RESEARCH PAPER | Medical Science | Volume : 4 | Issue : 4 | Apr 2014 | ISSN - 2249-555X

Anaesthetic Management for Splenectomy in Thalassemia Minor Complicated With SLE and Hypothyroidism

KEYWORDS

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Introduction

Thalassemia[1,2] is an inherited autosomal recessive disorder. It occurs in two major degrees of severity. Thalassemia minor is the heterozygous type with decreased severity of symptoms. Thalassemia major is the homozygous type characterised by decreased production of normal globin chains resulting in intravascular hemolysis, profound anemia, erythroid hyperplasia, extramedullary hematopoiesis, splenomegaly and severe bone deformities. It usually results in death by 2nd decade of life[3].

Splenectomy[2,4] is usually performed in these patients when their transfusion requirements increase and when they develop features of hypersplenism with splenomegaly. With careful and vigilant monitoring and perioperative management, splenectomy in children with massive splenomegaly is rewarding as the transfusion requirements decrease markedly postoperatively and there is marked improvement in hematological parameters post splenectomy[4].

We hereby describe the anesthetic management of splenectomy in thalassemia minor complicated with SLE, hypothyroidism. There is insufficient literature regarding anesthesia in such a combination of disorders.

Case Report

A 8 years old male child (Fig 1) weighing 11kg (ht. 38.5cm), a known case of thalassemia minor with massive splenomegaly presented for splenectomy.

Thalassemia was first detected at 8 months of age. At that time, the transfusion requirement was once per month which increased to twice per week when the patient was presented for surgery. On admission, he was pale with Hb of 6.7, Hct 20.2%, and platelet count of 11000/cu mm with no overt evidence of bleeding. On examination, there was enlargement of liver (3cm below right costal margin) and spleen (6cm below costal margin). On examination, his abdominal girth was 21cm at umbilicus and 24cm 3cm above umbilicus. He had failure to thrive with bronzing discoloration of skin. Examination of the respiratory system was unremarkable except a slight increase in respiratory rate. Examination of the cardiovascular system revealed tachycardia with hyperkinetic precordium . Tests for viral markers like Hepatitis C, Hepatitis B and HIV were negative.

He had skin ulcers on calf and medial side of leg (Fig 2) . On preoperative assessment, it was found that he was also suffering from SLE and hypothyroidism. He was diagnosed SLE with positive reports of serum antinuclear antibody, dsDNA +ve, ESR 90 mm in 1st hour, CRP +ve and was started on prednisolone by dermatologist on admission. His investigations revealed hypothyroidism with initial TSH of 10.4 which decreased to TSH 5.5 after starting treatment with levothyroxine on admission.

Peripheral blood smear showed mild anisopoikilocytosis, some microcytic hypochromic cells with reduced platelets with normal WBC counts.

USG scan of abdomen showed hepatosplenomegaly with liver span of 15.8cm and spleen 13.1cm.

Patient received vaccination with Hib, pneumococcal and meningococcal vaccine before splenectomy.

He received packed RBCs and platelets before surgery. Prednisolone therapy and levothyroxine treatment continued till the day of surgery with planning for intraoperative steroid coverage.

Preoperative blood investigations revealed Hb of 7.3, hematoctit 20.2%, MCV 84.9, MCH 28.2, MCHC 33.2 and platelet count 51000/cu mm. Serum electrolytes, LFT, RFT and blood sugar reports were within normal limits. Serum T4 and TSH were 1.33 and 5.53, respectively.

On the day of surgery, in the operation theatre, the child was premedicated with Inj. Midazolam to calm him and then monitors were attached. Pulse oximeter, NIBP, temperature, ECG and EtCO2 monitoring were used.(monitor with name). Preinduction heart rate was 115/min.,NI BP 142/103 mm Hg, SpO2 95% on room air with respiratory rate of 32/min. After preoxygenation, induction was done with preexisting I.V. line with Inj. Fentanyl 2µg/kg, Inj. Thiopentone 5mg/kg and Inj. Succinylcholine 1.5 mg/kg. The trachea was intubated with uncuffed ETT of 5.5 ID fixed at 15 cm. Bilateral air entry was checked. Post induction, one more I.V. line was secured with 22G I.V. cannula.

Splenectomy was performed through left subcostal incision (Fig 3) . Our anesthesia technique was balanced approach utilizing low concentration of isoflurane (0.4-0.6%) in N2O + O2, with Inj. Fentanyl and Inj. Atracurium. Patient was transfused with one unit of packed RBC's and one unit of packed platelets after the spleen was removed. The trend towards hypertension intraoperatively was managed successfully with Inj. Propofol repeated as required. The hemodynamic variables remained stable intraoperatively except for slight increase in NIBP (range). Nasogastric tube was inserted and secured.

Volume : 4 | Issue : 4 | Apr 2014 | ISSN - 2249-555X

The duration of surgery was 1 hr with urine output of 150mL and blood loss was around 100-150ml. After surgery, patient's trachea was extubated when he was awake and shifted to recovery room from where he was shifted to ward after monitoring for 1 hr.

The postoperative period was uneventful and the patient started accepting orally. However, on further follow up it was found that he had an episode of generalized tonic clonic seizure after 1 week postsurgery and developed neurological deficits. MRI scan was done which showed mutiple hyperdense areas suggestive of multiple infarcts likely due to vasculitis. The patient was managed conservatively in the ward and then discharged on steroid, antibiotic, levothyroxine and ecosprin.

DISCUSSION:

Thalassemia is an autosomal recessive disorder which may be associated with other hemoglobinopathies like sickle cell anemia[5] and autoimmune diseases like SLE with resultant increase in morbidity. Rarely the transfusion frequency may increase dramatically in these patients due to autoimmune hemolytic anemia due to SLE as reported by Paul et al[6].

Castellino et al[7] in their landmark study reported and analyzed the association of β thalassemia trait and SLE. They described the clinical manifestations in patients with SLE with and without β thalassemia. They concluded that when these two conditions coexist, SLE seem to have a more severe course and a greater prevalence of CNS involvement. In our patient seizures in the postoperative period were attributed to vasculitis(SLE) . However thromboembolic phenomenon could not be ruled out and the patient was started on ecosprin with steroid by the pediatrician.

Though autoantibodies used to diagnose SLE may be falsepositive in multitransfused thalassemic cases but anti ds DNA is rarely positive. In our patient both ANA and dsDNA were positive which confirmed the diagnosis of SLE.

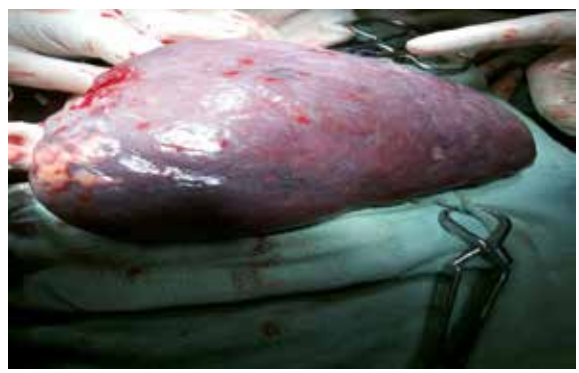
A case report by Pinto et al[8] described a case of SLE with sickle$/_{\beta+}$ - thalassemia.

Anaesthetic management of splenectomy in these patients deserves special attention due to various complications arising due to SLE and other autoimmune disease with multisystem involvement. However, in our patient, the manifestations of SLE were limited to skin in the preoperative period and the patient was started on prednisolone therapy preoperatively. The transfusion requirements had increased too much with failure to thrive which was amenable to treatment only by splenectomy.

In our patient, slight hypertension was observed intraoperatively which was managed successfully with repeated doses of Inj. Propofol. Various authors[9,10] have suggested that manipulation of large spleen during surgery leading to autotransfusion may be a possible cause of hypertension in these patients. A retrospective study[10] of 100 thalassemic children undergoing splenectomy reported intraoperative and postoperative hypertension requiring aggressive treatment.

Altnoz et al[11] in their study discussed the association of β thalassemia trait with autoimmune diseases. They attributed these associations to the proximity of hemoglobin β chain locus at 11p15.5 to eight genes with profound role in immunity : STIM1, CD151, TC21/RRAS2, SIGIRR/TOLL/IL1R8, pp52/LSP1 (lymphocytes specific protein), TRIM21, toll interacting protein (TOLLIP) and SLEN3. They also reported that the association with autoimmune disease may be due to altered concentrations of hemorphins. Reduced concentrations of hemorphins in thalassemia heterozygosity may lead to proinflammatory stage and autoimmune vulnerability.

Hence association and presence of autoimmune diseases like SLE in thalassemia patients need special consideration and aggressive management.



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