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CASE STUDY

ANAESTHETIC MANAGEMENT OF β – THALASSEMIA MAJOR WITH MASSIVE SPLENOMEGALY FOR SPLENECTOMY

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ABSTRACT

Six years old male child known case of β – Thalessemia Major from 6 months of age, who is on repeated blood transfusions admitted with failure to thrive, breathlessness and reduced activity. Complete investigation was done for this child who showed microcytic hypochromic anemia, thrombocytopenia, prolonged bleeding and clotting time, abnormal liver function test and Tricuspid regurgitation grade III with moderate pulmonary hypertension and cardiomegaly. With the above high risk complications the child was posted for elective splenectomy and managed successfully.

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INTRODUCTION

β Thalassemia is more common in individuals from the Mediterranean, south east Asia, India and part of Africa. It's an autosomal recessive trait, which causes severe microcytic, hypochromicanaemia, splenomegaly, and severe bone deformities and leading to premature destruction of maturing erythroblasts in the marrow (ineffective erythropoiesis) and lysis of mature red cells in spleen (hemolysis). Management consists of periodic blood transfusion; splenectomy if splenomegaly is present, and treatment of transfusion-caused iron overload (Nishkarsh Gupta, 2011 and Staikou, 2014). Splenectomy should be reserved for cases ofworsening anaemia leading to poor growth and development. Leading to worsening anaemia, leucopenia, or thrombocytopenia and causing clinical problems such as recurrent bacterial infections or bleeding. Hypersplenism accompanied by symptoms such as left upper quadrant pain or early satiety. Massive splenomegaly (largest dimension >20 cm) with concern about possible splenic rupture. Post-splenectomy sepsis remains a risk in all splenectomized thalassemia patients. Therefore, febrile splenectomized patients should undergo rapid evaluation and treatment. Splenectomized patients should receive prophylactic antibiotic therapy for at least two years following splenectomy. The importance of compliance with prophylactic antibiotic therapy should be stressed repeatedly to patients and parents while explaining that it does not entirely prevent post splenectomy sepsis and immediate presentation in cases of febrile illness is essential.

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Case Report

Six year old male child weight about 12 kg, known case of β – Thalassemia major with massive splenomegaly for elective splenectomy. Child was diagnosed during his 6th month of life as β thalassemia major from which he had repeated blood transfusions and frequency of transfusion increased in recently. Child has loss of appetite, breathlessness and reduced activity. Preoperatively managed with blood transfusion, chelating agents, pneumococcal, meningococcal, and influenza vaccinationalso was done. On examination patient conscious, oriented, pallor, thalassemicfacies (Maxillary hyperplasia, Flat nasal bone, Frontal bossing) (Figure 1) pulse rate-104/min, blood pressure- 90/60 mm of Hg, CVS – S1S2 heard, Pansystolic Murmur present, abdomen distended with massive splenomegaly extended up to right iliac fossa and hepatomegaly (Figure1). Investigation showed Haemoglobin – 6 grams initially which increased to 10.4 grams after transfusing two units of blood, preoperatively bleeding time prolonged more than 15 minutes, clotting time prolonged more than 17 minutes, thrombocytopenia, prothrombin time 15 seconds, liver function test shows elevated serum bilirubin and liver enzymesand renal function tests within normal limits. Peripheral smear shows microcytic hypochromic anemia, bone marrow shows no immature cells. Viral markers negative. Electrocardiogram shows sinus tachycardia, chest X ray shows

cardiomegaly and Echocardiography shows right atrial and right ventricle dilatation, Tricuspid regurgitation grade III , moderate pulmonary hypertension with minimal pericardial effusion. Case was assessed under ASA III. High risk consent was obtained from parents and we are planned for general anesthesia.

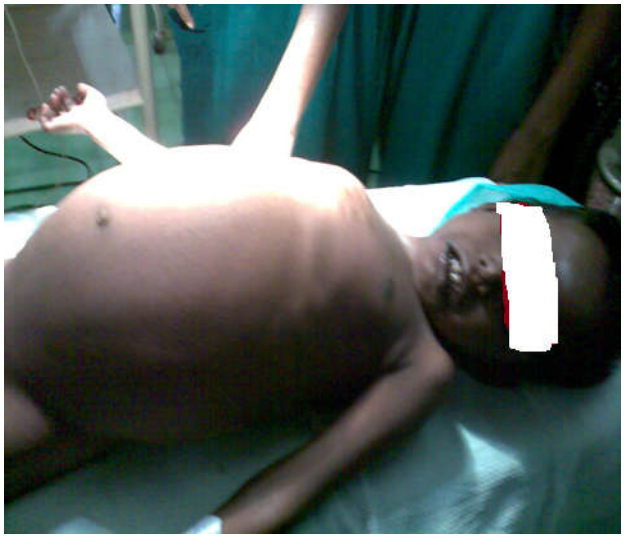

Figure 1. Distended abdomen

Inside the operative room intravenous line was secured, monitors were connected (pulse oximetry, NIBP, ECG, EtCO2, temperature). Aspiration prophylaxis done with ranitidine 25mg and ondansetron 2mg given. Child was premedicated with atropine 0.2 mgand midazolam0.5 mg. Preoxygenation with 100% O2 for 3 minutesdone. Induced with propofol 30 mg, fentanyl 25µg, suxamethonium 25 mg. Oral intubated with 4 mm ID uncuffed endotracheal tube and bilateral air entry was checked and connected to Modified Jackson-Rees Circuit. Maintenances with N2O: O2- 50:50 & 1% sevoflurane and titrated dose of atracurium and fentanyl. Intra-operatively during an insertion ofnasogastric bleeding occurringfrom nose and tooth loosed (Figure 2) bleedingcontrolled with nasal compression and blood products. Surgeryblood loss around170ml and replaced with150 ml of wholeblood and 1 unit of FFP was transfused, 150ml of crystalloids was infused. Splenectomy done and hemostasis were achieved (Figure 3, 4).The surgery lasted for 90minutes. Paracetamol suppository 160 mg was given for postoperative analgesia.

Figure 2. Bleeding and nasogastric tube

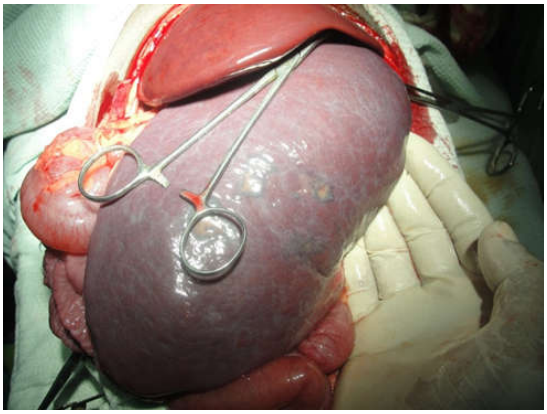

Figure 3. Hepatosplenomegaly

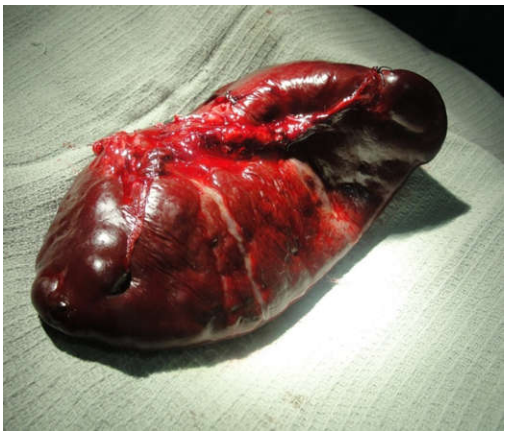

Figure 4. Splenectomy specimen

Intraoperatively hemodynamic was stable and throughout the surgery(HR -90 to 150 /min, BP - 80 to 100/ 50 to 60 mm of Hg, EtCO2 – 34 to 40 mm of Hg, Spo2 – 99 to 100%). Neuromuscularblockade reversed with neostigmine 0.6 mg andatropine 0.2 mg and then extubatedin awake state. (Figure 5) Post-operative periods were alsouneventful and child was discharged on 10thpostoperative day.

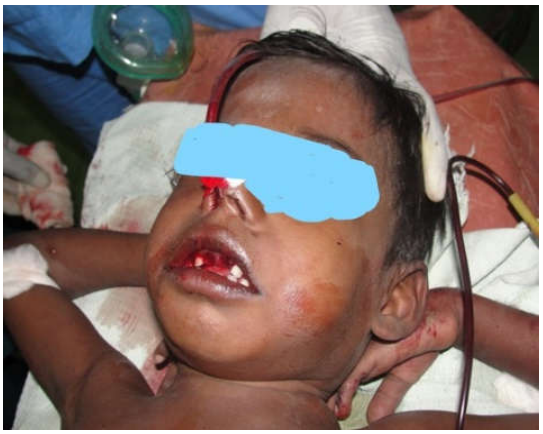

Figure 5. After extubation

DISCUSSION

Thalassemia major were decreased synthesis of one globin chain leads to fewer amounts of hemoglobin, hypochromicand microcytosis and the relative excess of other unpaired chain precipitates as insoluble inclusion leading to premature

destruction of maturing erythroblasts in the marrow (ineffective erythropoiesis) and lysis of mature red cells in spleen (hemolysis). Anemia results in increased erythropoietin production and extra medullary erythropoiesis and splenomegaly (Roberta, ?). In the most severe form, presentations with pallor, icterus, and enlarged abdomen are common. This patient presented with severe microcytic hypochromic anaemia, massive splenomegaly, thrombocytopenia and abnormal liver function test (Ali, 2010). Peripheral smear shows hypochromic microcytosis, anisocytosis, poikilocytosis, target cells, basophilic stippling and fragmented red cell. Reticulocyte count is elevated, bone marrow expansion and extramedullary erythropoiesis occurs and characteristically laeds to skull and facial deformities (Thalassemic facies). Haemolytic anemia causes hepatosplenomegaly, leg ulcer, gall stone, high output cardiac failure. Untreated children suffer from profound growth retardation, susceptible to infection, endocrine dysfunction and die at early age.[4] Blood transfusions are extended for survival. They improve anemia and suppress secondary features related to excess erythropoiesis. Iron overload and secondary haemochromatosis inevitably becomes a problem in healthy transfused patients (Roberta, ?). In this case child had failure to thrive, breathlessness and reduced activity. Patient was diagnosed as β Thalassemia major from 6 months of life, was on repeated blood transfusion and chelating agent (deferoxamine) for the past 5 ½ years.

Anaesthetic management

Pre-operative preparation depends on severity of anemia, skeletal deformity and secondary organ damage. Hematologicalinvestigations,cardiac, hepatic and endocrine function to be done and optimized (Staikou, 2015). Sevoflurane is favoured in paediatric practice for gaseous induction, but desflurane or isoflurane are marginally the preferred agents for maintenance of anaesthesia in children with liver disease undergoing major abdominal surgery.so we are avoided the halothane and use the sevoflurane for induction and maintenance agent for this child (Staikou, 2014; Kallenbacha, 2015 and Green, 2002). This child was presented with pulmonary hyper-tension and distended abdomen. Thus it is prudent intraoperatively to avoid conditions that will worsen pulmonary hypertension such as acidosis, hypoxia and hypercarbia (Green, 2002 and Cappellini, 2012). Pre-splenectomy antibiotics and immunization (pneumococcaland H. Influenza) to be given. Epidural and spinal anesthesia are relative contraindicated due to increased bleeding and hematoma formation. Possible chance of aspiration pneumonitis due to distended abdomen (Staikou, 2015).

Intra-operatively chance of hypoxia is more common in β thalassemia patient because of microcytic hypochromic anemia so pre-oxygenation is must. Over-distended abdomen can cause aspiration pneumonitis so premedication with aspiration prophylaxis is essential. craniofacial abnormality (Hyperplasia of facial bone, narrowing of nasal passage) may lead to difficult mask ventilation and intubation.[8] Careful positioning done becauseof demineralized(osteoporosis) extremitiesto prevent pathological fractures.[9]This patient have more chance to hemorrhagic complication due to prolonged bleeding time, clotting time so careful monitoring essential. Cardiovascular monitoring to prevent post-splenectomy hypertension. [4]Post-operatively monitor oxygen saturation and supplement oxygen. Watch for bleeding complication, monitor cardiovascular functioning.

Conclusion

β Thalassemia major includes various anaesthetic problems intra-operatively like difficult intubation, bleeding complication, sudden haemodynamic changes, hypothermia which had been managed successfully. Hence being aware of all the problem will lead to successful outcome.

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