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

Anaesthesiology

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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.

KEYWORDS

β – Thalassemia Major, cardiomegaly, splenectomy.

INTRODUCTION

β Thalassemia is more common in individual from the Mediterranean, south east Asia, India and part of Africa. It's an autosomal recessive trait, which causes severe microcytic, hypochromic anaemia, 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. [1] [4]

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 vaccination also was done.

On examination patient conscious, oriented, pallor +, thalassemic facies (Maxillary hyperplasia, Flat nasal bone,Frontal bossing) pulse rate-104/min, blood pressure- 90/60 mm of Hg, CVS : S1S2+, Pan systolic Murmur +, abdomen distended with massive splenomegaly extended up to right iliac fossa and hepatomegaly .

Investigation showed Haemoglobin – 6 gms initially which increased to 10.4 gms 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 secs, liver function test shows elevated serum bilirubin and liver enzymes and normal renal function tests. 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 atrium and right ventricle dilatation, Tricuspid regurgitation grade III , moderate pulmonary hypertension with minimal pericardial effusion. Case was assessed under ASAPS III. High risk consent was obtained from parents and we are planned for general anesthesia.

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 mg and midazolam 0.5 mg.Preoxygenation with 100% O2 for 3 minutes done. Induction done 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 of nasogastric tube bleeding occurring from nose and tooth loosed (Figure 1) bleeding controlled with nasal compression and blood products. Surgery blood loss around 170 ml and replaced with 150 ml of whole blood and 1 unit of FFP was transfused, 150ml of crystalloids was infused. Splenectomy done and hemostasis were achieved (Figure 2,3).The surgery lasted for 90minutes. Paracetamol suppository 160 mg was given for postoperative analgesia.

Figure 1. Bleeding and nasogastric tube

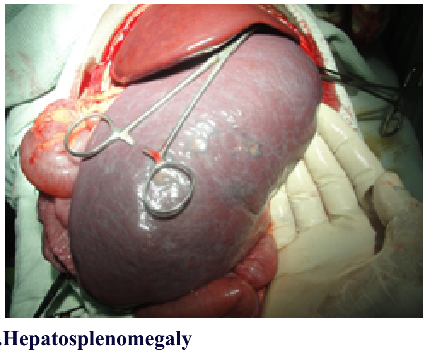

Figure 2 .Hepatosplenomegaly

Figure 3. Splenectomy specimen

Intra-operatively 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%).Neuromuscular blockade reversed with neostigmine 0.6 mg and atropine 0.2 mg and then extubated in awake state. (Figure 4) Post-operative periods were also uneventful and child was discharged on 10th postoperative day.

Figure 4 .After extubation

DISCUSSION

Thalassemia major were decreased synthesis of one globin chain leads to fewer amounts of hemoglobin, hypochromic and 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[2]. 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.[3]

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.[2]

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. Hematological investigations, cardiac, hepatic and endocrine function to be done and optimized.[4] 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. [4][5][6]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.[6][7]

Pre-splenectomy antibiotics and immunization (pneumococcal and H.Influenza) to be given. Epidural & spinal anesthesia are relative contraindicated due to increased bleeding and hematoma formation. Possible chance of aspiration pneumonitis due to distended abdomen.[4]

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 because of demineralized (osteoporosis) extremities to 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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