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ANESTHETIC MANAGEMENT OF PEDIATRIC SPLENECTOMY WITH THALASSEMIA MAJOR AND EVANS SYNDROME : A CASE REPORT

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ABSTRACT In Beta-thalassemia major disease, patient need multiple blood products which are regular and frequent which ultimately leads to autoimmunization reactions . In our case of an 8 year old male child , who received frequent blood transfusion because of Beta-thalassemia, diagnosed with Evans syndrome ; posted for Splenectomy. As Evans syndrome leads to autoimmune pancytopenia. Here we report such case in view to increase awareness about disease and prevent it by regular screening and prenatal diagnostic modalities.

KEYWORDS : Beta-thalassemia, Autoimmune pancytopenia, Evans syndrome, Splenectomy

INTRODUCTION

Thalassemia presents mainly with two main degrees of severity ; 1.Thalassemia Major 2.Thalassemia minor. Thalassemia Major is homozygous type while the minor one is hetrozygous type [1]. In which haemoglobin molecule,alpha or beta globin chain synthesis is decreased. Patients with β-Thalassemia major require life-long regular blood transfusions. They are at risk of developing antibodies against RBCs (both allo-and autoantibodies). When auto-antibodies are present against two or more blood cells it is known as Evan syndrome (ES). Management of these patients is difficult as these patients develop change in upper airway anatomy due to heavy iron deposition apart from that also there were high blood pressure variation intra-operatively, pulmonary hypertension & endocrinal abnormalities [2]. Iron overload, chelation therapy, multiple blood transfusions can affect multiple organ systems. Evans syndrome is mainly due to idiopathic thrombocytopenic purpura and autoimmune haemolytic anemia with no underlying disorder ,as it is autoimmune in origin.

Case Presentation

An 8 year old child weighing 18 kg born of non-consanguineous marriage diagnosed at the age of 7 year with Beta-thalassemia major,developed autoimmune pancytopenia (Evans syndrome), received multiple blood transfusion (monthly), and had complaint of abdominal pain that gradually progressed after the last treatment. A year and a half ago a splenectomy was prescribed. His history revealed that he had increasing blood transfusion requirement in recent time. Child taken all vaccines 3 months prior to surgery like Pneumococcal, Influenza & Meningococcal vaccines. Last PCV given was about 12 hour ago to surgery. Patient was on Tab DEFERIPRONE (500 mg)TDS, Tab DEFERISIROX (500 mg)OD, Tab PREDNISOLONE (5mg)TDS since 7 month. As hepato-splenomegaly progressively increase ; Splenectomy was planned.

On examination of airway of patient, It was found normal with adequate mouth opening with Mallampati Grade-2. He was pale, icteric,afebrile with HR-108/min, BP-106/72 mmHg, RR-24/min, Bilateral lung fields clear on auscultation, S1S2 was heard as normal and Oxygen saturation (SpO2) is 96% on room air. Patient had haemolytic facies like; frontal bossing, depressed nasal bridge, malar prominence & high arched palate, increase in size of liver and spleen(liver 6 cm and spleen 15 cm below costal margin in midclavicular line), severe pallor, jaundice, tachycardia, tachypnea, with no prominent neck veins and bilateral pedal edema.

Child had severe pancytopenia with Hemoglobin-1.8 gm/dL, Hematocrit-4.7 %, RBC counts-1.02 million/cumm, Platelet counts 18,000/cumm. Total serum bilirubin was 2.2 mg/dL.Two units of packed cell volume (PCV) transfusion was done at 12 hours interval on first day. He was further transfused with 2 units of Packed cell volume and 1 unit of random donor platelets (RDP). At the same time direct antiglobulin test (DAT) and indirect antiglobulin test (IAT) were sent, both were strongly positive. On day of surgery child had Hemoglobin-9.6 gm/dL,Platelet counts-2,37,000/cumm,S.ferritin-1000 ng/ml (17-140 ng/ml),Total serum bilirubin was 2.6mg/dL.Serum electrolytes and Renal fuction test were within normal limits.2D echo findings were 60 % Ejection fraction with mitral valve prolapse Grade-3 ,Midvalvular pulmonary stenosis and Midvalvular aortic stenosis. Ultrasonography abdomen suggested increases in Liver size with 14.5 cm and spleen size of18.5 cm. We decided to conduct this case under general anaesthesia after taking proper verbal & written consent from patient's relatives and patient was kept nil per orally for eight hours before surgery. Two units of packed cell volume (PCV) arranged for surgery.

Anesthetic Management:-

In Operation theatre patient was laid supine, routine monitoring were attached which includes non-invasive blood pressure, electrocardiogram, oxygen saturation(spo2).The Operation was performed under general anesthesia.Initial findings were HR-130 bpm ,BP-124/96 mmHg and SpO_2 of 98%.

Child was given intravenous Injection of midazolam 0.4 mg, Ondensetron 0.16 mg and Glycopyrrolate 0.1mg for premedication.Preoxygenation was done with 100% oxygen for adequate duration.Induction was done with intravenous sedatives like Injection propofol 40 mg,after checking proper mask ventilation short acting muscle relaxant like intravenous Injection succinylcholine 40 mg was given, after attempting direct laryngoscopy we showed partial view of glottis with Cormack Lehane grading of class IIA; endotracheal tracheal intubation was performed with using 4.5 mm cuffed endotracheal tube , Bilateral air entry checked which was found equal and adequate, tube was fixed at 17 cm depth. Anesthesia was maintained with Entonox and 2% sevoflurane and intermittent boluses of 2.0 mgiv atracurium. Ryles tube insertion was done. There were not much changes in hemodynamic parameters during intraoperative time. Estimated blood loss was 200 ml and replaced with 300 ml of ringer lactate and 1 unit PCV. Duration of surgery was about 2 hours with urine of 250 ml is collected. After surgery, when patient's spontaneous respiratory efforts were regular and adequate, neuromuscular blockade was reversed with Injection neostigmine 0.9 mg/iv and Injection glycopyrolate 0.2 mg/iv. ET tube was removed when patient was fully awake and perform all motor movements. Patient was shifted to recovery room. Postoperative period was uneventful. Paracetamol suppository was inserted for postoperative analgesia.

DISCUSSION

When patients with Beta-thalassemia posted for splenectomy; proper laboratory investigations depending upon patient's clinical condition is must before surgery.A careful systemic multimodality approach is required. In beta thalassemia patient erythropoesis is ineffective as it can not compensate for lack of beta chain so erythropoesis is increased but extramedullary in origin which causes craniofacial deformities like maxillary bone growth with dental protrusion and malocclusion, zygomatic bone expansion with nasal bone depression leads to characteristic chipmunk appearance ,ultimately leads to difficult airway so preoperatively bed side assessment of airway is required[3].
In patients with thalassemia major,there were chronic RBC destruction with nitric oxide changes which will cause vessel wall damage and high pulmonary artery pressure .[4]In these patients, treatment is mainly frequent blood transfusions and iron chleation. In case of splenomegaly,splenectomy is advised.For better outcome, stem cell transplantation [bone marrow transplantation] is required.[5] As requirement of blood transfusion is increases,splenectomy required between 6 to 8 year of age which can be postpone upto 5 more years in

case of sepsis.[6] As there is risk of iron overload which will deposit in heart, liver, pituitary; manifesting as various complications like cardiomyopathies, Liver cirrhosis & pituitary dysfunctions so pre-operative investigations include S.iron, ferritin, exercise testing.Preoperative echocardiography is recommended as it tells about contractility, size and pressure changes in both atria and ventricles. Intra-operatively low level of inhalational anaesthetic agent is recommended as high level leads to cardiovascular collapse. During long surgical procedures it is advised to kept legs up in slight trendelenburg position to prevent leg ulcers, deep vein thrombosis and pathological fractures. [7]

These patients are immunocompromised so increase chances of infections in them. As multiple blood transfusion are required, they are also at increase risk of diseases which are transmitted by blood products like Hepatitis, HIV, Syphilis etc. Intra-operatively to avoid hypercarbia, acidosis, hypoxia and cardiovascular dysfunctions in these patients ; diagnosis of any pre existing respiratory pathology, pre-operative PFT etc are required. [8] Precautions are taken for prevention of deep vein thrombosis as thalassemia is hypercoagulable state.[9]

Acute myocarditis is common in these patients which present as cardiac dysfuction. [10] In Beta-thalassemia major patients; repolarisation abnormalities are there, which present as increase QT interval.[11] Difficult intubation with Cormack Lehane IIA was observed in such cases. Possible causes of hypertension include repeated blood transfusion during whole life and autotransfusion during splenectomy.[8] fio2 was maintained at least 50% during surgery as chances of ventilation perfusion mismatch is high.[12] We managed successfully with combined use of volatile anesthetic agents and beta-blockers.Post-operative follow-up of the such patients showed that the Hb percentage improved and the requirement of blood transfusion reduced.

CONCLUSION

Beta-thalassemia major is present with difficult clinical conditions which required meticulous anaesthetic approach with preservation of blood products with required drugs for management of peri-operative complications of splenectomy. A team approach between various medical specialities is must for management of such cases and better prognosis in thalassemia major patients.

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