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Management of a patient suffering with Cherubism with Dental Implants

Karun Dewan* and Karl Bishop†

Abstract - *Cherubism is a rare non-neoplastic, fibro-osseous hereditary disorder characterized by bilateral expansion of the maxilla and mandible producing a characteristic facial appearance. It can affect the facial and dental growth of the individual and often results in gross aesthetic and functional deficiencies. The teeth may also be displaced or submerged and these problems can often compromise successful restorative rehabilitation. This paper describes the restorative management of an adult patient with Cherubism involving a fixed implant retained mandibular restoration. The care utilized 3D planning software and implant insertion guides to facilitate an early loading protocol and the use of optimum bone quality/volume areas.*

KEY WORDS : Cherubism, 3D planning, Implants, Adult

INTRODUCTION

Cherubism is caused by an autosomal dominant gene located on chromosome 4p16.3, and typically affects males with 2:1 predominance. The pathology is typically benign in nature beginning at the age of 2 to 3 years and progressing into childhood[2,3]. It was first described by Jones in 1933 and since then many cases have been reported in the literature [2,4-9].

The pathogenesis of Cherubim remains controversial. It is considered as a benign self-limiting fibro-osseous disorder and has been postulated to be a manifestation of fibrous dysplasia and a giant cell granuloma[10,11,12,13]. A molecular pathogenesis by means of SH3BP2 gene mutations have been proposed leading to disregulation of the Msx-1 gene involved in regulating mesenchymal interaction in craniofacial morphogenesis[2].

The diagnosis of Cherubism is based on clinical, radiographical and histological findings[2]. The mandibular angle, ascending ramus, retromolar region and posterior maxilla are the most often affected sites. The pathology may also involve the coronoid process but the condyles are often spared. The classical clinical presentation is that of a painless, firm and symmetrical enlargement of the mandible and maxilla resulting in fullness of the cheeks[3,4]. However, the physical manifestations are highly variable from a barely discernible posterior swelling of the maxilla or mandible to a marked expansion of both resulting in masticatory, speech and swallowing difficulties[13-18]. Radiographic bony changes are classical for this disease and often provide the only evidence of the problem[19].

The classical histological picture of Cherubism shows a loose, highly vascular, fibrous stoma with a moderate number of unevenly distributed multinucleated giant cells of osteoclastic nature[2]. Therefore, in addition to the typical histological features, the classical clinical manifestations, radiographic features and the familial occurrence of the disease are required for a definitive diagnosis[20,21]. Teeth are often displaced or unerupted and radiographically may appear to be 'floating' in the bony cyst- like spaces.[19,20,22,23,24]

Traditionally, the general approach to management has been to follow patients, perform biopsies, remove ectopic and erupted teeth, and provide surgical correction when appropriate [2-5,8,12 2,15,23,25]. Surgical treatments advocated involve contouring of the expanded lesions and complete curettage[22]. Liposuction has also been proposed to reduce the mass of the lesion in some cases[26]. Although radical surgery has been described it is often contraindicated when both the mandible and maxilla are affected bilaterally[2]. In addition, poor results and a tendency to relapse following surgical intervention have been reported, particularly when surgery is initiated as an early intervention[23,26]. For these reasons monitoring of the condition is often carried out since the disease process characteristically regresses during puberty with spontaneous bone recontouring occurring through third decade of life. This results in the gradual improvement in the altered facial contours[9].

Descriptions of the restorative rehabilitation of patients with Cherubism is very limited with only one case reported involving the construction of a removable mandibular overdenture in a 21-year-old male[19]. The use of osseointegrated implants in cases of Cherubism has not been previously reported. This may be due to the observation that systemic and local factors that affect bony healing such as metabolic bone diseases could have a negative outcome on osseointegration[27]. In addition the displaced and submerged position of the teeth can complicate the positioning and timing implants[19].

This paper presents a clinical case of a patient with known familial history of Cherubim restored with an implant retained fixed bridge using and 3D planning software. This approach facilitated good initial stabilization, optimization of implant sites and the use of an early loading protocol.

* BDS, MFDS RCS, LDS RCS, MSc
† BDS, MScD, FDS, DRD, MRD, FDSRCS (Rest Dent), LLM

Figure 1. *Extra-oral view showing enlarged mandible*

CASE REPORT

A 43-year-old female suffering with Cherubsim was referred by her general dental practitioner to the Department of Restorative Dentistry. The patient had a family history of Cherubism with one of her siblings and her daughter also affected. The patient also reported a history of ocular involvement for which she underwent corrective surgery during childhood.

The patients' physical appearance showed an enlarged mandible with a skeletal class III dental base relationship and an obtuse gonial angle (Fig 1). From the history it was evident that the facial and bony changes were stable. Clinical examination revealed the presence of a number of missing teeth in both the mandible and maxilla.

The mandibular teeth present were atypical in shape, spaced and irregularly aligned (Fig. 2a) with a number of retained deciduous teeth retained. Occlusal examination revealed a class I incisor relationship and a unilateral cross bite on the left. Radiographs showed that the typical widespread irregularly multilocular radiolucent areas were filled with osseous structure at the time of examination although a comparison was not possible. The radiographs also highlighted that the erupted teeth in the mandible had short and malformed root morphologies or evidence of resorption.. Unerupted teeth were present anteriorly.

The patient wore a removable partial cobalt-chromium mandibular prosthesis and expressed concerns about the malformed and displaced mandibular teeth. She was also unhappy with the overall aesthetics achieved with the existing partial denture (Fig. 2b).

Following an initial assessment, it was clear that the patient's aesthetic concerns mainly involved the mandibular arch and as the maxillary dentition was largely unaffected the treatment options considered were:

- The construction of a mandibular partial/complete overdenture using strategic use of the patient's natural teeth as abutments.
- Fabrication of conventional a mandibular fixed bridge using the remaining natural teeth as abutments with possible orthodontic alignment.
- Extraction of all remaining erupted and unerupted teeth and provision of a complete conventional or implant retained mandibular prosthesis.

Following discussions with the patient and in light of her experiences with the existing partial denture, it was decided that any treatment that involved a removable option would not be acceptable. A conventional fixed mandibular prosthesis was also excluded following a diagnostic wax-up due to the limited aesthetic changes it facilitated and the poor condition of the potential abutments. Orthodontic movement of the remaining natural teeth to create more favourable spacing was also considered inappropriate due to the presence of deciduous teeth and its unacceptability to the patient. The initial definitive treatment plan was:

- The construction of a mandibular immediate complete denture at the existing occlusal vertical dimension following the loss of all remaining erupted and unerupted mandibular teeth.
- Restoration of the mandibular arch following healing with dental implants and a fixed bridge.

The erupted and unerupted mandibular teeth were extracted under a general anesthetic and an immediate complete prosthesis inserted with a soft lining (Coe-comfort, GC America Inc., Illinois) to assist healing. The patient was then reviewed regularly and the denture relined to compensate for the alveolar bone changes.

Figure 2a. *Intra-oral view showing irregularly shaped, spaced, malaligned mandibular teeth*

Figure 2b. *Intra-oral view with mandibular partial denture insitu*

A copy of the mandibular prosthesis was constructed in clear heat cured acrylic to use as a stent to assist in 3 dimensional treatment planning. Using the Nobel Guide Protocol (Nobel Boicare, UK Ltd.) a CT scan was obtained approximately 8 weeks following the extractions. Using the computer based Nobel Guide planning software (Nobel Boicare, UK Ltd.); a surgical stent was constructed to aid the placement of 16 mm regular bodied implants and an immediate temporary bridge (3a & 3b). The choice of implant site and angulation was determined not only the optimum position in relation to the prosthesis but also in relation to the bone density[28].

Four dental implants (Replace Select) were subsequently inserted following the Noble Guide protocol under a local anasethetic. Fixture head impressions were obtained and the cast verified and centric jaw registration recorded at the same visit (Fig. 4) the interim bridge was then inserted. Three weeks later the definitive bridge was fitted. (Fig. 5a to 5c)

The patient has been followed-up for approximately 3 years without any adverse events.

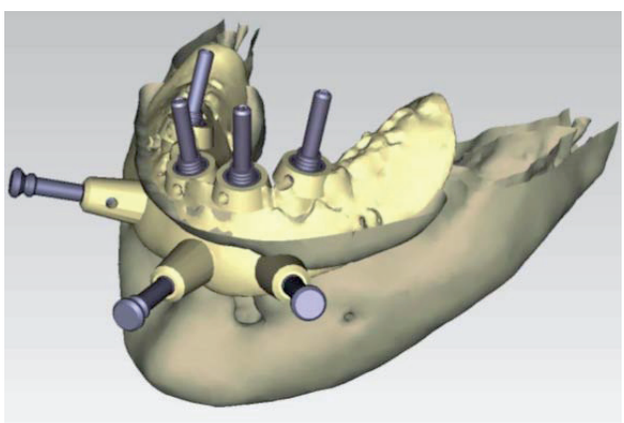

Figure 3a and b. *Surgical stent to assist in implant surgery and screen shot of Nobel Biocare software*

Figure 4. *Demonstrating fixture head impression copings of inserted implants*

DISCUSSION

A Cherub patient clinically presents as a challenge to clinicians' not only from the maxillo-manidublar skeletal deformity point of view but also due to the fact that they suffer with malaligned, unevenly spaced, ectopically erupted, and unerupted teeth.

The use of implants to restore a patient suffering with Cherubism has not been described in the literature previously. Yilmaz[19] described a prosthetic treatment approach for a Cherubism patient with a removable partial denture in the maxilla and a mandibular overdenture using erupted teeth as abutments. Although an implant option was considered in the treatment, authors were concerned that implant placement into porous bone might result in lack of osseointegration and complications.

However in the current case, based on the history, the patient was likely to be in the stable phase of the problem[9]. This appeared to be confirmed from the radiographic examinations which showed bony in-fill in previously resorbed areas. As such, treatment with dental implants was then considered a viable option providing the optimum conditions were present or could be developed.

Computer-guided flapless surgery for implant placement using CT guided 3-dimensional technique was used as it had the advantage of being minimally surgically invasive. It also allowed accurate implant placement into sites where good bone quality and quantity was predicted[28].

Brånemark[29] in 1977 described conventional implant protocols, which were based on the achievement of primary stability and prolonged, non-loaded healing periods. Conventional implant protocols based on clinical experience advocates loading of implant fixtures once osseointegration is complete with an unloaded period of three to six months through a two stage technique [29]. The arguments against early loading are based on the belief that the transfer of any micro-motion to the implant surface during healing would result in fibrous encapsulation rather than osseointegration[30]. However, there is a growing body of published literature supporting reduced implant loading times[31-35]. This is based on evidence that immediate loading of dental implants does not necessarily cause untoward effects on the formation of mineralized tissue at the interface. The evidence suggests that immediate loading may actually result in a higher bone volume deposition around the implant fixture at an early stage when compared to the traditional two stage procedure[36,37,38]. This is especially the case with prosthesis that utilize cross arch stabilization[32,35].

Figure 5a &b. *Illustrates definitive bridge in situ*

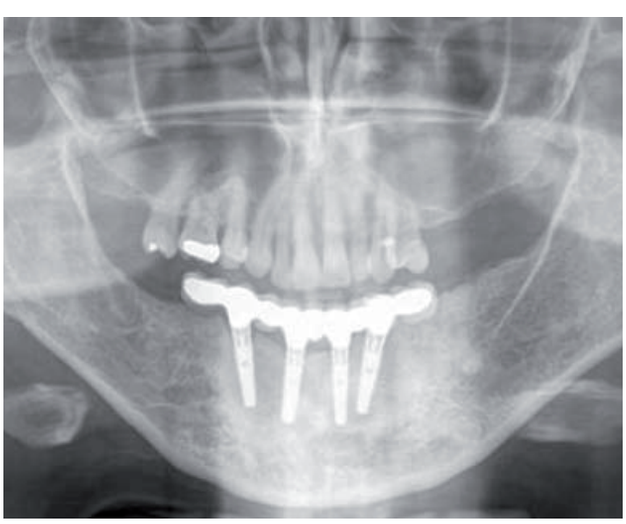

Figure 5c. *Demonstrating implants insterted at good bone sites with cross arch definative bridge*

These findings have also led to a number of 'early loading' protocols being described with a functional restoration being placed from between 48 hours to 3 months after the implant is placed[31]. An early loading protocol was chosen in this case as the perceived psychological, and functional advantage of shortened treatment periods was considered an advantage. In addition, there was a high initial stability of the fixtures as reflected by the insertion torque being greater than 40Ncm for all implants[31]. In addition cross arch stabilization was provided at the time of implant insertion by the use of an immediate interim bridge.

CONCLUSION

Cherubism is a rare, fibro-osseous lesion of the jaws that may have dental manifestations that could affect successful dental rehabilitation. This paper highlights that Cherubism patients' could be treated with dental implants if treatment is timed correctly. The use of 3-dimensional planning facilitates optimum and precise implant placement which in turn to allows the implants to be placed in areas with appropriate bone quality and volume. The technique may also allow early loading protocols to be utilized. This technique also has the potential to reduce the need for extraction of unerupted teeth.

ADDRESS FOR CORRESPONDENCE

Karun Dewan BDS, MFDS RCS, LDS RCS, MSc, Specialist Registrar and Honorary Clinical Lecturer in Restorative Dentistry, School of Dentistry, Birmingham Dental Hospital, St Chad's Queensway, Birmingham. B4 6NN. E:mail: karun.dewan@sbpct.nhs.uk

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