

Dental Management of Severe Malocclusion and Syndromic Multiple Odontogenic Keratocysts

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Abstract

The rare Gorlin-Goltz syndrome is an autosomal dominant disorder. One of its primary characteristics is numerous odontogenic keratocysts (OKC). Whilst the OKC are benign, they are recurring and locally aggressive cysts that can affect the maxillary and mandibular jaws but are most frequently observed in the posterior part of the mandible from ages 10 to 40 years. Dental management of this condition frequently necessitates surgical intervention and subsequent orthodontic consultation.

This case report presents the multidisciplinary clinical management of a patient (male aged 18) experiencing teeth malocclusion further affected by numerous OKC due to a diagnosis of Gorlin-Goltz syndrome. This case demonstrated positive bone healing following the surgical intervention for the OKC and teeth malocclusion treatment employing a comprehensive orthodontic treatment.

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Introduction

Gorlin-Goltz syndrome, also referred to as nevoid basal cell carcinoma syndrome, is a rare autosomal disorder. Characteristics include numerous OKC, multiple basal cell carcinoma, and other abnormalities (both skeletal and non-skeletal). This disorder affects approximately 1 of every 60,000 births, with the genders equally impacted¹.

Epidemiologically, OKC and other developmental odontogenic cysts are more common in males than females and account for approximately 0.8% to 45% of the diagnosed oral lesions^{2,3}. OKC are also far more common in adolescents than children, compared to other developmental odontogenic cysts such as dentigerous cysts, where they are more common in children⁴.

Philipsen (1956) initially introduced OKC⁵. This cystic lesion could occur in isolation or as a

part of Gorlin-Goltz syndrome. It is believed that it originates from the dental lamina remnants remained after tooth development is completed⁶. Whilst benign, the OKC are recurring, locally aggressive cysts that can affect both jaws but are most frequently observed in the posterior part of the mandible from ages 10 to 40 years. Skeletal destruction of the jaws and teeth displacement resulting in the malalignment of the teeth is a typical manifestation of OKC. Regarding surgical intervention and management of OKC, treatment ranges from minimally invasive approaches to radical jaw resections⁷.

The field of orthodontics centers on the treatment of orofacial abnormalities including malocclusion⁸⁻¹⁰. Frequently, other orofacial irregularities (for instance, OKC) are discovered inadvertently during routine orthodontic radiographic examinations¹¹. This case report presents the multidisciplinary clinical management of a patient experiencing teeth malocclusion that is exacerbated by numerous OKCs due to a diagnosis of Gorlin-Goltz syndrome.

Case Presentation

A patient (male aged 18) was referred to the Orthodontic Specialist Clinic for a consultation due to teeth crowding and anterior

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and buccal crossbite. The patients' chief complaint was "my upper jaw is disfigured". There were no noteworthy aspects of the patients' medical history, and he had no other apparent health anomalies. The patients' clinical examination revealed a symmetric brachycephalic face on the frontal view, with thick but competent lips (Figure 1a).

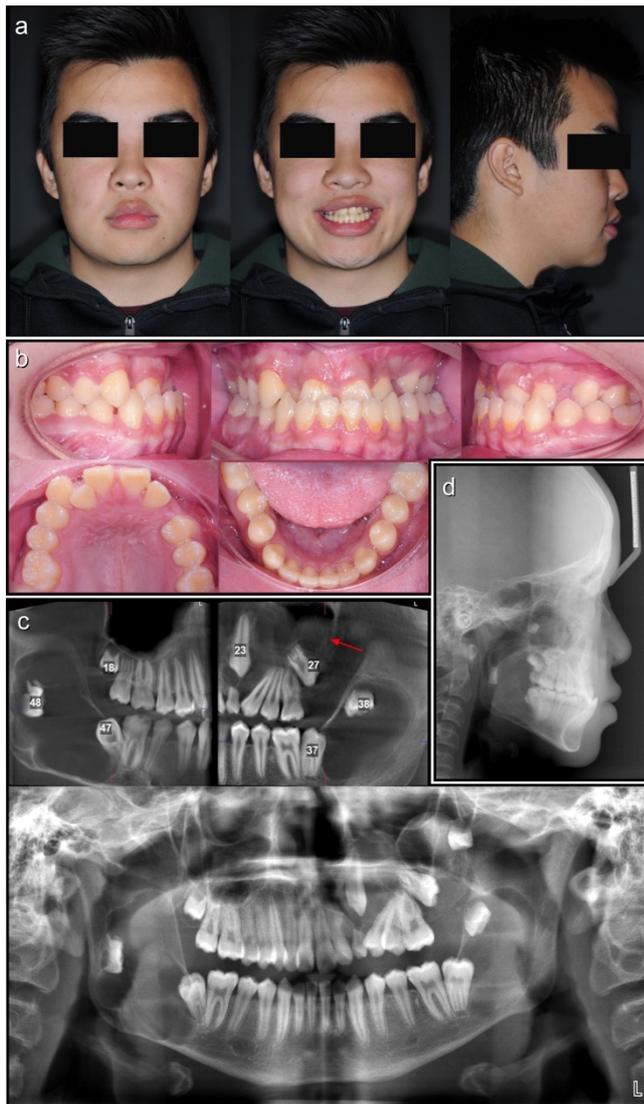


Figure 1. (a) Pre-treatment extra-oral patient photographs. (b) Pre-treatment intra-oral patient photographs. (c) Pre-treatment panoramic and CBCT radiographs illustrating the teeth displacement resulting from the OKC cystic lesions. (d) Pre-treatment lateral cephalometric radiograph.

The upper teeth midline was moved 1mm to the left in comparison to the facial midline. Furthermore, the lateral facial view demonstrates

that the patient had a straight profile with average vertical facial height.

An intraoral examination determined that the patient had average oral hygiene level and moderate gingival health (Figure 1b). Teeth occlusion was evident in an Angle Class I malocclusion, with a leaning towards Class III malocclusion on the left. The maxillary anterior teeth were observed to be in a crossbite. Moreover, tooth number 16 and the entire left posterior teeth segment number 24-26 were in buccal crossbite. An increase was noted in teeth overbite (7mm), while teeth overjet was negative. The facial and lower teeth midlines coincided. In addition, upper teeth crowding and lower teeth spacing were observed, along with a number of missing teeth.

A panoramic and CBCT radiographic assessment established the presence of numerous clearly defined, multilocular radiolucent lesions impacting the body of the maxilla and mandible and also displacing unerupted upper-left permanent canine and second molar, and upper and lower right and left permanent second and third molars (Figure 1c). Additionally, the cysts displaced the roots of the upper-left first and second premolars, and lower right and left permanent second molars. The initial diagnosis for the radiolucent lesions was OKC. A lateral cephalogram examination revealed that the patient had a prognathic mandible with skeletal pre-normal relationship (Figure 1d). Both the upper and lower central incisors were found to be retroclined, with a forward rotation of maxillary and mandibular planes. Furthermore, the soft tissue profile was observed to be concaved.

The patient was firstly referred to the Oral and Maxillofacial Surgery clinic so the cystic lesions could be diagnosed and enucleated. The OKC diagnosis was confirmed by a histopathologic examination of the cysts. In addition, the Gorlin-Goltz syndrome diagnosis was verified via genetic examination of the PTCH1 exons and their intron-exon borders. The Oral and Maxillofacial surgeon enucleated the mandibular jaws' OKC lesions, while conducting a Lefort I osteotomy in the maxilla so as to uncover and enucleate the OKC. Subsequently, a rigid internal fixation was performed. A panoramic radiographic assessment carried out six months post-surgery showed that the mandibular and maxillary jaws were healing well

(Figure 2a). Regular follow ups were conducted with the patient post-surgery in order to monitor any radiographic indications of the cystic lesions' relapsing (Figures 2b & 2c). This course of action was taken due to the aggressiveness of the OKC and their significant risk of relapse.



Figure 2. (a) Panoramic radiograph six months post-surgery for the removal of the OKC lesions. (b) and (c) Panoramic radiograph 12 months and 24 months post-surgery, respectively.

Follow-ups were conducted for two years post-surgery, and there were no apparent indications of the OKC lesions relapsing. It was noted that the patient had established good oral hygiene practices. Hence, complete orthodontic treatment began with a non-extraction plan comprised of the following goals:

1. Resolve the upper teeth crowding and lower teeth spacing
2. Resolve the anterior and unilateral left posterior crossbite

3. Establish Class I molar and canine relationship
4. Establish normal overjet and overbite
5. Preserve a space for future implant in the position of tooth number 23
6. Establish permanent retention of the orthodontic treatment outcomes

This orthodontic treatment began while the patient remained under the care of the Oral and Maxillofacial Surgery clinic, where he was constantly monitored for relapse of the cystic lesions.

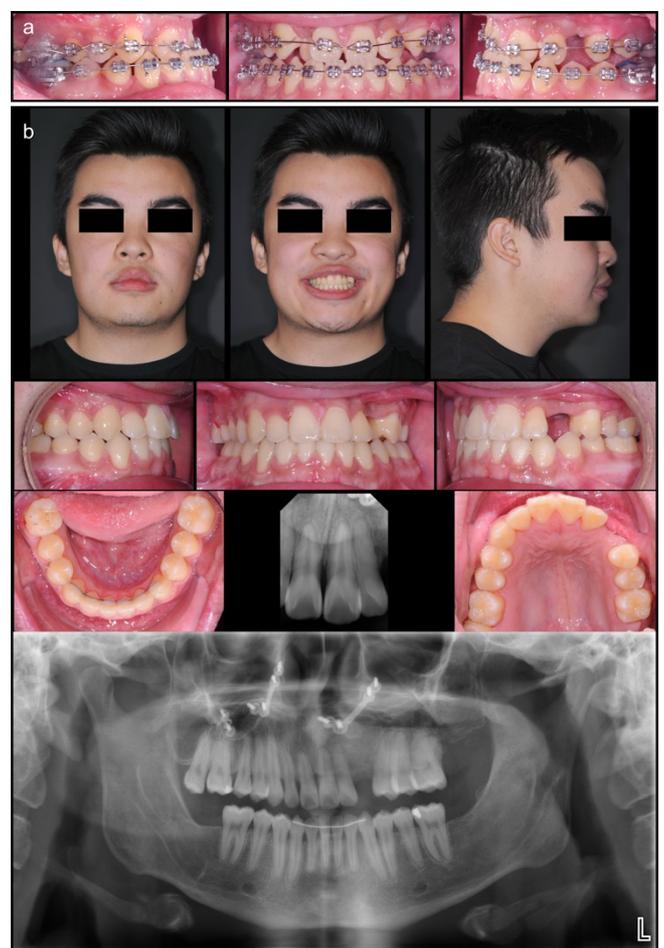


Figure 3. (a) MBT brackets bonded on the upper and lower teeth. (b) Extra- and intra-oral photos as well as the routine radiographic records taken after the removal of the orthodontic brackets.

Fixed upper and lower MBT brackets were bonded with bite blocks to allow bonding the upper anterior teeth (Figure 3a). Firstly, soft nickel titanium wires were utilized to align the teeth and enable forward inclination of the upper anterior teeth so as to rectify the crowding issue.

Nickel titanium spring coils were employed to open the space for tooth number 12 and preserve an adequate implant space for missing tooth number 23. Twelve months after the orthodontic treatment began, heavy rectangular stainless-steel wires were utilized to fully express the brackets' 3D prescription, close the teeth spaces in the lower jaw and to expand the upper-left posterior segment. This action placed the upper anterior teeth in a positive sagittal position, with the overjet and overbite at normal levels. Subsequently, short-term Class III elastics were utilized to rectify the sagittal relationship in the left side, and finally, teeth occlusion detailing and finishing were performed. Overall, the entire orthodontic treatment took place over a duration of twenty-four months. The patient was provided with an upper vacuum-formed retainer for permanent use at night-time, and a stainless-steel fixed retainer bond on the lingual surfaces of the lower anterior teeth (Figure 3b).

Following the orthodontic treatment, the patient was regularly monitored in the Oral and Maxillofacial Surgery clinic in order to check for a potential relapse of the cystic lesions and carry out any required management. The patient was also offered a consultation with a periodontist for an implant in position of tooth number 23.

Discussion

Previous studies have reported the influence of oral cystic lesions on teeth eruption and alignment¹²⁻¹⁵, while the multidisciplinary management of OKC is scarce in literature. This case report details the multidisciplinary clinical management of a patient (male aged 18) experiencing teeth malocclusion and numerous OKC as a result of Gorlin-Goltz syndrome. The patient had Angle's Class I malocclusion with upper teeth crowding and numerous teeth displacements due to the OKC. Initially, the treatment entailed surgical enucleation of the OKC, and subsequently, a comprehensive orthodontic treatment was provided.

During early childhood, it is important to perform routine dental radiographic examinations in order to identify any abnormalities in teeth formation, eruption, and position, and any pathological lesions. In this patients' case, the OKC lesions were inadvertently discovered during a routine radiographic orthodontic assessment. At that point, the patient was in

early adulthood, but it is possible that an earlier diagnosis of the OKC lesions could have reduced the bone damage and tooth loss experienced.

In this case, the cystic lesions were entirely enucleated along with the other teeth involved. Another adjunctive method entails the use of marsupialization (which is deroofting of the lining of the cyst) in order to decrease the intra-cystic pressure resulting in cyst decompression and followed thereafter by cystic enucleation¹⁶. This method is particularly beneficial for pediatric patients as it maintains the bony structure and enables the impacted teeth to erupt. However, this approach was not followed in this case due to the patients' age.

As discussed, OKC are aggressive cystic lesions with high relapse risk, varying from 3% to 60%¹⁷. Many theories have been put forward to explain this high rate, including the partial removal of the delicate cystic lining, cystic lining outpouching, and satellite cells originating from the odontogenic epithelial remnants resulting in new OKC lesions forming⁷. Hence, it is essential that periodic radiographic monitoring of patients treated for OKC is performed in order to monitor for any potential cystic lesion relapse. If this occurs, the required management should be undertaken.

In this case, the orthodontic treatment outcomes were satisfactory despite being complicated by the Gorlin-Goltz syndrome. The usual treatment retention protocol was undertaken. The patient was provided with a maxillary removable vacuum-formed retainer for night-time usage, and a metallic retainer was bonded to the lingual surfaces of the lower anterior teeth.

Conclusions

OKC lesions are a primary feature of Gorlin-Goltz syndrome. Whilst the OKC is benign, it is recurrent and locally aggressive. This can impact the maxillary and mandibular jaws and also displace the dentition. Dental management of this condition frequently necessitates surgical intervention and subsequent orthodontic consultation. This case demonstrated positive bone healing following the surgical intervention for the OKC and teeth malocclusion treatment employing a comprehensive orthodontic treatment.

Declaration of Interest

The author declares that there are no conflicts of interest.

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