
Case Report

Anaesthetic Management in an Adult Thalassemia Major Patient Undergoing Splenectomy: A Case Report

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ABSTRACT

Thalassemia major patient requires multiple transfusions therefore splenectomy is a common surgery performed in this group of patients to decrease the frequency of transfusion and mechanical effect of enlarged spleen. Anaesthetic management in these patients are challenging being associated with unanticipated difficult airway, cardiac diseases, restrictive respiratory pattern, perioperative high blood pressure, endocrinological abnormalities, iron overload and pulmonary hypertension. We report a case report of anaesthetic management in an adult thalassemia major patient with massive splenomegaly posted for splenectomy. A 25 year old female, suffering from thalassemia major since the age of six months, with increasing frequency of blood transfusions ranging from once in a month to once in a week and presently upto twice in a week. She presented with chief complaints of pain and mass per abdomen, which was gradually progressive since last one and a half year. Systemic examination revealed prominent abdominal veins, hepatomegaly 10 cm below right costal margin (RCM) and splenomegaly 15 cm below left costal margin (LCM). Ultrasonography suggested liver measuring 23 cm with fatty infiltration and spleen measuring 23 cm with splenic vein diameter of 13mm. She was given general anaesthesia for the surgery. Many case reports in medical literature are about anaesthetic management of paediatric thalassemia major patients undergoing splenectomy and some case reports of anaesthetic management in adult beta thalassemia intermedia (milder disease) patients undergoing splenectomy. To the best of our knowledge, this is the first case report of anaesthetic management in an adult female with thalassemia major undergoing splenectomy.

Key Words: Thalassemia major, Anaesthetic management, splenectomy, spleen.

INTRODUCTION

Thalassemias are a group of hereditary disorders, with two major degrees of severity: Thalassemia major is the homozygous type with severe degree of symptoms; thalassemia minor is the heterozygous type with decrease severity of symptoms. ^[1] Thalassemia major patient requires multiple transfusions therefore splenectomy is a common surgery performed in these groups of patients to

decrease the frequency of transfusions and mechanical effects of enlarged spleen. Anaesthetic management in these patients are challenging being associated with unanticipated difficult airway, cardiac diseases, restrictive respiratory pattern, perioperative high blood pressure (BP), endocrinological abnormalities, iron overload and pulmonary hypertension. ^[2] We hereby present a case report of anaesthetic management in an adult

thalassemia major patient with massive splenomegaly posted for splenectomy and review of literature.

CASE REPORT

A 25 year old female, weighing 43 kg, unmarried, suffering from thalassemia major since the age of six months, presented with chief complaints of pain and mass per abdomen, which was gradually progressive since last one and a half year. Her history revealed that she was on multiple blood transfusions with increasing frequency from once in a month to once in a week and presently up to twice in a week. On admission her haematological parameters were haemoglobin (Hb) as 6.1 gm/dl and platelet count as 50,000/cm³ without any overt sign and symptom of bleeding.

On examination, she had frontal bossing, malar prominence, depressed nasal bridge and high arched palate. Airway assessment was found normal and Mallampati grade 2 with bucking of teeth. She was pale, afebrile with a heart rate (HR) of 126 bpm, Blood pressure (BP) of 116/78 mmHg, respiratory rate of 16 cycles/min, bilateral lung fields clear, S₁S₂ was heard as normal with haemic systolic murmur and oxygen saturation of (SpO₂) 96% on room air. Systemic examination revealed prominent abdominal veins, hepatomegaly 10 cm below right costal margin (RCM) and splenomegaly 15 cm below left costal margin (LCM) [Fig. 1].



Fig. 1: Clinical photograph of patient showing protruding abdomen because of Hepatosplenomegaly

Hematological investigations revealed Hb of 10.7 gm/dl after five units of packed cell transfusion, total leucocyte counts 12,000/cm³ and platelet count of 80,000/cm³ after four units of platelet concentrate transfusion. Peripheral blood smear revealed microcytic hypochromic anemia with thrombocytopenia. Serum ferritin 8157 ng/ml (N 17–140 ng/ml), liver function tests, renal function tests, coagulation profile and serum electrolytes were within normal limits. Viral markers (HIV, HbsAg and HCV) were negative. Ultrasonography suggested liver measuring 23 cm with fatty infiltration and spleen measuring 23 cm wherein splenic vein diameter was 13mm. She had received pneumococcal, haemophilus influenza and meningococcal vaccinations prior to surgery.

In view of massive hepatosplenomegaly which was still progressively increasing and requirement for multiple blood transfusions, splenectomy was planned by the surgeon. We decided to conduct this case under general anaesthesia and patient was kept nil per orally for eight hours before surgery. Premedication was done with tablet ranitidine 150 mg and tablet alprazolam 0.5 mg a day and two hour before surgery. Upon arrival into the operating room, patient was laid supine, routine monitoring like non-invasive blood pressure (NIBP), electrocardiogram (ECG) and oxygen saturation (SPO₂) were established. Peripheral venous access was secured with 16 gauze cannula. Baseline HR was 124 bpm, NIBP 122/76 mmHg, SPO₂ 96% on room air and respiratory rate (RR) 18 cycles/min. Preoxygenation was done with 100% oxygen for 3 minutes. Induction was done with injection fentanyl 100 ug/iv and injection propofol 100 mg/iv, after checking ventilation injection atracurium 25 mg/iv was given, patient was then ventilated for 3 minutes and airway was secured with cuffed endotracheal tube (ETT) of size 6.5 mm. Bilateral air entry checked, found equal and adequate, tube was fixed at 20 cm depth.

Anaesthesia was maintained with oxygen: nitrous (50%: 50%), 1% sevoflurane and intermittent boluses of 5mg/iv atracurium. On the request of surgeon nasogastric tube was inserted without any difficulty and secured. Surgery started with left subcostal incision. The haemodynamic variables remained stable throughout the intraoperative period. Two ringer lactate, one hydroxy ethyl starch (HES), one units packed cell, two units platelet concentrate and one unit fresh frozen plasma was transfused intraoperatively. Surgery lasted for two hour with urine output of 350 ml and blood loss of 850 ml [Figure 2 and 3]. Injection paracetamol and ketorolac was given for postoperative analgesia.



Fig. 2: Photograph showing resected spleen of size 28cm×16cm.



Fig. 3: Post splenectomy clinical photograph showing enlarged liver bulging from the surgical wound

After surgery, when patient's spontaneous respiratory efforts were equal and adequate, neuromuscular blockade was reversed with injection neostigmine 2 mg/iv and injection glycopyrolate 0.2 mg/iv. ETT

was removed when patient was awake and obeying commands. Patient was shifted to recovery room from where she was shifted to surgery ward after observation and monitoring for one hour. Postoperative period was uneventful and patient was discharged five days later from the surgery ward.

DISCUSSION

Thalassemia is an autosomal recessive disorder. It is characterized by impaired production of normal globin chain (alpha and beta) resulting in excess of one type of chain. Beta thalassemia is characterized by impaired synthesis of the beta chain, whereas alpha thalassemia results from impaired synthesis of the alpha chain. [1] This globin chain synthesis imbalance ($\alpha \gg \beta$) results in intravascular hemolysis, extramedullary hematopoiesis, erythroid hyperplasia, profound anemia, severe bone deformities, hepatosplenomegaly, growth retardation and death by second or third decade. [2]

Pulmonary hypertension and progressive vascular damage occur because of chronic hemolysis and disturbed nitric oxide physiology. [3] Maxillary bone enlargement due to extramedullary hematopoiesis results in airway difficulty. [3] Preoperative evaluation should thus include examination of organs like heart, liver, spleen and endocrine system (pancreas and pituitary) affected by hemochromatosis following multiple transfusions. Management includes supportive treatment like multiple blood transfusions. Splenectomy is indicated only when there is splenomegaly or treatment of transfusion related iron overload. Allogenic bone marrow (stem cell) transplantation can cure severely affected patients. [4]

Many case reports in medical literature are about anaesthetic management of paediatric thalassemia major patients undergoing splenectomy [2,5] and some case reports of anaesthetic management in adult beta thalassemia intermedia (milder disease) patients undergoing splenectomy. [6,7] These

authors discuss about the perioperative problems such as perioperative hypertension, potential difficult airway, severe anemia, etc. There are case reports about thalassemia with other diseases like hypothyroidism, systemic lupus erythematosus, and Eisenmenger's syndrome too. [8,9]

To the best of our knowledge, there is paucity of literature regarding anaesthetic management in an adult female with thalassemia major undergoing splenectomy, as in this patient, her disease was diagnosed very early in her childhood (at 6 months of age) and had a history of frequent blood transfusions with massive splenomegaly. Challenges for anaesthesiologists in such patients are management of difficult airway (due to extramedullary hematopoiesis), anemia (which persists in spite of transfusion), pulmonary hypertension and intra operative systemic hypertension severe V/Q mismatch due to restrictive lung disease caused by hypersplenism and decreased oxygen carrying capacity (severe anemia). There is a high incidence of blood transfusion related diseases like hepatitis hence precaution must be taken to avoid exposure to blood and body fluids. [10]

In our patient, her MPG grade II, bucked teeth, frontal bossing, malar prominence, depressed nasal bridge and high arched palate made the airway slightly difficult and hence intubation was planned to secure it. Anaemia was an important sign in our patient with preoperative Hb ranging from 6.1 gm% to 10.7 gm%. Hence blood transfusions were given as required. Our patient also received iron chelation therapy, which is actually beneficial in preventing cardiac dysfunction and reducing liver iron concentrations. The massive size of spleen could lead to decompressive symptoms, which were taken care of by timely loading the patient with colloids and iv fluids. Short acting drugs like propofol, fentanyl and atracurium for muscle relaxation were given, keeping in view of hepatomegaly although LFTs were within normal limits. The FiO_2 was kept 50% or above

throughout the surgery, to avoid any V/Q mismatch and pulmonary complications. Immunization with pneumococcal, haemophilus influenza and meningococcal vaccinations prior to surgery were given to prevent post-splenectomy infection.

CONCLUSION

Thalassemia major is a common form of haemoglobinopathy in developing countries like ours and many times it present with clinically challenging situations with difficult management. A carefully planned balanced anaesthesia technique with adequate supplementation of blood, blood products and required drugs for surgeries like massive splenectomies become a must in these patients. Moreover, a well qualified, vigilant and collaborative team work between multiple medical specialties is essential for optimal management and better survival of the patient with thalassemia major, despite their incurable genetic disease.

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