Anaesthetic management of a case of hereditary spherocytosis for splenectomy and cholecystectomy

INTRODUCTION

Hereditary spherocytosis (HS) is a familial haemolytic disorder characterised by the production of red blood cells (RBC) that are sphere-shaped rather than bi-concave disk shaped (doughnut-shaped) and therefore more prone to haemolysis leading to chronic haemolytic anaemia. The incidence in Northern Europe and America varies from 1:2000 to 1:5000.[1] Patients with HS often present for surgery because of gall stones or for splenectomy. Exact incidence of HS in India is not known; however, the case is reported because of the challenges to the anaesthesiologist due to 'sickling' based complications. Peri-operative management of HS should include blood transfusions, prevention of hypoxia, acidosis and temperature regulation.[2]

CASE REPORT

We report a case of 28-year-old female weighing 40 kg, a known case of HS posted for laparoscopic splenectomy and cholecystectomy. She presented with pain abdomen and fever. She gave a history of jaundice since childhood. On physical examination, she had pallor, icteric sclera and splenomegaly. Rest of the systemic examination was normal. Her haemogram revealed haemoglobin (Hb) level of 6.2 g% and reticulocyte count of 18.5%. Peripheral blood smear showed spherocytes with increased osmotic fragility and Coombs test negative. Unconjugated bilirubin was 5.4 mg/dL, prothrombin time (PT), international normalised ratio (INR) was 27 s, and 2.0, respectively. Platelet count was 1.0 lakh/mm^3. Rest of the liver function tests, creatinine and electrolytes revealed no abnormalities. Abdominal ultrasound revealed gall stones and splenomegaly. Patient was immunised with pneumococcal and haemophilus influenza vaccines. She was hydrated with 1.5 L/day for 2 days and received 5 units of packed cells, 5 units of fresh frozen plasma (FFP). Injection vitamin K (10mg, IV BID) was administered preoperatively for two days. Post-transfusion, Hb was 9.9 g%, platelet count was 1.0 lakh/mm^3 and PT, INR was 18.1 s and

1.35, respectively. 3 units of FFP, 2 units of platelet concentrates and 2 units of packed cells volume were kept ready for intraoperative use.

On the morning of surgery, venous access was secured with a wide bore 16G intravenous (i.v) canula and infusion of Ringer's lactate started. She was pre-medicated with injection glycopyrrolate and fentanyl. After pre-oxygenation for 3 min, anaesthesia was induced with injection thiopentone sodium and succinylcholine was used to facilitate intubation which was done with 6.5 no. cuffed Portex® tube. Anaesthesia was maintained with O_2, N_2O, fentanyl, isoflurane and vecuronium. End tidal CO_2 was maintained between 30 and 35 mmHg. Intra-operatively hypoxia, acidosis were avoided. Hypothermia was avoided by infusing warm i.v fluids, warm blood and by wrapping the patient's extremities with cotton. Patient received 3 units of FFP, 2 units of platelet concentrates, 1 unit of packed cells after clamping of splenic vessels. Surgery lasted for 4 h. She received 2 litres of crystalloids and her urine output was 425 ml. Blood loss was 400 ml. At the end of the surgery, port sites were infiltrated with 0.25% bupivacaine. Trachea was extubated at the end of surgery. The patient received 1 unit of packed cells post-operatively. I V paracetmol infusions and I V fentanyl were used to alleviate post-operative pain.

DISCUSSION

Hereditary spherocytosis is a heterogeneous form of haemolytic anaemia associated with the presence of spherocytes in the peripheral blood. Classically, the inheritance of HS is autosomal dominant and some severe forms are autosomal recessive.[3] In HS, erythrocyte shape changes are caused by defects in membrane protein spectrin resulting in cytoskeleton instability resulting in spherical RBCs.

Spherocytic RBCs are removed rapidly from the circulation and destroyed by the spleen leading to anaemia, jaundice, enlarged spleen, and often gallstones. Moderate cases of HS may present in infancy with severe anaemia, hyperbilirubinemia, whereas mild cases are commonly seen in young adults or even later in life. Indeed, it is often the finding of gallstones in a young person that triggers diagnostic investigations. A characteristic feature of HS is an increase in mean corpuscular haemoglobin concentration (MCHC); this is almost the only condition in which an increased MCHC is seen. In most cases, the diagnosis can be made on the basis of red cell morphology and abnormal osmotic fragility, a modified version of which is called the "pink test." In some cases, a definitive diagnosis can be obtained only by molecular studies demonstrating a mutation in one of the genes underlying HS.[4]

The commonly encountered complications of HS are haemolytic crisis, aplastic crisis and megaloblastic crisis. High RBC turnover and heightened erythroid marrow activity in HS makes children vulnerable to develop aplastic crisis due to parvovirus and various other infections. Death may also occur due to severe anaemia, heart failure, and cardiovascular collapse.[5] Folate therapy is recommended in severe and moderate HS in order to prevent megaloblastic crisis.

Surgical treatment involves splenectomy, which results in cessation of haemolysis, return of the Hb to normal and clearance of jaundice. Surgery is indicated in all patients with splenomegaly to prevent gallstone formation and haemolytic crisis. Those who are well-compensated and asymptomatic may be treated conservatively.[6] Immunisation with pneumococcal and haemophilus influenza vaccines should precede splenectomy. If gallstones are present, cholecystectomy may be performed simultaneously or at a later date.

CONCLUSION

We could successfully manage the patient with HS posted for laparoscopic splenectomy and cholecystectomy. Peri-operative management of HS largely depends on the severity of anaemia and the degree of haemolysis. Anaesthetic goals include avoidance of hypoxia, acidosis, and hypothermia. Vaccination before splenectomy is a must to prevent post-operative infections.

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