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ANAESTHETIC MANAGEMENT OF A PATIENT WITH HEREDITARY SPHEROCYTOSIS UNDERGOING SPLENECTOMY AND CHOLECYSTECTOMY: A CASE REPORT

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INTRODUCTION

Hereditary spherocytosis (HS) is a genetic hemolytic anemia characterized by spherical red blood cells prone to premature destruction. Clinical manifestations typically include anemia, jaundice, and enlargement of the spleen.

CASE PRESENTATION

We describe a patient diagnosed with HS presenting with splenomegaly and gallstones, scheduled for open cholecystectomy and splenectomy with common bile duct exploration. Clinical examination revealed pallor, jaundice, and splenomegaly, with stable vital signs. Laboratory studies showed hemoglobin of 11 g/dL, platelet count of 340,000/cu.mm, elevated prothrombin time/international normalized ratio (PT/INR 16.4/1.34), markedly raised total and direct bilirubin (91.3 and 55.7 respectively), elevated liver enzymes (SGOT 101, SGPT 114), and alkaline phosphatase (482). Peripheral smear demonstrated classic spherocytes with increased osmotic fragility.

After obtaining informed consent, the patient was transferred to the operating theater. Two 18-gauge intravenous cannulae were secured alongside right internal jugular vein cannulation for central access. The left radial artery was cannulated for invasive blood pressure monitoring. Baseline parameters were recorded. An epidural catheter was inserted at the T11-T12 interspace, with the catheter tip positioned 8 cm cephalad.

Premedication included glycopyrrolate 0.2 mg intravenously. After preoxygenation for three minutes, anesthesia was induced with fentanyl (2 mcg/kg), propofol (2 mg/kg), and muscle relaxation achieved with atracurium (0.5 mg/kg). The trachea was intubated with an 8.0 mm cuffed endotracheal tube. Bilateral lung ventilation was confirmed. Anesthesia maintenance consisted of oxygen, nitrous oxide, sevoflurane, and atracurium (0.1 mg/kg). Monitoring included electrocardiogram, invasive blood pressure, pulse oximetry, capnography, temperature, and urine output.

An epidural infusion of 0.25% bupivacaine with fentanyl 1 mcg/ml was initiated at 3 ml/hr and continued intra- and post-operatively. The surgical procedure lasted six hours, with an estimated blood loss of 800 ml. Fluid management included 3 liters of crystalloid solution. Rotational thromboelastometry (ROTEM) indicated prolonged clotting time in the EXTEM assay. Transfusion was guided by hematological consultation, resulting in administration of 1 unit of packed red blood cells and 5 units of fresh frozen plasma. Neuromuscular blockade was reversed with neostigmine (50 mcg/kg) and glycopyrrolate (10 mcg/kg). The patient was extubated following adequate respiratory effort and subsequently transferred to the intensive care unit for further monitoring.

DISCUSSION

HS is an autosomal dominant disorder caused by mutations in genes encoding ankyrin or beta spectrum proteins, leading to fragile spherical erythrocytes vulnerable to hemolysis. Perioperative management necessitates

correction of anemia through transfusion, vigorous hydration, and prevention of hypoxia, hypothermia, and acidosis that may exacerbate hemolysis. Maintaining normothermia and adequate hepatic blood flow is essential to prevent vasoconstriction and circulatory stasis. Stress should be minimized by providing effective analgesia and avoiding hepatotoxic drugs.

CONCLUSION

The anaesthetic approach for patients with HS must be tailored to the severity of anemia and hemolytic activity. The main objectives include avoiding hypoxia, acidosis, and hypothermia perioperatively. Vaccination against pneumococcus, Haemophilus influenzae, and meningococcus prior to splenectomy is critically important to reduce postoperative infection risk.

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