

Anaesthetic Management of a Case of Hereditary Spherocytosis for Splenectomy and Cholecystectomy

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Abstract

Case Report

We could successfully manage the patient with HS posted for laparoscopic splenectomy and cholecystectomy. Perioperative management of HS largely depends on the severity of anaemia and the degree of haemolysis. Anemia should be corrected preoperatively before major surgery. Surgery should be avoided in the presence of haemolytic crisis. Anesthetic challenge includes prevention of hypoxia, acidosis and hypothermia. Vaccination before splenectomy is a must to prevent postoperative infections. Perioperative HS management should include blood transfusions, aggressive hydration, and prevention of hypoxia, acidosis, and temperature regulation [1, 2]. We report the case of a patient with this disorder presenting for laparoscopic cholecystectomy and splenectomy, under general anesthesia.

Keywords: Hereditary spherocytosis; anesthesia management; splenectomy; cholecystectomy.

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INTRODUCTION

Hereditary spherocytosis (HS) is a common cause of non-immune hemolytic anemia due to transmembrane protein abnormalities in RBCs [1]. The case is reported because of the difficulties encountered by the anesthetist due to complications due to 'sickling' based complications. Perioperative HS management should include blood transfusions, aggressive hydration and prevention of hypoxia, acidosis, and temperature regulation [2]. We report the case of a patient with this disorder presenting for laparoscopic cholecystectomy and splenectomy, under general anesthesia.

CASE REPORT

A 17-year-old girl weighing 48 kg, a known case of HS, was candidate to laparoscopic cholecystectomy and splenectomy. Her mother was splenectomized and a brother treated too for HS. She gave a history of jaundice since she was 4 years old. There were also complaints of abdominal pain and episodes of jaundice for which she had been admitted in the past. On examination, a subicteric was seen. His vital signs were stable. Per abdomen examination revealed mild splenomegaly. Rest of the systemic review was unremarkable. A hemogram revealed a hemoglobin level of 9.1 g/dL, leukocyte and platelet counts within the normal range, a peripheral smear showing typical

spherocytes at 3% with normal mean corpuscular volume (figure 1), a raised reticulocyte count, osmotic fragility tests was normal, proteins electrophoresis showed the persistence of hemoglobin F, prothrombin time, international normalized ratio (INR) was 68%, and 1.7 respectively. Patient was Rhesus A⁺. The serum bilirubin level was 2 g / dL (unconjugated). Rest of the liver function tests, serum creatinine, and electrolytes were normal. On abdominal ultrasound and scanner, gallstones were present and there was an enlarged spleen (figure 2). The patient was immunized with pneumococcal and Haemophilus influenzae vaccines. 3 units of packed cells, 2 units of FFP were kept ready for an eventual peri-operative transfusion. Patient was premedicated with Atarax[®] (hydroxyzine dihydrochloride).

On the morning of surgery, after securing an 18G intravenous line, an infusion of saline solution was started. Routine monitoring were used (electrocardiogram, pulse oximeter, non-invasive blood pressure cuff and the capnograph was checked). Catheter for epidural analgesia was implanted and then a loading dose of 0.125 mg of bupivacaine was administered. After preoxygenation for 3 minutes, anesthesia was induced with intravenous propofol 3 mg/kg, fentanyl 5 µg/kg and rocuronium 0.6 mg/kg was used for relaxation. The trachea was intubated with a

6.5-mm internal diameter cuffed tracheal tube. Anesthesia was maintained with 50% oxygen in air with isoflurane. IV Fentanyl along with rocuronium were used intraoperatively. End-tidal carbon-dioxide was maintained between 30 and 35 mmHg, and intraoperative vitals were stable. Intraoperatively hypoxia, acidosis were avoided. Hypothermia was avoided by infusing hot intravenous fluids and increasing the operating room temperature. Cholecystectomy was done followed by extraction of the spleen through 6 cm left subcostal incision ("Hand-assisted laparoscopic splenectomy"). The surgery lasted 1 hour 30 minutes. She received 1 L of crystalloids during the surgery and her urine output was 120 mL. Blood loss was around 100 ml. Patient is extubated once awake with an estimated modified Aldrete score at 10. At the end of surgery, epidural morphine, IV paracetamol and nefopam was given for postoperative analgesia. Postoperative analgesia was multimodal using epidural morphine 5 mg once a day, nefopam and paracetamol if pain. Lovenox 12 hourly was started as thrombo-prophylaxis. From the 1st POD prophylaxis injection Penicillin was also started in addition to other medication. She had a good recovery.

DISCUSSION

Hereditary spherocytosis is the most frequent hereditary disease of erythrocyte membrane. It results from mutations affecting several sub-membrane proteins (Ankyrin, Spectrin, Band 3, Protein 4.2, Rhesus Complex, Protein 4.1) [3]. Genealogical study shows that 75% of cases are transmitted in a dominant mode, 25% in a recessive mode. Hemolysis severity is very variable, and it is often compensated by effective regeneration, explaining the frequency of not very obvious forms. Most pronounced forms seem to result from recessive transmission.

Clinically, anemia is the presenting complaint in nearly half of patients, although the disease severity varies widely among individuals. Mild compensated hemolysis is observed in about 20% of individuals, with the majority of affected people (60%) having moderate haemolysis with a hemoglobin 8-11 g / dL. The more asymptomatic forms of SH may not be identified until a haemolytic crisis develops during childhood, often triggered by a viral infection. Less commonly, an aplastic crisis develops secondary to parvovirus B19 infection.

The most severe forms of the disease are recessive and associated with α -spectrin and some ankyrins defects. The diagnosis is usually supported by examination of red cells on a simple blood smear: excess of spherocytes is usually obvious [4]. Osmotic fragility test is often positive. A recent cytometry technique using labeling with eosin 5'maleimide (EMA) seems to give sensitive and specific results [5]. In difficult cases, ektacytometric study, eventually supplemented by membrane biochemical study, may be

necessary. Splenectomy is the only efficient treatment of spherocytosis. It normalizes life of red cells and therefore suppresses any sign of excessive haemolysis. It is not required in each case, especially in clinical latency. Precocious revelation and when the anemia is not compensated, it should be so possible postponed until second infancy or adolescence, because of extreme sensitivity of asplenic children to *Streptococcus pneumoniae* and *Haemophilus b* infections so immunisation with their vaccines should precede splenectomy and penicillin prophylaxis would be started whose modalities and duration are not consensual [6;7]. If splenectomy is not indicated, a continuous supply of folic acid is required. If gallstones are present, a cholecystectomy may be performed simultaneously or at a later date. Moreover many patients with such sickling disorder have impairment to oxygen delivery secondary to pulmonary damage, widespread macro and microvasculopathy, increased blood viscosity, anaemia, impaired vascular regulation and disturbed nitric oxide signaling [8]. They therefore may have limited reserve to cope with further reduction in oxygen delivery. Thus the avoidance of hypoxaemia is the foundation of anaesthetic management of this patient. The risk of late post splenectomy thrombosis [9] is also sought out for our patient, and standard thrombo prophylaxis that is subcutaneous heparin was also started.

CONCLUSION

Peri-operative management of HS depends on the severity of anaemia and the degree of haemolysis. Anesthetic challenge includes prevention of hypoxia, acidosis and hypothermia. Vaccination before splenectomy is a must to prevent postoperative infections.

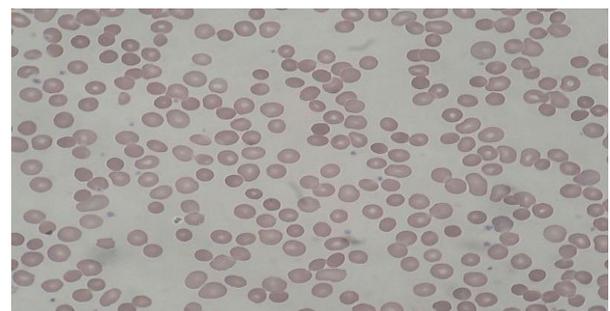


Fig-1: Abnormal peripheral blood smears of the patient



Fig-2: Abdominal scanner: gallstones and an enlarged spleen

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