

Anaesthetic Management of a Patient with Glycogen Storage Disease Type 1A

 Esra ÖZER^{a,b}

^aDepartment of Oral and Maxillofacial Surgery,

^bDivision of Anesthesiology and Reanimation, Gazi University Faculty of Dentistry, Ankara, TURKEY

Received: 31.03.2018

Received in revised form: 24.04.2018

Accepted: 25.04.2018

Available online: 05.12.2018

Correspondence:

Esra ÖZER

Gazi University Faculty of Dentistry, Department of Oral and Maxillofacial Surgery,

Division of Anesthesiology and Reanimation,

Ankara, TURKEY

tacesra@yahoo.com

ABSTRACT Glycogen storage diseases (GSD) are a group of inherited disorders which result in deficiencies of enzymes involved in glycogen metabolism and lead to an accumulation of glycogen in the liver. Glycogen storage disease type 1a, a form of glycogen storage disturbance, is a rare metabolic disorder with important implications for anaesthesiologists. It is caused by the lack of the glucose-6-phosphatase, which is necessary for the liver to convert glycogen to glucose. The aim of this case report is to present an uncomplicated surgery with an uncomplicated general anaesthesia and an anaesthetic follow up in a patient with GSD type 1a in an elective day case surgery. In this case, there was a serious hypoglycemia problem due to the glycogen storage disturbance related to disease. To avoid severe hypoglycemia, it is crucial to keep oral feeding 2-3 hour for maintaining a normal blood sugar level.

Keywords: Glycogen storage diseases (GSD); hypoglycemia; anaesthesia, general

The glycogen-storage diseases (GSD) are a group of inherited disorders which result in deficiencies of enzymes involved in glycogen metabolism and lead to an accumulation of glycogen in the liver. GSD type 1a is a condition in which there is an abnormal deposit of glycogen and a hepatorenal deficiency of glucose-6-phosphatase. GSD type 1a is a multisystem disorder with significant morbidity and mortality. The clinical features of the disease include hypoglycemia, lactic acidosis, hyperuricemia, hyperlipidemia, hyperproteinemia, hepatomegaly, truncal obesity, rounded "doll-like" face, growth retardation, muscle wasting and bleeding tendency (Figure 1).¹⁻³ Previous reports of anaesthesia in patients with this condition are limited, but indicate that serious complications may arise.

More recently Huang detailed two cases in which the clinical course appeared satisfactory, but in both of them blood samples showed the presence of moderate and severe hypoglycemia during surgery.⁴

From the anaesthetist's point of view; hypoglycemia and hepatic dysfunction are the major problems to deal with GSD type 1a patients at the perioperative period.²⁻⁴ Moreover, difficulties in intubation and ventilation and aspiration risks are the other encountered problems in GSD patients during surgery.^{2,3,5}

The application of modern methods of management can correct or improve the clinical and biochemical features of GSD type 1a disease. If peri-



FIGURE 1: Rounded "doll-like" face in Glycogen Type 1a storage disease.

ods of hypoglycemia and metabolic acidosis can be avoided then these patients appear to tolerate anaesthesia and surgery satisfactorily.

The aim of this case report is to present an uncomplicated surgery with an uncomplicated general anaesthesia and an anaesthetic follow up in a patient with GSD type 1a in an elective day case surgery.

CASE REPORT

Tonsillectomy, adenoidectomy and circumcision were planned in a 2-yr-old boy with GSD-1a. He was diagnosed in the infancy period after a liver biopsy and an enzyme assay. His physical examination, the abdomen was distended and a remarkable hepatosplenomegaly was detected. His preoperative evaluation revealed normal platelet count and liver enzymes. His prothrombin time (PT) and activated partial thrombin time (PTT) levels were within normal limits (PT:11,6 sec [12-18 sec], aPTT: 28,8 sec [25-40 sec], INR:0.87% [0.80-1.22]). Blood count revealed leukopaenia (4620 leukocytes per mm³). The patient was consulted to pediatrics and close follow up for hypoglycemia during and after surgery was advised.

The patient was fasted for 4 hr preoperatively. His preoperative blood sample was drawn for the

assessment of blood glucose level after the insertion of an intravenous cannula and was found 59 gr.dl⁻¹ (70-100 gr.dL⁻¹). The infusion of 10% dextrose with 0.9% NaCl was started thereafter. His blood glucose was measured before premedication. He was premedicated with midazolam (Dormicum, Roche) 0.5 mg (IV) after blood glucose reached 91 gr.dL⁻¹. Anaesthesia was induced with 3 mg.kg⁻¹ propofol (Propofol, Abbott) and muscle relaxation was achieved with rocuronium bromide (Esmeron, MSD) 8 mg. Orotracheal intubation was easily accomplished using a 4.5 mm ID endotracheal tube, and the lungs were mechanically ventilated. N₂O/O₂ combination and sevoflurane 2% was used for maintenance. Oxygenation, ventilation, and hemodynamic status were normal and stable. Venous blood was taken for gas analysis and for plasma glucose concentration measurements. Except for severe hypoglycemia (plasma glucose concentration of 65 mg/ dl⁻¹) at the beginning of the procedure, which was corrected by intravenous administration of 10% dextrose, there were no episodes of metabolic acidosis. At the end of the surgery, volatile agents were discontinued and the patient was extubated. Recovery from anaesthesia was uneventful. At the postoperative period his venous blood sample was taken and his blood glucose and lactate levels were found 99 gr dL⁻¹, and 13 mg.dl⁻¹ (4,5-14,4 mg.dl⁻¹) respectively. The patient was infused with 10% dextrose and 0.9% NaCl for 4 hours and fed at the 2nd postoperative hour. He was discharged at the 12th postoperative hour without any other problems.

DISCUSSION

Glycogen storage diseases occur as a result of enzymatic abnormalities that lead to abnormal concentrations or structures of glycogen. In type 1a (GSD-1a), absence of glycogen-6-phosphatase 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.¹⁻³

The hypoglycemia is the major problem in the 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. Our patient experienced hypoglycemia, although we offered four hours fasting to our patient. The patient hospitalized and preoperative dextrose infusion started for the adequate control of blood glucose concentrations. Surgery is a major stress to the body and results in glucose accumulation into the blood due to do release of counter regulatory hormones in patients undergoing surgery. In regard to GSD-1a patients, the surgical stress results in glucose-6-phosphate accumulation in blood. The accumulation of this intermediate metabolite causes hypoglycemia in acute terms. The perioperative hypoglycemia can only be prevented by glucose infusion. We started infusion of dextrose solution 4 hours before induction of anesthesia. The overall stay time of the patient in the operating theatre was about 1 hour. We did not experience lactic acidosis during this period. At the

postoperative period, as fasting lasted, we continued dextrose infusions for four hours. The patient was fed at the postoperative 2nd hour. We discontinued the dextrose infusion as soon as he tolerated his oral diet.

Source of Finance

During this study, no financial or spiritual support was received neither from any pharmaceutical company that has a direct connection with the research subject, nor from a company that provides or produces medical instruments and materials which may negatively affect the evaluation process of this study.

Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

This study is entirely author's own work and no other author contribution.

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