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Anaesthetic Management of Laparoscopic Cholecystectomy and Splenectomy in Hereditary Spherocytosis: A Case Report

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Abstract

Hereditary Spherocytosis (HS), a non-immune hemolytic anaemia, results from red blood cell (RBC) transmembrane protein abnormalities. Surgical management, like laparoscopic cholecystectomy and splenectomy, poses challenges due to altered liver function and chronic hemolysis. A 20-year-old female with known HS underwent laparoscopic cholecystectomy and splenectomy. The preoperative assessment revealed a blood transfusion history, altered liver function, and anaemia. Examination showed pallor, icterus, and hepatosplenomegaly. Laboratory findings indicated microcytic hyperchromic RBCs and elevated bilirubin. Anaesthetic management involves premedication, induction, and intraoperative monitoring, with attention to hemodynamic stability. Anaesthetic considerations include avoiding acidosis, hypothermia, and hypotension, and managing deranged liver function and preoperative anaemia. Preoperative optimization and prompt complication management are crucial. Anaesthetic management in HS patients undergoing surgery requires an understanding of pathophysiology and complications. Multidisciplinary collaboration and meticulous perioperative care are essential for optimal outcomes.

Keywords: *Hereditary Spherocytosis, Anesthetic Management, Laparoscopic Cholecystectomy, Splenectomy, Hemolytic Anemia.*

Introduction

Hereditary spherocytosis (HS) is an extremely rare autosomal dominant family haemolytic disorder with significant clinical heterogeneity. Features that vary from the absence of symptoms to fulminant haemolytic anaemia. The estimated incidence in the Caucasian population ranges from 1:2000 to 1:5000 [1]. In HS, autosomal dominant inheritance accounts for 80% of instances; the remaining genes are either recessive or sporadic. It is characterized by the lack of two transmembrane proteins, spectrin and ankyrin, which attach the red blood cell bilayer to the membrane skeleton [2]. Aberrant spherocytes are sequestered and removed in hemolysis caused by the spleen. These patients may have anaemia, jaundice, splenomegaly, cholelithiasis, and altered liver function, which may affect how anaesthetic medications are metabolized. This is because the spherocytes are susceptible to osmotic lysis [3]. This case presents a unique scenario of a young patient with Hereditary Spherocytosis undergoing laparoscopic cholecystectomy and splenectomy, complicated by a history of blood transfusion, altered liver function, and anaemia.

Case Report

A 20-year-old female with hereditary spherocytosis (HS) is scheduled for

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laparoscopic cholecystectomy and splenectomy. She has a history of blood transfusion, with a height of 157 cm and a weight of 67 kg. Since 2014, she has been taking folic acid (0.5 mg OD). Additionally, she experienced jaundice two months ago. Preoperative evaluation reveals fair general condition with pallor and icterus, but no clubbing, cyanosis, or pedal edema. Vital signs are within normal limits, with a heart rate of 88 bpm, blood pressure of 120/80 mmHg, and oxygen saturation of 99% at room air. Airway assessment indicates adequate mouth opening, thyromental distance, and neck mobility, with no loose teeth or dental prostheses. Laboratory investigations show microcytic hyperchromic RBCs, elevated reticulocyte count, and positive osmotic fragility test consistent with HS. Imaging reveals cholelithiasis, splenomegaly (16 cm/13.7 cm/13 cm), and moderate hepatomegaly (Figure 1, 2).

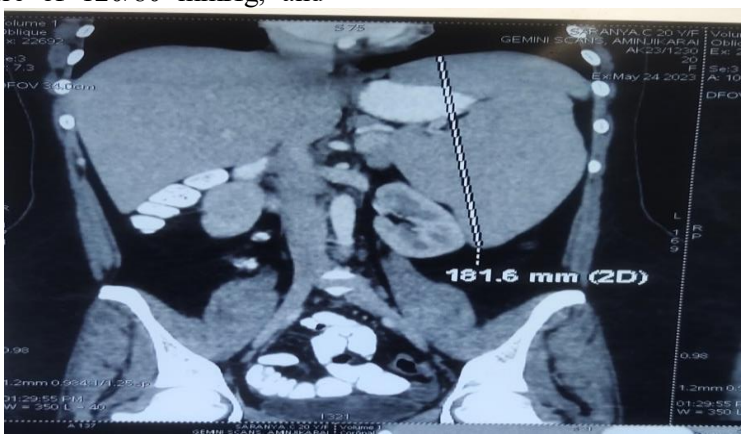

Figure 1. Spleen Enlargement in CT Scan

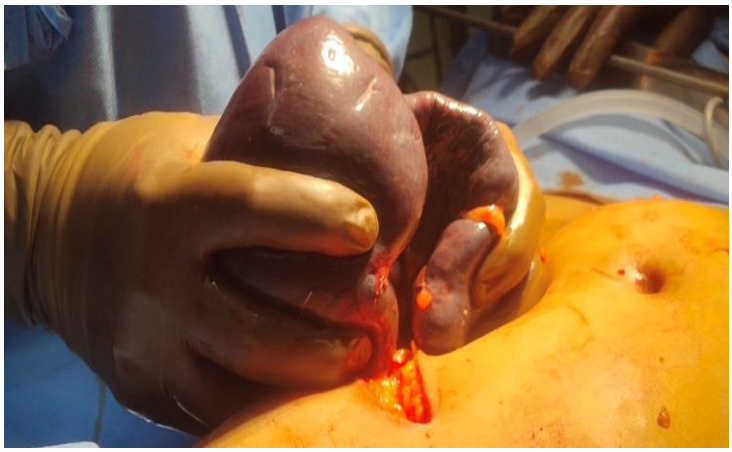

Figure 2. Splenectomy Intraoperative Pic

NPO status is maintained for 8 hours preoperatively, with premedication consisting of pantoprazole (40 mg) and metoclopramide. Adequate blood and blood products are reserved, and two wide-bore IV lines are secured, with standard ASA monitoring in place. Preoxygenation with 100% oxygen is performed, followed by premedication with IV glycopyrrolate (0.2 mg) and fentanyl (100 mcg). Anaesthesia induction is achieved with IV thiopentone (100 mg) and succinylcholine (75 mg), and intubation is facilitated using a Macintosh blade 3 and a COTT-7 tube. Bilateral equal air entry is confirmed with an ETCO2 trace, and a Ryles tube is inserted. Ventilation is initiated in VC mode, with muscle relaxation maintained using atracurium (initial dose of 20 mg and 5 mg boluses with TOF monitoring).

Intraoperative monitoring included ventilation in VC mode and administration of atracurium for muscle relaxation.

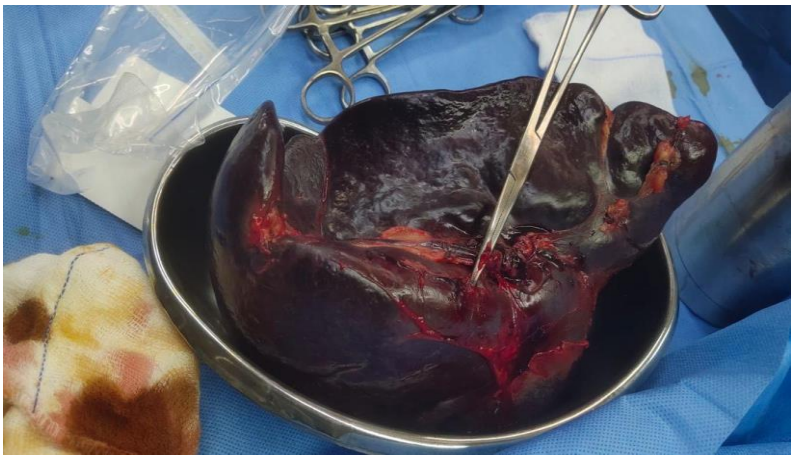

Figure 3. Hemosiderotic Spleen

Figure 3 shows that a large spleen measuring 22 cm necessitated meticulous surgical technique and intraoperative blood transfusion due to a significant blood loss of 400 ml. Postoperative analgesia was ensured through preperitoneal infiltration with bupivacaine, providing effective pain relief. The intraoperative period lasted for 4 hours, during which 2 units of packed red blood cells (PRBC) were administered. Fluid balance was carefully monitored, with a total input of 2500 ml and an output of 900 ml. Intravenous medications, including paracetamol and ondansetron, were administered to manage pain and prevent postoperative nausea and vomiting. Upon completion of the procedure, the patient was successfully extubated awake after the reversal of neuromuscular blockade. This comprehensive perioperative management approach ensured the successful completion of the surgery and facilitated the patient's smooth recovery postoperatively.

Discussion

Spherocytes in the peripheral circulation are linked to a heterogeneous form of haemolytic anaemia called hereditary spherocytosis. In the past, HS was inherited autosomally dominantly, however, some severe variants were inherited autosomally recessively [4]. Erythrocyte shape alterations in haemolytic anaemia (HS) are attributed to abnormalities in the membrane protein spectrin, which lead to cytoskeleton instability and spherical red blood cells. A rapid removal of splenocyte red blood cells from the bloodstream and their subsequent destruction by the spleen can result in anaemia, jaundice, an enlarged spleen, and frequent gallstones. While mild cases of HS are typically found in young adults or even later in life, moderate cases can manifest as severe anaemia and hyperbilirubinemia in infancy. Diagnostic tests are frequently initiated in response to the discovery of gallstones in a young individual.

An increase in mean corpuscular haemoglobin concentration (MCHC), which is almost unique to haemoglobin-storage syndrome (HS), is one of its defining characteristics. Red cell morphology and aberrant osmotic fragility—a modified test known as the "pink test"—can typically be used to make the diagnosis. In certain instances, only molecular testing proving a mutation in one of the HS-causing genes can lead to a conclusive diagnosis [5, 8].

Haemolytic crisis, aplastic crisis, and megaloblastic crisis are the most often occurring HS problems. Children with HS are more likely to experience aplastic crises as a result of parvovirus and other infections

because of their high RBC turnover and increased erythroid marrow activity. Heart failure, cardiovascular collapse, and severe anaemia can potentially be fatal [6, 9]. In cases with severe and moderate HS, folate medication is advised to avert megaloblastic crises. Splenectomy is the surgical procedure used to cure the condition. It stops haemolysis, returns the haemoglobin level to normal, and eliminates jaundice. For every patient with splenomegaly, surgery is necessary to avoid the development of gallstones and haemolytic crises. Patients who are asymptomatic and well-compensated might receive conservative care [7, 10].

The case demonstrates successful surgical management of laparoscopic cholecystectomy and splenectomy in a patient with Hereditary Spherocytosis (HS), showcasing effective multidisciplinary collaboration and meticulous anaesthetic management. Despite the complexity of the procedure and the patient's medical history, the favourable postoperative outcome underscores the expertise of the healthcare team and the comprehensive care provided.

Conclusion

In conclusion, the case underscores the intricacies of managing Hereditary Spherocytosis (HS) patients during laparoscopic cholecystectomy and splenectomy, highlighting the need for tailored perioperative strategies. By navigating through challenges associated with chronic hemolysis and altered liver function, the successful outcome emphasizes the significance of individualized care and interdisciplinary collaboration in optimizing patient safety and well-being.

Conflict of Interest

The authors declared no conflict of interest in the manuscript.

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