two large mandibular KCOTs is presented. The case demonstrates conservative treatment with enucleation of the tumours and preservation of all involved teeth. Subsequent orthodontic treatment resulted in all teeth erupting successfully. The tumour cavities regenerated with bone.

Keywords: Keratocystic odontogenic tumour; Conservative management; Gorlin-Goltz.

Introduction

The recommendations regarding the management of keratocystic odontogenic tumours (KCOTs) vary widely in the literature. This is a case report of a young patient, who was conservatively treated, with a successful outcome; highlighting that conservative surgical management should still be considered in some cases.

Gorlin-Goltz syndrome was defined by Robert J. Gorlin and Robert W. Goltz in 1960 [Gorlin, 1987]. It is also known as basal cell nevus syndrome (BCNS).

It is an autosomal dominant inheritable disease, with no sex predominance.

One of the common findings of this syndrome is multiple keratocystic odontogenic tumours of the jaws.

The treatment of KCOTs varies, with some surgeons favouring an aggressive approach because of the tendency of these tumours to recur [Williams and Connor, 1994]. The terminology has recently been updated by the WHO, due to the aggressive nature of these lesions, behaving more like a tumour, rather than a cystic lesion [Madras and Lapointe, 2008].

Case report

T.G. is a 12-year-old boy who was referred by his general dental practitioner to the Department of Orthodontics, at the Royal Gwent Hospital.

History and examination revealed Gorlin-Goltz Syndrome, learning difficulties, frontal bossing and previous hydrocephalus not requiring a shunt. Family history revealed a father with Gorlin-Goltz Syndrome. The diagnosis of Gorlin-Goltz Syndrome in TG was made at age 11 based on the family history, his previous medical history and subsequent genetic testing.

An orthodontic and clinical assessment showed:
- Class II, division 1 malocclusion.
- Mild crowding in the upper arch.
- High Frankfort mandibular plane angle and increased lower face height.
- Buccal expansion in the anterior and left body of the mandible with displacement of the anterior teeth.

Radiographic investigation revealed (Fig. 1):
- Full complement of teeth in the upper arch.
- Congenitally missing lower right second premolar (45), and retained lower right deciduous second premolar (85).
- Unerupted lower left canine (33).
- Two large unilocular radiolucencies in the
C. WILSON AND M. MURPHY

mandible; one in the right body and one in the anterior mandible extending from the right premolar (44) to the left molar (36) region displacing the involved teeth.

Treatment

From the history of Gorlin-Goltz syndrome a clinical diagnosis of KCOTs was made. Treatment initially involved surgical management of the cystic lesions followed by conventional orthodontic treatment to align the dental arches.

Under general anaesthetic, buccal mucoperiosteal flaps were raised and the mental nerves identified and protected. Bone was carefully removed to expose the cystic tumours which were then enucleated. The lower left canine (33) was found to be almost devoid of bony support within the tumour cavity. A decision was made to preserve this and all the other involved teeth but without compromising removal of the tumour linings. Primary closure was achieved. The upper right first premolar (14), upper left first premolar (24) and lower right second deciduous molar (85) were also extracted, as part of the orthodontic treatment plan.

The post operative recovery was uneventful and the patient was discharged the following day on a course of antibiotics and appropriate analgesia.

At subsequent review primary healing had occurred without complication, and histopathology confirmed the clinical diagnosis of KCOTs.

Fixed appliance orthodontic treatment commenced six weeks following surgery. Two months later, the lower left canine (33) became visible, and the following month a bracket was applied (Fig. 1b).

FIG. 1 - A: radiograph showing two large KCOT's. B: splayed appearance of teeth. C: treatment with a lower fixed appliance.

FIG. 2 - A: radiograph following enucleation of KCOT's and fixed orthodontic appliances. B, C: alignment of teeth.
In just under a year, the teeth were aligned, and the appliances removed. Following 8 months of retention, the treatment was complete. The anterior open bite is accepted, and the patient is pleased with the result. There is the option of orthognathic surgery in the future, should the patient wish.

At the last surgical review, two years post surgery the affected teeth were asymptomatic and well aligned, except for some rotation of the lower left canine (33). Vitality testing revealed positive responses to all teeth adjacent to the lesions. The radiograph showed good bony regeneration of the cystic tumour cavities, and no signs of recurrence (Fig. 3).

Given the nature of Gorlin-Goltz syndrome and KCOTs this patient will be kept on long term review.

Discussion

There continues to be two schools of thought in the surgical management of KCOTs. A number of clinicians favour conservative treatment [Meiselman, 1994], whilst others advocate aggressive treatment based on the risk of recurrence of these tumours, which is on average 30% when treated with enucleation alone [Madras and Lapointe, 2008]. KCOT’s also have the potential for aggressive behaviour, such as extension and persistence in adjacent structures including soft tissues [Stoelinga, 2001], and the ability to undergo transformation into ameloblastoma or malignancy [Williams and Connor, 1994]. An aggressive approach to treatment includes mechanical curettage, chemical curettage using Carnoy’s solution, the sacrifice of involved and adjacent structures including overlying mucosa, peripheral ostectomy and in some cases resection with a margin of normal tissue depending on the site of the tumour [Stoelinga, 2001; Dammer et al., 1997; Morgan et al., 2005].

The disadvantages of an aggressive approach to treatment in this case would have been:
- Loss of the involved teeth
- Long term loss of sensation in the distribution of the inferior alveolar nerve
- Potential facial disfigurement.

The conservative approach includes enucleation with or without local curettage and marsupialisation either alone or with subsequent enucleation of residual tumour [Meiselman, 1994; Madras and Lapointe, 2008]. This minimises the negative effects associated with aggressive surgery, safeguarding the delicate anatomical structures of the jaws.

When considering KCOTs associated with Gorlin-Goltz syndrome as opposed to sporadic cases some differences should be taken into account. In Gorlin-Goltz Syndrome, KCOTs tend to be multiple occurring concurrently or synchronously, occur at a younger age (as illustrated in this case), reportedly have a greater tendency to recur [Dominguez, 1988] and may have enhanced neoplastic properties based on heparanase gene and protein expression [Katase et al., 2007].

This leads to some contradictions when considering appropriate surgical treatment for these cases. There is a pressure to adopt a conservative approach because the patient is young as in this case and may develop further tumours in the future, yet there is a contradictory pressure to adopt an aggressive approach because the potential for recurrence is high and the neoplastic properties may be enhanced.

In this case a conservative approach was adopted in order to preserve the affected teeth, particularly the lower left canine which was unerupted and almost devoid of bone following enucleation of the KCOT. This approach has been successful in that at two years post surgery all affected teeth are erupted, in function and vital and there is no evidence of recurrence of the KCOTs. It will of course be necessary to maintain a long term follow-up of this patient in order to diagnose and treat any recurrent or new KCOTs early.

Conclusion

When planning the surgical management of KCOTS, there is a balance to be struck between reducing the risk of recurrence, whilst maintaining adjacent or involved structures.

In this case of a young patient with two KCOTs associated with Gorlin-Goltz Syndrome, conservative management was favoured with preservation of the involved teeth, the inferior alveolar and mental nerves and the continuity of the mandible, while still achieving complete removal of the cysts. He remains...
C. WILSON AND M. MURPHY

disease free at two years post surgery.
Recent publications concentrating on sporadic KCOTs tend to favour an aggressive approach to treatment with at least the use of Carnoy’s solution in addition to enucleation. This case illustrates that a conservative approach should still be considered particularly with KCOTs associated with Gorlin-Goltz Syndrome in young patients with unerupted and affected teeth.

Acknowledgements
Thanks to James Dean for providing orthodontic treatment.

References