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Case Report

Anesthetic Management Of A Glycogen Storage Disease Type 1a With Air Embolism During Liver Transplantation

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ABSTRACT

Anesthetic Management of a Glycogen Storage Disease type 1a with air embolism during liver transplantation.

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Glycogen storage disease type 1a is a rare inborn error of metabolism. It causes severe fasting intolerance and lactic acidosis due to the deficiency of glucose-6-phosphatase enzyme. Anesthetic management of these patients is very difficult due to their course with different system involvements. Our case report evaluates the anesthetic management and considerations of a glycogen storage disease type 1a patient with air embolism during liver transplantation. Although orthotopic liver transplantation is an important treatment option in these patients, anesthesia management requires a multidisciplinary approach and care should be taken in terms of perioperative complications.

Keywords: Glycogen storage disease type 1a, liver transplantation, air embolism.

INTRODUCTION

Glycogen storage disease type 1a (GSD-1a) is a rare autosomal recessive inborn error of metabolism caused by deficient activity of glucose-6-phosphatase (G6Pase). In GSD-1a, endogenous glucose production is impaired by defects in both glycogenolysis and gluconeogenesis. Clinically, patients present with severe fasting intolerance, failure to thrive, and hepatomegaly.

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Biochemically, the disorder is associated with nonketotic hypoglycemia, hyperlactacidemia, hyperuricemia, and hyperlipidemia[1].

Orthotopic liver transplantation (OLT) may be considered in patients with difficult metabolic control and hepatic, renal or immunological complications despite medical treatments. Anesthesiologic management of these patients requires special perioperative planning due to multisystemic involvement. In our case report, we present an anesthesiologic approach in a rare patient with GSD-1a who developed venous air embolism (VAE) during OLT.

CASE PRESENTATION

OLT was planned a 1-year-old, 14 kg, 72 cm boy diagnosed with GSD-1a. His physical examination, the abdomen was distended and hepatosplenomegaly was detected. He was fed with nasogastric tube continuously due to hypoglycemia attacks. Despite this, he had poor metabolic control and resistant lactic acidosis. He was under $NaHCO_3$ and allopurinol treatment.

On preoperative evaluation, his neurological examination was normal, but he had growth retardation. There was known secundum ASD and that was showed with transthoracic echocardiography. His protrombin time and activated partial trombin time levels were within normal limits. His platelet count was normal, but there was a slight elevation in liver enzymes. CHILD-score was calculated as A and PELD-score as 0. The patient was fasted for 4 hours before surgery. To prevent hypoglycemia, infusion was started with dextrose solution and close blood glucose monitoring was performed. He was premedicated with midazolam.

After induction of anesthesia with propofol and rocuronium, he was intubated with a size 4.0 endotracheal tube without complication. After induction, central venous and arterial catheters were performed. Maintenance of anesthesia was provided by infusion of remifentanyl, sevoflurane and rocuronium. At the 130th minute of the surgery, during the organ dissection, sudden hypotension and a drop in end-tidal CO_2 ($EtCO_2$) occurred after rupture of the venous vessel wall due to dilatation of the hepatic vein. VAE was considered and an attempt was made to aspirate air from the central venous catheter. After the new air supply was blocked by the surgical team, the patient was ventilated with 100 % oxygen. However, the patient was immediately placed in Trendelenburg and left lateral positions, and vasopressor support in the form of norepinephrine infusion was started. With all these interventions, hemodynamic stabilization was achieved.

Surgery continued for 9 hours. Although the total bleeding amount of the patient, who had occasional severe surgical bleeding, could not be calculated precisely, fluid replacement was performed with close arterial blood gas (ABG) monitoring and hemodynamic monitoring. A continuous infusion of dextrose solution was

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given intraoperatively and the blood glucose levels remained in the range of 74-286 g/dL. In ABG monitoring, the lowest hemoglobin value was 5,4 g/dL and the highest lactate value was 26 mmol/L. The patient was transferred to intensive care unit (ICU) postoperative as intubated with vasopressor support. At the postoperative 4th hour, the patient was extubated according to the weaning criteria following consciousness. Patient's neurological examination was normal after extubation, therefore no complication due to intraoperative VAE was suspected (Figure 1).

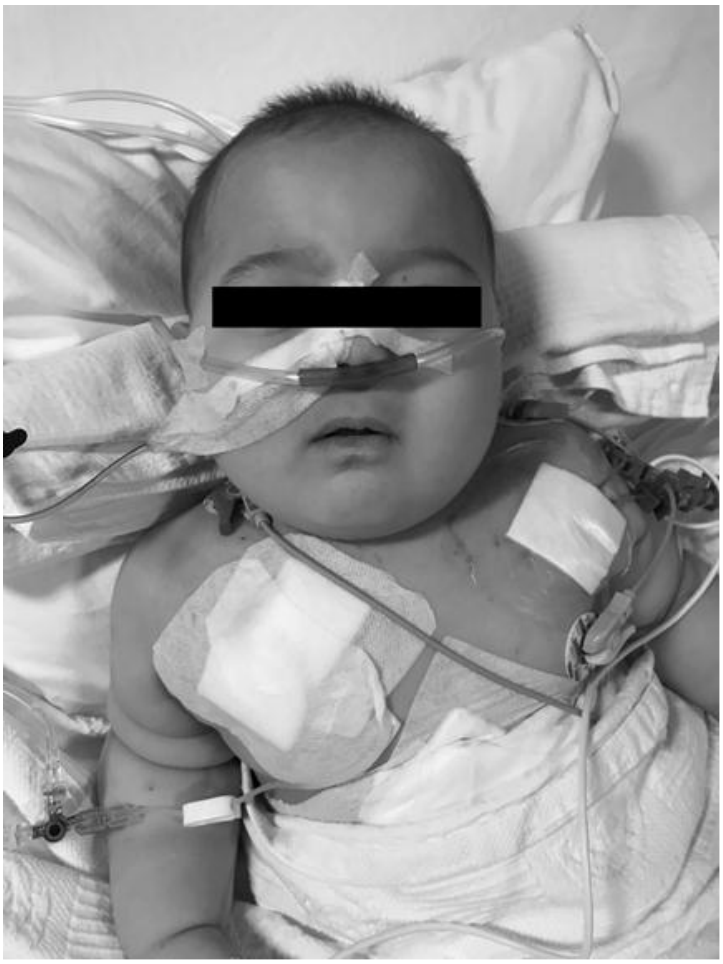

Figure 1. The patient in transplant ICU immediately after extubation.

DISCUSSION

Glycogen storage diseases occur as a result of enzymatic abnormalities that lead to abnormal concentrations or structures of glycogen. In GSD-1a, absence of G6Pase is characterized with fasting hypoglycemia. As both glucose producing pathways are blocked due to this inborn error, glycogen accumulation in liver, kidney and intestine is unavoidable. Hypoglycemia and glycogen storage are the main reasons of the clinical findings and other biochemical abnormalities of the disease[2]. The hypoglycemia is the major problem in the

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GSD-1a patients. This problem is caused by fasting before surgery. As fasting hypoglycemia is the most important problem of the disease, a short duration of preoperative fasting is recommended for such patients. We offered four hours fasting to our patient. We started infusion of dextrose solutions before induction of anesthesia and did not experience any difficulties in maintaining normoglycemia in the patient.

VAE is the condition in which air enters the systemic venous circulation from the operation area or during interventional procedures. Intraoperative VAE in OLT is an important complication that can be seen during dissection, vascular anastomoses, and liver reperfusion. A sudden decrease in $EtCO_2$ value in anesthetized patients may appear as the earliest sign of air embolism. Hypoxemia, hypercarbia, hypotension, tachyarrhythmias and ST-segment changes were seen[3]. In our case, almost all of these findings were seen. To avoid VAE during surgical procedures, the patient should be positioned so that the surgical area is above the right atrium level, adequate hydration is needed to keep the central venous pressure between 10 and 15 cmH_2O[4]. After the diagnosis of VAE, early intervention is very important. First of all, new air entry should be prevented by the surgical team[5]. It should be ensured that the patient is breathing with 100 % oxygen. It may be possible to relieve the airlock on the right side of the heart by placing the patient in the partial left lateral decubitus (Durant maneuver)[6] and Trendelenburg position. If the patient has a central venous catheter, air aspiration from the right atrium should be attempted although the success rates are reported to be low[5]. Appropriate inotrope and vasopressor support should be provided to ensure hemodynamic stabilization. CPR algorithms should be initiated in patients with cardiac arrest. In our case, we intervened in our patient by following this intervention algorithm. No complications were observed due to VAE in our patient in the postoperative period.

Bleeding disorders due to hepatic dysfunction and airway difficulties due to growth retardation or macroglossia may also be encountered during surgery in patients with GSD-1a. We did not experience any problems in our patient due to these reasons.

In conclusion, GSD-1a is a rare disease that can cause serious multisystemic problems; therefore anesthesia management of these patients requires a multidisciplinary approach. Although the frequency of air embolism in OLT surgery is not very high, intraoperative close follow-up and early intervention is important to avoid long term complications.

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KE: conceptualisation, data collection, paper draft, literature review, manuscript preparation and is the lead author. ÇN: paper draft, literature review, TA: paper draft, literature review,. HM: paper draft, literature review. All authors approved the manuscript.

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