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Case Report / Olgu Sunumu

Management of Anesthesia in Hereditary Spherocytosis

Herediter Sferositozda Anestezi Yönetimi

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ABSTRACT

Hereditary spherocytosis (HS) is a type of hemolytic anemia. The severity of these outcomes varies from patient to patient, and splenomegaly occurs in every patient with HS. HS occurs mostly in young patients and very rarely in adults. The management of anesthesia is specific for these patients. We aimed to share our experience of a case of cholecystectomy of an 18-year-old female patient diagnosed with HS, under general anesthesia.

Keywords: Hereditary spherocytosis, cholecystectomy, general anesthesia

ÖZ

Herediter sferositoz (HS) bir tür hemolitik anemidir. Hastalığın şiddeti hastadan hastaya değişir ve HS'li her hastada splenomegaligörülür. HS çoğunlukla genç hastalarda ve çok nadiren yetişkinlerde görülür. Anestezi yönetimi bu hastalara özeldir. HS tanısı almış 18 yaşındaki kadın hastanın,genel anestezi altında kolesistektomi vakasıyla ilgili deneyimlerimizi paylaşmayı amaçladık.

Anahtar Kelimeler: Herediter sferositoz, kolesistektomi, genel anestezi

INTRODUCTION

Hereditary spherocytosis (HS) is a type of hemolytic anemia. The management of anesthesia is specific for these patients. Patients with HS are at risk for hypothermia, hypoxia, and acidosis during general anesthesia and should be monitored closely for postoperative pain. Our patient, diagnosed with HS, was taken to the operating room for cholecystectomy, and the surgery was carried out under general anesthesia. In addition to routine monitoring, we monitored the plethysmographic variability index (PVI), hemoglobin (Hb), oxygen reserve index, and oxygen content using Masimo Radical 7.

CASE PRESENTATION

We report the case of cholecystectomy in an 18-year-old female patient who was 176 cm tall and weighed 110 kg with a body mass index of 35.5. Informed consent was obtained from the patient. Her preoperative anesthesiology assessment was determined as American Society of Anesthesiology II. Her medical history revealed HS diagnosis and an ongoing prescription for 100 mg Lamictal (Lamotrigine Glaxo Smith Kline, London England) tablet for concurrent epilepsy. The patient's laboratory results showed normal values, except for the following: Hb: 14.6 g/dL, hematocrit (Htc): 46.1, mean corpuscular volume: 84.3 fL, thrombocyte count 240,000, white blood cell count: 1,040, fasting blood glucose level: 94 mg/dL, total bilirubin: 0.42 mg/dL, INR: 1.04, PT: 9.8, and APTT: 25.1. Ultrasonography (USG) showed the liver and spleen sizes of 164 mm and 156 mm, respectively and demonstrated an abundance of calculi in the gallbladder with the largest one around 13 mm in size. Peripheral blood smear confirmed the presence of spherocytes with increased osmotic fragility. The patient underwent a preoperative 8-hour fasting period. Before anesthesia, we monitored pulse, blood pressure, oxygen saturation, perfusion index (PI), PVI, HB, oxygen reserve index, and oxygen content SPOC using the probe of a Masimo Radical 7 (Masimo Corp. Irvine, CA, USA). We sedated the patient with 2 mg midazolam with preoxygenation for 3 minutes. Induction was performed with 1 mg/kg lidocaine (Osel Pharmaceutical Industry and Trade Inc., Istanbul, Turkey), 1 mg/kg fentanyl (Vem Pharmaceutical Industry, Ankara, Turkey), 4 mg/kg thiopental sodium (Ulugay Pharmaceutical Industry, Istanbul, Turkey), and 0.6 mg/kg rocuronium (Mustafa Nevzat Pharmaceutical Industry, Istanbul, Turkey) for

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muscle relaxation. Anesthesia was maintained with 2% sevoflurane + 50% air + 50% O_2. In addition to temperature monitoring, we used an electric blanket under the patient and fan heater on her arm and chest to prevent hypothermia. Furthermore, we preheated the intravenous (IV) liquids for infusion, and 50 mg IV Dolantin and 50 mg IV Contramal were administered for analgesia. Throughout the procedure, her hemodynamic parameters were stable.

We performed extubation after administering 200 mg IV sugammadex (Patheon Manufacturing Services LLC Greenville, North Carolina, ABD). We followed up on the patient in the recovery room. Furthermore, we prescribed 75 mg diclofenac sodium twice a day as postoperative analgesia. The patient was discharged one day postsurgery.

DISCUSSION

The diagnosis and treatment of HS has changed over the last 30 years, and most of the HS research has been on understanding the structural organization of the red blood cells. Abnormal red blood cell morphology has been associated with decreased spectrin, ankyrin, band 3, and protein 4.1 levels or dysfunction. The severity of hemolytic anemia is proportional to the degree of loss of these proteins.[1,2] Observation of spherocytes in the peripheral blood smear is important for diagnosis.[3]

Increased osmotic fragility, presence of spherocytes in the peripheral blood smear, and a family history was diagnostic for HS in our patient.[4]

Immune hemolytic anemia, clostridial sepsis, hemolytic transfusion reactions, hereditary pyropoikilocytosis, severe hypophosphatemia, ABO incompatibility, dyserythropoiesis, and other hereditary hemolytic anemia leading to hypersplenism are some clinical problems that can be seen. The presence of spherocytes in the peripheral blood smear should be kept in mind for the differential diagnosis of HS.

Loss of ankyrin may be because of either an autosomal dominant or recessive genetic disorder. The recessive disorder is more symptomatic than the autosomal dominant disorder, leading to mild anemia in 25% of the patients. Moreover, lack of membrane proteins prevents other proteins from binding to the lipid bilayer. As a result, lack of vesiculation and loss of membrane occurs in the red blood cells causing them to lose elasticity and become microspherocytes. This abnormal morphology prevents the translocation of spherocytes to the sinuses in the spleen, causing them to get stuck in the spleen cord. These cells are phagocytosed by the macrophages.[5]

Such an increase in the red blood cell phagocytosis may lead to anemia, jaundice, and splenomegaly. Although the severity of these outcomes varies from patient to patient, splenomegaly occurs in every patient with HS. The extent of splenomegaly is proportional to the increase in hemolysis. Splenectomy does not correct the cytoskeletal membrane defects of HS.[6,7]

Total splenectomy exposes the patient to a life-long risk of potentially lethal infections.[8]

In our case, the patient had a spleen size of 156 mm. HS occurs mostly in young patients and very rarely in adults. Pigment stones can occur even at a young age, blocking the bile duct and causing jaundice.

USG revealed many calculi in the gallbladder, the largest being 13 mm in size; therefore, a cholecystectomy was performed.

During cholecystectomy, it is essential to avoid hypoxemia, respiratory stress, and sickling. Therefore, careful management of sedatives and opioid analgesics is strongly advised. In case of intraoperative blood loss, they should be replaced. Hypothermia may lead to vasoconstriction and circulatory stasis and should thus be prevented.[9]

Intraoperatively, we used heating fans and increased the temperature of the theater to prevent potential hypothermia of the patient. We performed preoxygenation and validated the hemodynamic stability throughout the procedure. Furthermore, during the procedure and in the recovery room, we followed the levels of hypoxia, hypothermia, and hemodynamic stability. Sufficient analgesia of the patients with HS is essential during post operative care. Therefore, we visited the patient postoperatively and confirmed analgesic relief. We continuously monitored our patient's Hb using a Massimo Radical 7, which was observed as 15.1 g/dL and Htc as 46.8. Previously, the role of PVI and PI in intravascular volume and response to fluid administration was studied on patients via mechanical ventilation.[10] Thus, we preferred to use Masimo Radical 7 monitoring in our case.

In conclusion, patients with HS should be monitored vigorously in terms of hypothermia, acidosis, and hypoxia during surgery, and the necessary precautions should be taken. Moreover, maintaining analgesia is very important for the care of these patients. New technologies developed for the continuous monitoring provide valuable information to the surgical team to modulate care and help prepare for the potential complications. We aimed to suggest the preliminary preparation in anesthesia and points to be considered during the surgery and the postoperative period in patients with HS.

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