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Case Report

Orthodontic Treatment in a Beta-Thalassemia Major Patient

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A B S T R A C T

Surgical procedures are the most popular choice of treatment for beta-thalassemia major patients. Although these procedures could be life-threatening, orthodontic treatment modality has received little or no attention in the past five decades. Only two case reports have described the treatment of these patients with fixed orthodontics in conjunction with orthopaedic appliances. The following report describes a combination of orthodontic and functional appliances for the treatment of moderate dentofacial deformities in an adolescent male patient suffering from beta thalassemia major.

Keywords: Beta Thalassemia, Flared Maxillary Incisors, Class II Skeletal Base, Blood Transfusion, Functional Appliance

Introduction

Numerous reports exist in the literature describing the craniofacial features of beta (β) thalassemia major patients.[1,6] Surgical correction remains the popular choice of treatment for patients with this deformity. Authors have mentioned various surgical procedures such as alveolar contouring,[7] tissue resection,[8] anterior maxillary osteotomies, Le Fort I osteotomy, bilateral sagittal split ramus osteotomy, anterior mandibular segmental osteotomy, genioplasty,[9,10] and maxillary set back[11] for the treatment of the various dentofacial deformities. Although surgery involves the risk of excessive bleeding and morbidity, scant attention is given to the orthodontic treatment model.[10,12] Few authors have recommended early interceptive procedures such as maxillary growth restriction, and mandibular advancement to prevent worsening of the dentofacial deformities.[10,13]

The following case report describes the successful treatment of the pertinent oro-facial complications in an adolescent β thalassemia major patient using fixed orthodontic appliances in conjunction with light class II elastics to correct the Class II skeletal base discrepancy.

Case Report

A 13-year-old male patient was referred for orthodontic consultation regarding poor facial aesthetics and function. Their parents informed them he was a known case of β thalassemia major, diagnosed at 6 months after birth. The patient was placed under the care of a paediatrician and received fortnightly blood transfusions at a daycare facility. The patient was hailing from a far-off location and sometimes needed hospitalization for related medical issues such as an infection or fever at his native place. He frequently suffered from headaches, general body pain and fatigue. These symptoms would worsen a couple of days after transfusion. He was irregular at school and the family

spent most of their time obtaining medical treatment for both parents as well as two out of the three siblings who were anaemic and were carriers of this condition.

The cranio-oro-facial clinical examination revealed moderate symptoms of β thalassemia major, such as pale skin, frontal bossing, exophthalmos, depressed nasal bridge, pinched nares, prominent malar regions, mandibular retrognathia, flared maxillary anterior teeth, and lip trap during rest and smile (Figure 1).

Intraoral examination revealed reduced salivary flow, expanded maxillary lateral alveolar plates, discoloured teeth with the pinkish hue of maxillary left central incisor, bimaxillary proclination, Class II molar relation on the right side, Class I molar relation on the left side due to mesial rotation and drifting of mandibular left first premolar, increased overjet, closed overbite, exaggerated bimaxillary curves of Spee, missing mandibular right lateral incisor, sharp cusps and incisal edges, slightly small sized crowns, blocked out mandibular left second premolar, and scissor bite with respect to maxillary first premolars (Figure 1). The maxillary left central incisor was found to be vital and was cleared for tooth movement by an endodontist.

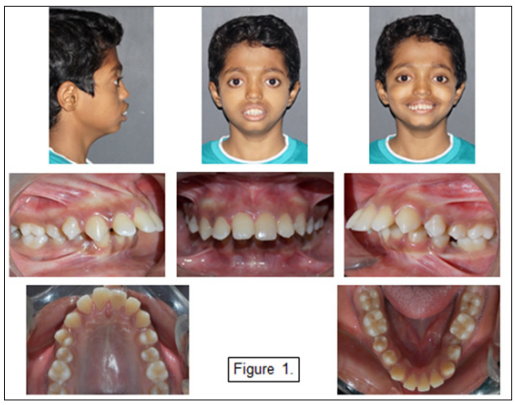

Figure 1.Pre-Treatment Extraoral and Intraoral Photographs

A panoramic radiograph and a lateral cephalogram were reconstructed from a Cone-Beam Computed Tomography (CBCT) scan. The panoramic radiograph revealed a missing mandibular right lateral incisor as well as missing third molar tooth buds in all but the fourth quadrant. Roots were generally narrow and spiky (Figure 2, A). The cephalogram, its tracing and analysis revealed a high mandibular plane angle and a retrusive mandible with a small ramal height. The ANB angle was increased (7.2°), maxillary and mandibular incisors were proclined (+1/SN; 117.2°, -1/MP-abo; 112.2°) and overjet was increased (10 mm) (Figure 2, B and C; Table 1). The control values in the Table were obtained from the published literature.[1,2,6] A hand wrist radiograph revealed deformed and rectangular-shaped metatarsals and phalanges indicating sites of extramedullary erythropoiesis in these bones (Figure 2, D).

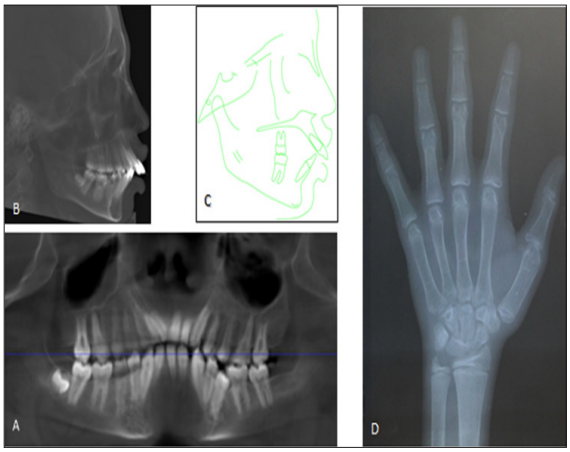

Figure 2.Pre-Treatment Panoramic Radiograph, Cephalogram, Tracing and Hand Wrist Radiograph

Table 1.Cephalometric Analysis

Measurement	Normal value	Pretreatment	Posttreatment
Skeletal Analysis			
Angle SNA (°)	80	73.8	71.1
Angle SNB (°)	82	66.6	70.9
ANB (°)	2	7.2	0.2
SN-MP-abo (°)	32	40.3	36
FMA-abo (°)	25	27.1	26
Co-A (mm)	100	84.39	81.97
Co-Gn (mm)	134	97.09	109.14
S-Go (mm)	70-75	67.95	77.42
Ar-Go (mm)	44-45	40.71	50.19
Go-Me (mm)	65-72	60.34	62.35
N-Me (mm)	116-123	108.09	113.36
ANS-Me (mm)	75	61.07	69.27
Dental Analysis			
+1i/NA (mm)	4	15	8
+1/SN (°)	103	117.2	103.1
-1i/NB (mm)	4	9	5
-1/MP-abo (°)	99	112.2	98.6
Overjet (mm)	3.5	10	3
Overbite (mm)	2	3	1
Facial Analysis			
Ls/E-line (mm)	-4	1	-1
Li'/E-line (mm)	-2	8	3

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Treatment Plan

The treatment plan was aimed at improving facial aesthetics and function, with minimal medical risks, maximum patient comfort and enhanced stability. An extraction orthodontic treatment plan combined with a removable functional appliance to advance the mandible was devised for the patient. Extraction of maxillary first premolars and mandibular left first was planned to allow space for the retraction of the flared incisors. The mandibular right first premolar would be reshaped to be substituted for the missing right lateral incisor. The mandibular right third molar may as well require extraction depending upon its eruption status. The moderate premaxillary alveolar overgrowth would be addressed by torquing the incisor roots palatially allowing the alveolar bone to resorb and relocate distally.

Light Class II elastics would be employed to gently advance and/or remodel the mandible. Elastics occupy minimal oral space, do not rest on teeth or cover the mucosa, and apply barely perceptible forces and go unnoticed by the patient. Patient compliance would not be a problem if elastic use brings about planned changes, since the patient is able to appreciate the beneficial changes that occur with respect to aesthetics and function, and remains motivated.

Treatment Progress

The patient and his parents agreed with the plan. In view of the various medical treatment needed by the patient, the appointments were agreed to be scheduled every 6-8 weeks. Extractions were carried out under antibiotic prophylaxis and were uneventful. Conventional two-step orthodontic treatment was instituted with fixed appliances (Roth Prescription). One year into treatment, working 0.019 × 0.025-in stainless steel archwires were placed for bimaxillary incisor retraction. Light Class II elastics exerting 1 (oz) (28 gms) of force were employed to advance the mandible. Transverse discrepancies were corrected by the use of expanded mandibular archwires and constricted maxillary archwires. Midline deviations in the mandibular arch were corrected by using asymmetrical elastic arch wires exerting 4-5 oz of force.

Upon application of conventional retraction forces, it was found that the roots moved suddenly while the crowns moved minimally. In order to bring about bodily tooth movement where in roots moved in tandem with the crowns, the force levels had to be reduced. This increased the overall treatment duration. In order to minimize mucosal injury initially elastic ligatures were placed, but their placement, as well as removal, was painful to the patient. In addition, it served as a nidus for plaque and calculus deposits. Hence, elastics were replaced by stainless steel ligatures, and closed coil NiTi springs for space closure were used. Caution was exercised not to cause mucosal lacerations during their placement.

Forty months into treatment, at the finishing stage, the patient requested appliance removal for consultation and possible therapy with a bone marrow transplant. The patient has been informed of the importance of achieving optimum finishing. He informed his desire to complete the treatment at a later date. Consequently, all appliances were removed. Soft vacuum-formed retainers were placed, but the patient-reported discomfort in wearing them, due to the slightly raised bite. Hence routine upper and lower Hawley retainers were fabricated and placed on the same day. Four months later after being found unsuitable for marrow transplant therapy, the patient returned for a dental check-up. At this point once again fixed appliances were placed and maximum intercuspation was obtained. Root canal treatment was performed for the maxillary left central incisor, which was now found to be non-vital. In addition, the mandibular right first premolar was re-shaped to mimic a lateral incisor. There after, at 50 months the case was debonded and appropriate records were made. The patient did not agree to the extraction of the mandibular right third molar. Fixed retention was provided for the mandibular anterior teeth, in combination with upper and lower Hawley retainers to be worn full-time for one year and part-time for the following year. The patient was scheduled for biannual follow-up appointments. The patient and his parents were pleased with the results.

Treatment Results

Post-treatment photographs revealed excellent improvement in aesthetics, a well-interdigitated Class I molar occlusion and normal overjet and overbite. Wider arches (to ensure stability) and compression of the expanded maxillary cortices could be appreciated. The advancement of the mandible to a normal position is remarkable. The patient was checked for any dual bite problems before the case was debonded, hence advancement is factual (Figure 3). Post-treatment panoramic radiograph revealed root parallelism and mildly obliterated maxillary sinuses (Figure 4, A).

Figure 3. Post-Treatment Extraoral and Intraoral Photographs

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Post-treatment cephalogram, its tracing and analysis confirmed the clinical as well as photographic findings indicate that the mandibular plane angle, ANB angle, maxillary and mandibular incisor inclinations, and lip procumbence were within the normal values. Remarkable advancement of B-point (SNB; from 66.6° to 70.9°) and retrusion of A-point (SNA, from 73.8° to 71.1°) was seen. This was coupled with an increase in effective mandibular corpus length (Co-Gn, from 97.09 mm to 109.14 mm), an increase in ramal height (Ar-Go, from 40.71 mm to 50.19 mm), and an increase in mandibular base length (Go-Me, from 60.34 mm to 62.35 mm) (Figure 4, B and C; Table).

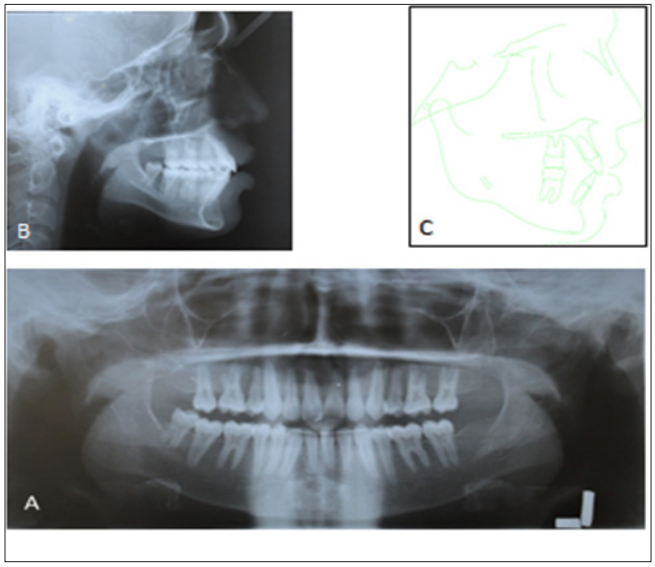

Figure 4.Post-Treatment Panoramic Radiograph, Cephalogram, and Tracing

Superimposition of pre-and post-treatment cephalometric tracings revealed remarkable growth of the posterior cranial base, as well as remodelling of the mandibular corpus. The usefulness of Class II elastic wear may be assessed by comparing the actual mandibular changes (Figure 5, B) with that of the predicted growth changes occurring during the entire duration of treatment (Figure 5, A). Post-treatment change in the mandibular form can be easily appreciated. It is associated with a considerable apposition of bone in the region of the angle of the mandible, mandibular base and also in the posterior border of the ramus. A peculiar characteristic is the concavity appearing midway of the mandibular base and also mild concavity below the neck of the condyle. The concave regions depict the areas where the mandible flexes when masticatory forces act upon it. The change in mandibular form suggests that greater forces are being borne by the mandible due to occlusal improvement, while concomitant advancement of the B-point and closure of the mandibular plane suggest an overall physiological improvement of the functioning spaces essential for the well-being of the patient.

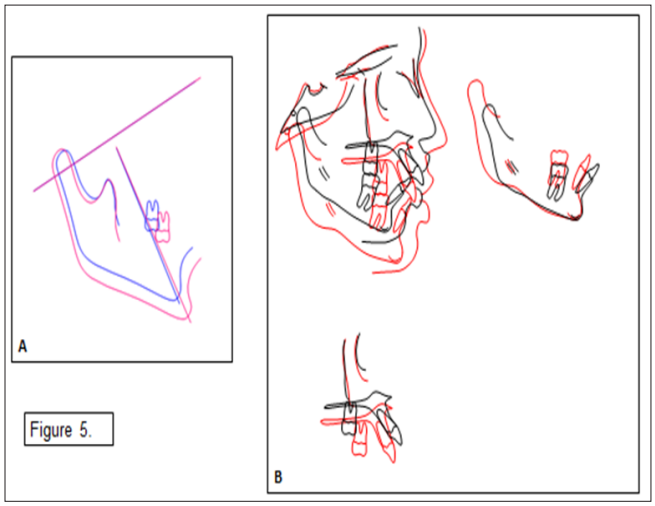

Figure 5. Growth Prediction and Superimposition of Pre-Treatment and Posttreatment, Cephalometric Tracings

Discussion

Currently no standard protocol for the orthodontic treatment of the various dental as well as orofacial complications in individuals afflicted with β thalassemia major exists in the literature. A Cochrane review is underway to assess the same.[14]

Until 1969, dentofacial deformities were primarily treated by a combination of orthopaedic appliances and fixed orthodontics.[9] However, with the advent of improved hypertranfusion techniques and iron chelation therapies, surgical treatment modalities have become the choice of treatment over the past five decades.[9,15] The main reasons to avoid orthodontic treatment have been the lack of awareness, willingness,[10] as well as relatively poor treatment outcomes, especially with respect to correction of the vertical and lateral bone deformity.[9,16]

Only 3 published case reports could be found where orthodontic treatment was instituted and no adjunct surgical procedures were performed. Kharsa[13] and Adelman[16] reported the use of cervical and high-pull headgear in a 7-and 10-year-old patient respectively to retract and intrude the anterior maxillary dentition. In both cases the maxillary protrusion was reported to decrease, however, vertical control could not be achieved, and hence facial aesthetics did not improve dramatically. The third case report authored by Einy et al[15] instituted in addition, a Twin block functional appliance to advance the mandible in a 10-year-old female patient, to correct the class II skeletal base. The functional appliance was employed for 7 months after a 5-month therapy with high pull headgear. The case was finished with conventional fixed appliances. The authors reported correction of the protrusive premaxilla, however, the skeletal base correction was limited (ANB from 15° to 11°). The cephalometric superimposition did

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not show any appreciable change in the ramal height or condylar growth. The mandibular auto rotation as well as advancement could have been hindered by the tight anterior dental contacts. Although the counter clockwise movement of the maxilla was attempted mandible did not follow suit, perhaps the intrusion of the entire maxillary arch could have affected the desired reduction of the mandibular plane. The use of temporary anchorage devices to bring about orthognathic-like changes seems promising in the treatment of thalassemia-associated alveolar overgrowth.[10] Another reason for scant changes in the condylar position might be attributed to the early fusion of the posterior cranial base, which precludes mandibular advancement.[1]

In contrast, the treatment outcomes in the present case are encouraging as far as modulation of mandibular growth and control of vertical dimension is concerned. The use of light class II elastics appears to bring about changes that are in sync with the natural physiological processes. In other words, the mandible was not actively protracted. This might be the reason why the treatment duration was much longer than the conventional orthodontic treatment, which aims to bring about only dental movements. It might be argued that the mandibular changes during treatment were a result of catch-up mandibular growth. This may not be the case since a considerable amount of growth occurred in the posterior cranial base as well. Furthermore, the mandibular changes were much greater than those predicted due to growth.

Fixed orthodontic treatment in combination with orthopaedic and functional appliances as well as temporary anchorage devices must be given due consideration while planning treatment of the dentofacial deformities in thalassemia patients.

Conclusions

Oro-facial deformities in a β thalassemia patient may be successfully treated by fixed orthodontic therapy in conjunction with light class II elastics to advance the mandible, in adolescent patients.

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Conflict of Interest: None

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