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VIRTUAL EVENT

Challenges in the surgical management of sickle cell gnathopathy: a case report and literature review.

Thanh-Thuy Nham, Romain Deransy, Marielle Surbled-Leproux, Pierre Corre, Justine Loin, Hélios Bertin.

Background

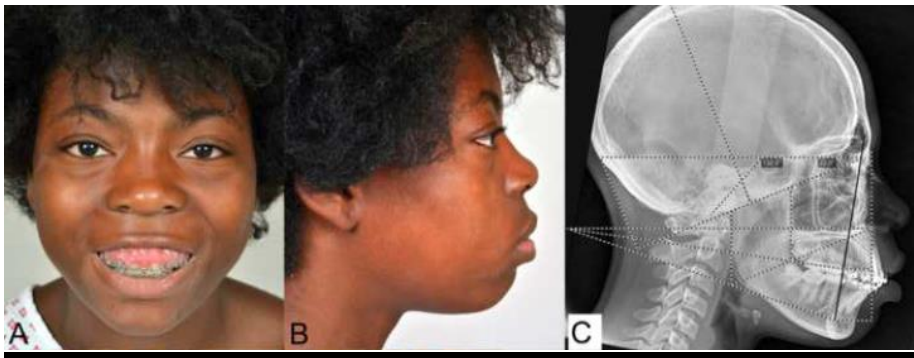
Sickle cell disease (SCD) is one of the most common inherited haemoglobin disorders[1]. The craniomaxillofacial dysmorphosis and malocclusion induced by SCD have been described[2,3,4]. However, the maxillary protrusion also called sickle cell gnathopathy remains poorly described in the literature and it is probably underestimated by physicians[7]. Maxillary protrusion is thought to be triggered by the chronic hemolytic anemia, this can lead to hypoxemia and cellular hypoxia thereby causing compensatory erythropoiesis and bone marrow hyperplasia[2]. The maxilla undergoes an exaggerated reactional development and can have a major functional and aesthetic consequence in patients, hence carranting orthodontic and surgical correction[7]. We report the case of a patient who underwent surgical correction of her sickle cell gnathopathy, and we discuss the preventive measures to safely undertake the procedure.

Case report

A 15-year-old black female with severe homozygous SCD consulted for possible correction of her sickle cell gnathopathy. She was adversely affected by a gummy smile maxillary protrusion and retrogenia with class II dental occlusion.

Surgical procedure

A one-stage surgical procedure with an ascending Lefort I osteotomy, an advancement genioplasty and gingivoplasty was performed.



BEFORE SURGERY *A: Front B: Profile C: Cephalometric analysis on x-ray*

AFTER SURGERY: *Front E: Profile F: Cephalometric analysis on x-ray*

The perioperative measures[5,6]

Pre-operative

1. Personalized treatment plan, including the surgeon, the haematologist, and the anesthesiologist
2. Blood test: CBC, platelets, reticulocytes, blood group, and complete phenotyping, irregular agglutinin test, PT, APTT, ionogram, serum creatinine, liver function panel test, LDH
3. Monitoring of oxygen saturation (pulse oximetry)
4. Depending on the background: echocardiography, pulmonary function test, transcranial Doppler ultrasound, or magnetic resonance angiography
5. If prosthetic material is required: search for a dental infectious or ENT outbreak, digestive focus, and urinalysis
6. Evaluation of the need for transfusion therapy: to be performed at least 8 days before the procedure
7. Hydration per os 48 hours before, then IV on fasting: 2-3 L/24 h
8. Respiratory physiotherapy by incentive spirometry: three per day for 10 minutes, 1 week before any intervention
9. Nasal oxygen therapy (2 L/min)

Per-operative

1. Monitoring of oxygen saturation and blood pressure
2. Compensation for blood loss by isotonic solutes for normovolemia
3. Usual pre-oxygenation
4. Normothermia
5. Avoid controlled hypotension, with mean arterial pressure ≥ 70 mm Hg
6. Avoid rinsing with iced serum
7. Antibiotic prophylaxis (curative antibiotic to be discussed with the surgeon)
8. Control of bleeding with transfusion therapy

Post-operative

1. Immediate observation in the intensive care unit
2. Maintaining intravenous hydration until normal oral nutrition
3. Pulse oximetry monitoring with the objective of an oxygen saturation level > 96%
4. Mobilization
5. Respiratory therapy incentive spirometry to continue in the postoperative period
6. Monitoring of body temperature
7. Avoid facial icing

Discussion

Sickle cell gnathopathy is an unusual maxillary protrusion considered as the result of a bone marrow hyperplasia induced by chronic haemolytic anemia. It concern between 20% and 30% patients[2]. There appears to be a correlation between the severity of SCD and craniofacial impairment. The orthognathic surgery must be proposed tactfully, measured in these patients at risk of severe and acute decompensation[7]. It involves the adaptation of the surgical and anaesthetic parameters.

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