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ANAESTHETIC MANAGEMENT OF β-THALASSEMIA MAJOR POSTED FOR SPLENECTOMY IN PAEDIATRIC AGE GROUP- A CASE REPORT

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ABSTRACT B –thalassemia is the most common cause of Haemolytic Anaemia in India.Hereby,we are reporting a case aged 13 years,who presented with massive splenomegaly and hypersplenism posted for splenectomy over a period of 4 months.The child was on repeated blood transfusions since the day of diagnosis.The purpose is to emphasize on the anaesthetic concerns,prevent the complications, raise awareness of the disease and its prevention by aggressive screening and prenatal diagnosis.

KEYWORDS :

INTRODUCTION:

Beta thalassemia [1,2] is an autosomal haematological disorder that occurs as a result of genetically deficient synthesis of β-globin chains of haemoglobin .Patients may present with severe microcytic hypochromic anaemia,massive hepatosplenomegaly and bony deformities[3].Anaesthetic management of these children is challenging and can be associated with unanticipated difficult airway,high perioperative blood pressure, iron overload, endocrinological abnormalities,cardiac disease, restrictive respiratory pattern and pulmonary hypertension.We present this case which highlights anaesthetic concerns in children of beta thalassemia major with hypersplenism posted for splenectomy.

Case Report

A 13 yr old child weighing 26 kgs born of a non consanguineous marriage and a known case of β-thalassemia major with massive splenomegaly presented to us for splenectomy.He was diagnosed with it at 4 months of age and his requirement for blood transfusion increased gradually and underwent 234 blood transfusion since birth.On perioperative examination child had frontal bossing, high arched palate and enlarged abdomen.

The airway assessment was found to be normal with Mallampati grade 1 and mouth opening was adequate. He was pale, icteric, afebrile with heart rate of 82 beats per minute,blood pressure of 90/60 mmHg,respiratory rate of 28 breaths/min and oxygen saturation of 98% on room air.Systemic examination revealed clear lung fields,massive splenomegaly and hepatomegaly. Perioperative investigation revealed Haemoglobin of 7.4 gm/dl, WBC count of 2,900/cumm and platelet count of 1.5 lakh/cumm. LFT showed increased bilirubin with a total bilirubin of 2.9mg/dl and direct bilirubin of 1.0mg/dl,APTT was 37 seconds,PT of 18 sec with International normalised ratio of 1.4.CUE, RFT, Serum electrolytes are within normal limits.CECT Abdomen revealed enlarged liver of 18cm and enlarged spleen of about 18.8cm.ECG and 2DEcho are normal,Sr. ferritin around 8900ng/ml.Sickling test was negative.Serum electrophoresis revealed increased HBA2 levels.Peripheral smear revealed haemolytic anaemia with microcytic hypochromic cells.

Patient was shifted to the operation theatre after transfusion of 1 unit of FFP.All standard monitors were connected and baseline vitals were recorded as blood pressure of 120/90mmhg,pulse rate of 94 beat/min,Spo2 was 98% on room air. The child was premedicated with Inj. Ondansetron 3mg IV, Inj. Glycopyrrolate 0.2mg IV,Inj. fentanyl 50mcg IV, Inj. Midazolam1 mg IV,Induction was done with Inj. Propofol 60mg IV,Inj. Succinyl choline 50mg IV was given,Intubation was performed successfully with 5.5 uncuffed ET tube and a 4.5Fr Triple lumen was catheterized Ins into right internal jugular vein.Blood pressure showed a trend toward Hypertension intraoperatively with Systolicblood pressure around 150mmhg and diastolic blood pressure around 90mmHg. Anaesthesia was maintained with Oxygen and N2O in 2:2 ratio; intermittent Vecuronium boluses and isoflurane. Multimodal analgesia was given. Ventilation was adjusted to prevent Hypercarbia and airway pressures were monitored. Owing to the risk of alloimmunization, a blood transfusion unit was initiated at the end of the procedure titrated to blood loss.Warm fluids were transfused to prevent hypothermia.

With the return of adequate respiratory efforts neuromuscular blockade was reversed with inj. Neostigmine 4mg IV and inj. Glycopyrrolate 0.2mg IV.Patient was extubated after thorough throat suctioning.Child was transferred to PICU and kept on Inj.Paracetamol for post operative analgesia and O2 supplementation. Haemoglobin improved gradually and the patient was discharged 7 days later from the ward.

DISCUSSION:

Beta thalassemia Major is [3,4,5] Characterised by impaired production of Beta Globin chain.

This imbalance of Globin chain synthesis results in Extravascular Haemolysis,profound anaemia, Erythroid hyperplasia,extramedullary haematopoiesis, hepato splenomegaly, severe bone deformities and growth retardation. Extramedullary Haematopoiesis results in maxillary bone enlargement causing difficulty in airway management.Preoperative evaluation should focus on organs affected by Hemochromatosis following multiple transfusions such as heart, liver, and endocrine system.

Management of β-thalassemia is mainly supportive such as frequent blood transfusions and definitive therapy includes new strategies[3] such as iron chelating agents ,bone marrow transplantation, haematopoietic stem cell transplantation, gene therapy and splenectomy in case of a splenomegaly.

Splenectomy is usually needed around 6-8 years of age when transfusion requirement is 1.5 times the normal.Patient may present with asymptomatic restrictive lung disease,Lung function tests are useful for diagnosis of this condition.

Patients may present with Pulmonary Hypertension.As its prudent intraoperatively, the conditions that worsen pulmonary hypertension such as hypoxia, hypercarbia & acidosis should be avoided.

Challenges for anaesthesiologists in such patients would be management of anaemia, difficult airway [4,6] due to extramedullary haematopoiesis; pulmonary hypertension and intraoperative Systemic hypertension.Severe ventilation perfusion mismatch due to restrictive lung disease caused by hypersplenism and decreased oxygen carrying capacity. The patients are immunocompromised thus,aseptic vigilance should be maintained at all times and appropriate antibiotic prophylaxis must be recommended.In these patients there is an increased risk of wound healing postoperatively[7]. Osteopenia and Microfractures necessitate careful transfer and positioning.

CONCLUSION:

Haemoglobinopathies are large group of disorders with many challenging situations and a close collaboration between the Surgeon, Anaesthesiologist and Haematologists is essential to ensure safe and optimum management of β thalassemia major.

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