Orthodontic treatment in a patient with sickle cell anemia

Matheus Melo Pithon
Jequiê, Bahia, Brazil

Sickle cell anemia is a common hereditary hematologic disease. It affects many systems and tissues in the body, including the mouth. Delayed tooth eruption, an uncommon degree of periodontitis, alterations in the cells of the tongue surface, hypomaturatation and hypomineralization in enamel and dentin, pulp calcifications, hypercementosis, and bone alterations resulting in maxillary protrusion and formation of a thick trabecular pattern are some oral manifestations of the disease. The aim of this study was to report the orthodontic treatment of a patient with sickle cell anemia. Treatment consisted of correcting a Class II dental and skeletal pattern with an extraoral appliance combined with a fixed orthodontic appliance. From the orthodontic point of view, the results were satisfactory: the occlusion was normalized, and the patient’s health was maintained throughout the entire treatment period. (Am J Orthod Dentofacial Orthop 2011;140:713-9)

Sickle cell anemia is one of the most common hereditary hematologic diseases worldwide, and it is usually considered a matter of public health.1 It is a hereditary type of chronic hemolytic anemia, caused by a genetic mutation of the hemoglobin A molecule, which is called hemoglobin S.2,3

Hemoglobin is a respiratory protein in the erythrocytes with the primary function of transporting oxygen throughout the body. Molecular biology explains sickle cell anemia as a mutation of the hemoglobin that leads to cells that look like sickles.1,4 In situations of low oxygen tension, the affected hemoglobin molecules change from their normal disk shape to sickle shape. Initially, when the oxygen level increases, this sickling is reversible; however, the constant changes of shape of the red blood cells harm their cell membranes, which become rigid and no longer return to their normal state. This results in decreased capacity to transport oxygen to the tissues, vaso-occlusion conditions, and diminished lifespan of the red blood cells.

These physiopathologic events are the determinant in the origin of most signs and symptoms in the clinical condition of patients with sickle cell anemia: algic crises, high susceptibility to infections, hemolytic crises, ulcers in the lower limbs, splenic sequestration, priapism, strokes, and chronic compromise of multiple organs, systems, or processes.5-7

The clinical spectrum of involvement can vary greatly from patient to patient. The pathologic effects of sickle cell anemia, demonstrated in mineralized and connective tissues in other areas of the body, also occur in oral tissues.1,3 The most common findings described in the literature involving the oral region are paleness of the oral mucosa, delayed tooth eruption, uncommon degree of periodontitis, alterations in the cells of the tongue surface, hypomaturatation and hypomineralization in enamel and dentin, pulp calcifications, hypercementosis, and bone alterations, resulting in maxillary protrusion and formation of a thick trabecular pattern.1,3,5,8

There is no specific treatment for sickle cell anemia, and it is necessary to include preventive measures to ameliorate its consequences. These measures include good nutrition, prophylaxis, early diagnosis of and treatment for infections, maintenance of good hydration, and avoidance of adverse climate conditions. Allied with the preventive measures, folic acid is indispensable, because of its fundamental importance in the maturation and speed of red blood cell production, and should be prescribed for patients with sickle cell disease. In some cases, it is associated with vitamin B12.9,10

Orthodontists should have knowledge of the oral manifestations and possible interferences of sickle cell anemia.
anemia in orthodontic treatment. Nevertheless, no article describing orthodontic treatment in these patients was found, only cephalometric evaluations in population groups with this pathology and descriptions of the effects of this pathology on craniofacial growth. With the intention of filling this gap, the objective of this article was to describe the clinical orthopedic and orthodontic treatment of a patient with sickle cell anemia, its particularities, and the care to be taken.

DIAGNOSIS AND ETIOLOGY

The patient, a 12-year-old girl, came for orthodontic consultation with the chief complaint of misaligned and protruding teeth (Fig 1). Her mother said that she had sickle cell anemia and was under the care of a hematologist (Fig 2). According to the mother, the girl took supplements of folic acid associated with vitamin B12, prescribed by the hematologist. Clinically, she patient had a good state of general health, without any complications because of this pathology.

For adequate orthodontic treatment, it is indispensable to know the medical history of a patient with sickle cell disease, as well as his or her family history, to gain knowledge of the degree of systemic compromise of the disease.

The patient had an Angle Class II dental relationship, 12 mm of overjet, and an exaggerated anterior overbite (Fig 3). The gingivae showed discrete paleness, particularly in the posterior region. Enamel hypoplasia was noted on the maxillary incisors on the right side (Fig 1).

Cephalometrically, the patient had a Class II skeletal relationship (ANB, 6°) with maxillary protrusion (SNA, 85°) and a slightly retruded mandible (SNB, 79°), a trend toward a vertical facial growth pattern (SnGoGn, 34°), vestibularization and protrusion of the maxillary and mandibular incisors (1.NA, 30°; 1-NA, 8 mm; 1.NB, 38°; 1-NB, 7 mm), and a convex facial profile (LS-S, 4 mm; LI-S, 7 mm) (Figs 4 and 5).
TREATMENT OBJECTIVES

The goals were to correct the skeletal Class II relationship, improve the Class II dental relationship, harmonize the facial profile, improve the esthetics of the smile, and maintain the patient’s general health.

TREATMENT ALTERNATIVES

The treatment alternatives were to (1) extract the first premolars and retract the maxillary anterior teeth; (2) wait for cessation of craniofacial growth and plan for future orthodontic-surgical treatment with posterior repositioning of the maxilla and anterior repositioning of the mandible; and (3) correct the skeletal and dental Class II relationships with a combined extraoral traction appliance.

TREATMENT PROGRESS

The treatment plan chosen was the use of an extraoral appliance combined with a fixed orthodontic appliance. Initially, orthodontic bands were placed on the maxillary first molars, followed by fitting the extraoral traction appliance. After this, the fixed orthodontic brackets (0.022 × 0.030-in brackets; Morelli, Sorocaba, Brazil) were bonded to the remaining teeth. All banding appointments were preceded by antibiotic prophylaxis with 2 g of amoxicillin 1 hour before the procedure.

After the appliance was fitted, the dental alignment and leveling stage began, with a sequence of 0.014-, 0.016-, and 0.018-in stainless steel archwires. With the 0.018-in maxillary archwire, retraction of the maxillary second premolars began to create space for the distalized molars produced by the extraoral appliance. After this, the first premolars and the canines were moved distally.

Once the canines were well positioned in a Class I relationship, a 0.019 × 0.026-in maxillary retraction archwire was made with inverted drop-shaped loops, to achieve retraction of the maxillary anterior teeth.

When retraction was achieved, 0.019 × 0.026-in ideal arches were made to conclude the orthodontic treatment. Then the appliances were removed, with every care to control transitory bacteremia (antibiotic prophylaxis). After this, a mandibular canine-to-canine bonded lingual bar and a maxillary circumferential retainer were given to the patient.

TREATMENT RESULTS

The results obtained with orthopedic-orthodontic treatment were the correction of the dental and skeletal Class II malocclusion with reduction of maxillary protrusion and an improved relationship between the bony bases (ANB, 4°). The facial profile improved with the reduction in labial protrusion (LS-S, +3 mm; LI-S, +4 mm) and the passive lip seal at rest (Figs 6–10).

The maxillary incisors were better positioned in the bony base of the maxilla (1.NA, 15 mm; 1-NA, 3 mm). There was a discrete increase of the mandibular plane.
in a clockwise direction, which contributed to a slight increase in the lower third of the face (54%).

**DISCUSSION**

Sickle cell anemia is a common hereditary hematologic diseases worldwide, affecting a significant portion of the Brazilian population. It is usually pointed out as a question of public health. Thus, the objective of this article was to describe the orthodontic treatment of a patient with sickle cell anemia.

The oral manifestations in patients with sickle cell anemia are not pathognomonic of the disease but could help the dentist to identify it. The paleness of the oral mucosa is the result of chronic anemia or jaundice resulting from hemolysis of the red blood cells. In this patient, paleness of the mucosa was not noted, since she was under the constant care of a hematologist, thus preventing any anemic condition. Nor was any delay in tooth eruption seen. Nevertheless, there were defects in enamel mineralization affecting the crowns of the maxillary central and lateral incisors (Fig 1).

According to Franco et al., certain characteristics are found on the periapical radiographs of subjects with sickle cell anemia, including the formation of a thick trabeculated pattern, attributed to the erythroblastic hyperplasia and medullary hypertrophy that result in loss of the thin trabeculated bone and formation of wide medullary spaces. There is greater alteration in alveolar bone trabeculation, and the stepladder trabecular pattern is found more frequently in the posterior teeth. These characteristics of thick trabeculated bone were seen in this patient, particularly in the mandibular premolar region (Figs 4 and 8). The evident bone alterations of sickle cell anemia alert one to the importance of the radiographic examination as an aid in diagnosis of the disease.

Although the oral manifestations are not pathognomonic of the disease, the cephalometric alterations of the bone tissues are characteristic. The process of red blood cell sickling results in a lower capacity of oxygen transport to the tissues, circulatory difficulties, and a reduction in their lifespan, which falls from 120 days to approximately 20 days, when they are removed from circulation by the spleen. The early destruction of red blood cells makes it necessary for them to be constantly produced, causing hyperplasia and compensatory bone marrow expansion, which might result in changes in the bony structures that can be observed radiographically. 

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**Fig 4.** Pretreatment radiographs.

**Fig 5.** Pretreatment cephalometric tracing.
The gnathopathy resulting from the exaggerated growth of the midface was first described by Brown and Sebes who observed expansion of the maxilla and protrusion of the middle third of the face in patients with sickle cell anemia. The author attributed this fact to medullary hyperplasia and found maxillary protrusion in 31.6% of anemic patients in Ghana. These skeletal alterations were seen in the patient of this report. Cephalometrically, she had a Class II skeletal relationship with maxillary protrusion (SNA, 85°) and slight mandibular retrusion (SNB, 79°) (Fig 5). These findings agree with the cephalometric measurements for Brazilians with sickle cell anemia (SNA, 85°; SNB, 78°) found by Souza et al in 2008.

The issues in treating these patients are concentrated on avoiding bleeding during orthodontic procedures. If it is not possible to prevent bleeding, the procedures should be performed under prophylactic antibiotic therapy. Habitual orthodontic procedures that might generate some type of bacteremia would be the placement and removal of orthodontic separators and orthodontic mini-implants. Therefore, every orthodontic separation procedure was preceded by antibiotic prophylaxis with 2 g of amoxicillin, as suggested by Franco et al. The use of mini-implants was not necessary because an extraoral appliance was used to correct the skeletal Class II relationship.

The choice of treatment was strictly related to the pathology; ie, any more aggressive procedure was avoided. The option was to correct the Class II malocclusion with an extraoral appliance instead of extractions, mini-implants, or orthognathic surgery.

Our choice of treatment without tooth extractions was necessary because extraction of asymptomatic teeth is contraindicated in patients with sickle cell anemia. In these patients, teeth should be extracted only when it is impossible to perform adequate restorative procedures.

Special care was taken throughout the treatment period, which included periodic consultations with the
hematologist, strict control of oral hygiene, and antibiotic prophylaxis during all procedures that could lead to a bacteremic condition. Other concerns would be to avoid treatment during the chronic stage of the disease and avoid appointments during algic crises, except in cases of emergency.

The major cause of death in patients with sickle cell anemia is infection. Pneumonia, renal infection, and osteomyelitis occur with greater frequency in children and adults with sickle cell anemia, caused by the functional splenectomy resulting from continuous fibrosis of this organ over the years. Episodes of fever must be regarded as situations of risk, requiring immediate therapy. Oral surgeries are invasive procedures involving high risk; therefore, they must be judiciously planned so that the intervention will be safe. Susceptibility to infection justifies the use of prophylactic antibiotic therapy before invasive procedures that could cause bleeding and promote bacteremia. When surgery is necessary, the dental surgeon must request a complete hemogram of the patient.

CONCLUSIONS

Performing orthodontic treatment on a patient with sickle cell anemia is a reality, requiring special care from both the local and the systemic points of view. Because we took special care, our results were satisfactory, with correction of the malocclusion and maintenance the patient’s general health.

REFERENCES