Case Report

ANAESTHETIC MANAGEMENT OF A FIVE AND HALF YEARS OLD CHILD WITH BETA THALASSAEMIA MAJOR(COOLEY'S ANAEMIA) UNDERGOING TOTAL MOUTH REHABILITATION : A CASE REPORT

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ABSTRACT

Thalassaemias are a group of haematological disorders characterized by deficient or total lack of normal haemoglobin chains with extra vascular haemolysis and ectopic marrow expansion leading to anaemia, splenomegaly and bony abnormalities. Survival is associated with various multisystem complications primarily caused by chronic anaemia, iron overload, adverse effects of chelation, and transfusion associated infections We report a five and half years old child with beta thalassaemia major diagnosed at the age of one and half years receiving blood transfusions every three weeks on chelation therapy for iron overload. This child presented to the dental outpatient department with extensive dental caries and was scheduled for a total mouth rehabilitation under general anaesthesia. Due to early diagnosis, regular blood transfusion and chelation therapy for iron overload this child did not have obvious oro facial deformities and the typical facial appearance (chipmunk face)

KEYWORDS : Beta thalassaemia major, facial deformities, transfusion, iron overload, difficult airway

INTRODUCTION

The thalassaemias are a group of autosomally recessive inherited conditions characterised by decreased or absence of synthesis of one of the two polypeptide chains (**α** or **β**) that form the normal adult human haemoglobin molecule (haemoglobin A $\alpha_2\beta_2$), which results in reduced haemoglobin in red cells, and anaemia. **β** globin gene defects may give rise to **β** thalassaemia, while mutations of the **α** globin gene may cause **α** thalassaemia. Thalassaemia is prevalent in Mediterranean, Transcaucasus, Middle East, African and Asian regions. Evolutionary biologist J.B.S.Haldane postulated that the thalassaemias were common in human populations because they protected against malaria. Alpha thalassaemia is common in Asia, the Mediterranean and Melanesia where malaria is or was prevalent.

Evidence has accumulated that the first line of defence against malaria is provided by genetically controlled innate resistance, mainly exerted by abnormal haemoglobins and glucose-6-phosphate dehydrogenase deficiency. The three major types of inherited genetic resistance - sickle cell disease, thalassaemias, and G6PD deficiency were present in the Mediterranean world by the time of the Roman Empire. There are many forms over 300 mutations giving rise to thalassaemia. The most serious forms are called beta thalassaemia major (Cooley's anaemia) and alpha thalassaemia major. Although it is possible to live with treatment for beta thalassaemia major, alpha thalassaemia major is not compatible with life. The genes that encode the alpha globin chains are on chromosome 16. Those that encode the non-alpha globin chains are on chromosome 11 The most common treatment for beta thalassaemia major is to have regular blood transfusions every four-to-six weeks to top-up haemoglobin in the body. This treatment can raise iron levels in the body too high, risking heart, liver and hormone problems. Chelation may be given to reduce iron levels.In some cases, bone marrow transplants may cure thalassaemia, especially in under-16s, and when the marrow is donated from a brother or sister. Stem cells in umbilical cord blood transfusion are a newer technique used to cure for thalassaemia in some instances. Genetic counselling and genetic testing are recommended for families who carry a thalassaemia trait.

The newborn infant with **β** thalassaemia major

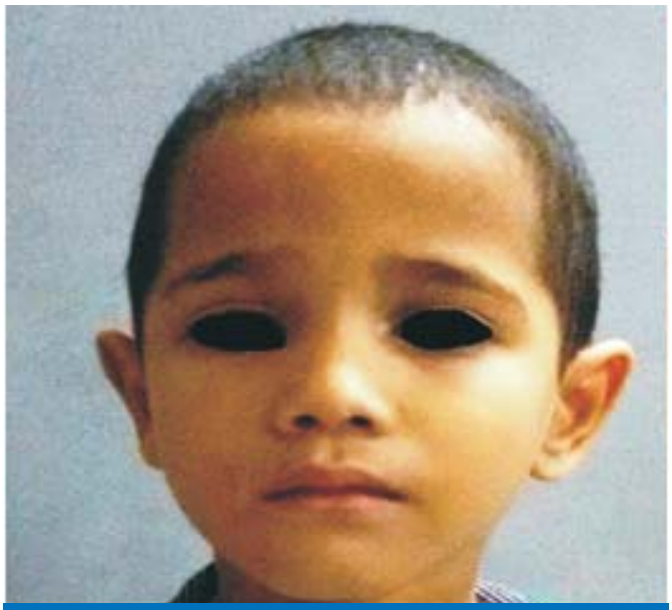
patients is born healthy and not anaemic; however symptoms such as anaemia, growth retardation, jaundice and bone changes usually develop within the first year of life, thus making regular blood transfusion and iron chelation therapy necessary for survival. Severe anaemia results in cardiac enlargement and splenomegaly develops within three years. Moderate hepatomegaly may be present with mild jaundice The clinical course of **β** thalassaemia major in most cases is severe. It is a life threatening condition characterized by severe, hypochromic and microcytic anaemia. Endocrine abnormalities encountered in **β** thalassaemia major are evident during the second decade of life and are secondary to the chronic iron overloading. Due to expansion of marrow in malar bones, skull, long bones and feet the characteristic skeletal changes are seen. Cortical thinning causes pathological fractures and bone pain. In addition skeletal changes are seen due to hypertrophy and expansion of erythroid marrow and susceptibility to infections is increased. Extramedullary haemotopoiesis may occur with masses of haemopoietic tissue compressing the spinal cord. Other clinical features include leg ulcers, epistaxis, skin pigmentation and gall stones. Hypersplenism worsens anaemia and may also reduce platelet count. The increase in body iron resulting in pancreatic haemosiderosis which may cause diabetes and cirrhosis from iron deposition in the liver. Haemosiderosis in the cardiac muscle causes arrhythmias, heart blocks and congestive cardiac failure. Radiological changes are not apparent until one year of age. These include large bone marrow spaces, one of the most important radiographic features of thalassaemia. This enlargement is explained by the fact that, when ineffective erythropoiesis damages the RBC membrane leading to severe anemia, the body responds by increasing the production of red blood cells, consequently causing expansion of the bone marrow up to 15-30 times the normal amount. The skull radiograph shows the increased diploid space and arrangement of trabeculae in the vertical rows, causing "hair on end" appearance.

CASE REPORT

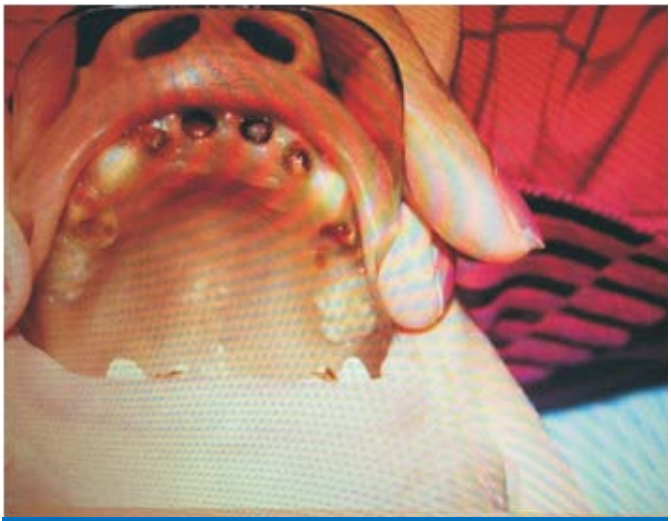
We report a five and half years old male child with β thalassaemia major who presented to the department of oral medicine with extensive dental caries. The child had a height of 110 cm and weighed 19 kg. He was normal at birth but after one year of life he manifested with anaemia, growth retardation and failure to thrive and was detected to have β thalassaemia major. He received blood transfusions from the age of one and half years receiving a unit every three weeks and was on chelation therapy since then. He did not have the classical maxillary hypertrophy except for frontal bossing, migration of spacing of upper anterior teeth, discoloured teeth with short crowns and roots, pale gums and mucosa. He had a depressed nasal bridge, a saddle nose , telecanthus, low set ears and a high arched palate. Preoperatively airway was assessed as normal. Patient had a haemoglobin value of 8.7 gm%, haematocrit 25 %, with HbA2 1.5 % and HbF 98.5%. ECG ,X-ray chest ,liver function tests ,renal function tests, serum electrolytes and blood sugar were within normal limits. Serum ferritin levels were 1979.3 ng/ml(grossly elevated),serum transferrin saturation was 80% and osmotic fragility of RBCs was increased. Peripheral smear showed microcytic hypochromic anaemia with anisocytosis and poikilocytosis.

The surgery was scheduled after obtaining a written informed consent from his parents. On the day of surgery he was premedicated with intravenous midazolam 0.5 mg and fentanyl 30μg. After preoxygenating with 100% oxygen for 5 minutes anaesthesia was induced with intravenous 30 mg propofol and 10 mg atracurium. Nasotracheal intubation was done with a cuffed 4.5 mm ID (internal diameter) RAE tube. After checking air entry, cuff was inflated, endotracheal tube was fixed and throat was packed with a sterile wet tape gauze. Direct laryngoscopy was Cormack and Lehane grade I. Anaesthesia was maintained with O2:N2O 1:1, sevoflurane 2% and bolus doses of atracurium. Intra operative monitoring included ECG, non invasive blood pressure, pulse oximetry and capnography (ETCO2). Baseline heart rate was 110 beats/minute and it was maintained between 110 -120 beats/minute. Blood pressures in the beginning were 100/60 mm Hg and remained in the range of 100-110 mmHg systolic blood pressure and 60-70 mm

Hg diastolic blood pressure. Blood loss was minimal. Surgery lasted for 120 minutes. Two hundred and fifty milliliters of Ringer lactate solution was administered. At the end of surgery throat pack was removed, neuromuscular blockade was reversed with neostigmine and glycopyrrolate and patient was extubated awake after thorough oropharyngeal suctioning. Paracetamol suppository was inserted for post operative pain relief.The post operative period was uneventful and oral fluids were started after four hours.



Frontal bossing, telecanthus & low set ears



Marked dental decay. high arched palate and root stumps in upper anterior.

DISCUSSION

Thalassaemia is one of the common genetic disorders, a hereditary blood disease transmissible through an autosomal recessive gene. Thalassaemia patients present with marked maxillofacial deformities. Thalassaemia major patients present with problems at a very early age of 1- 2 years. Children who are adequately transfused and with haemoglobin levels maintained at 9–10 gm % show less maxillofacial manifestations. Regular blood transfusions and iron chelation have dramatically improved the quality of life. The anaesthesiologist should be aware of airway problems, anaemia, hepatosplenomegly and cardiomypathy and organ involvement due to iron deposition. Splenomegaly produces hypersplenism which worsens the anaemia. This necessitates splenectomy in such patients which itself is fraught with problems like sepsis, recurrent infection and immunosuppression. This disease poses financial and social hardships to the patient and passes from one generation to another. Hence genetic counselling and screening is a must to prevent spread of this disease.

CONCLUSION

Late diagnosis and patients receiving inadequate blood transfusion in childhood will present with more bone changes, face and skull abnormalities and anaemia. Hence, early diagnosis, regular and repeated blood transfusion along with iron chelation lessens the severity or even prevents maxillo facial deformities. Bone marrow transplantation is the only treatment available to cure thalassemia. This therapy should be considered in all patients who have a HLA-identical donor.

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