

Case Report

Management of Sleep Apnoea (due to nose block) in a Case of Thalassemia Major

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Abstract

This paper presents a case of Thalassemia Major (Beta Type), a major bleeding disorder where major surgery is usually avoided. The patient had almost a total nose block which was corrected satisfactorily by widening the nasal air passages by a simple noninvasive maneuver [with a long bladed nasal speculum]. The nasal turbinates were fractured and pushed laterally; the patency was maintained by inserting and keeping nasal airway tubes for five weeks. This is an innovative trial with minimal bleeding.

Keywords: Thalassemia major (Beta); CT scan report; Rat face [hair on head] deformity; Nasal airway passage; Nasal airway tubes

Case Presentation

An African 12 years old boy, a known case of Thalassemia Major (beta type), born normal and was asymptomatic but developed short of breath, snoring and headache from the age of 3 years onwards. He was diagnosed by the blood test and his CT scan of skull showed involvement of both maxillary and frontal sinuses with expansions of its outer tables producing a rat face deformity and near total bilateral nasal blockage, producing sleep apnoea, short of breath on physical exertion and day time drowsiness. He was being stabilized with blood transfusions under the supervision of the pediatricians. Pathophysiology: Thalassemia is an inherited blood disorder of an imbalanced globin chain synthesis where haemoglobin protein molecule in red blood cells is abnormal in form leading to less oxygen carrying capacity and destruction of red blood cells, producing severe anaemia requiring repeated blood transfusions. There are two main forms of beta thalassemia, thalassemia minor [less severe form] in which one of beta globin gene has mutation or abnormality and in Thalassemia major (severe form) two of the beta globin genes are affected. A Low oxygen carrying capacity status stimulates an increased haemopoiesis in bone marrows, the increased amount of marrow tissues leads to expansion of thin cortical bones, more in skull bones, causing expansion of sinuses predominantly maxillary and frontal. This medial expansion of maxillary sinuses, lead to nasal obstruction (Figure 1) and Frontal sinus expansion with a protuberance produces a face deformity (Figure 2), called "hair on head" (rat face).

Maneuver

The inferior and medial turbinate's of each nostril were pushed

and fractured outwards (Laterally) by a long bladed nasal speculum (KELIAN'S). The resulted widened bilateral nasal passages were maintained by two airway tubes, 4 inches long pieces of endotracheal tubes (size 5) for 5 weeks. The nasal airway tubes were kept one inch outside the nostrils (Figure 3) and mild antibiotic soaked nasal pack was put around the nasal airway tubes. The patient tolerated the procedure well and there was hardly any bleeding. Normal saline irrigation through the airway tube was advised to avoid its blockage and to remove the crusts and secretions, twice in a day for a week. Post operatively antibiotics and steroids were given. Once the airway tubes were removed, the patient had a satisfactory nasal breathing (Figure 4-6), found patent even after one year of the maneuver. This is a simple and easy procedure and can be repeated if required.

Declaration

Consent was taken from relative of the patient and from the institution for its publication and there is no financial constrain for it.

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Figures 1 and 2: The excessive destruction of red cells leads to Hepato-splenomegaly and repeated blood transfusions result into an excessive haemoglobin in blood (reducible by a Chelation therapy).



Figure 3: Post operatively antibiotics and steroids were given.



Figures 4-6: The patient had a satisfactory nasal breathing.