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Case Report

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Anesthetic Management in a Patient With Glucose-6-Phosphate Dehydrogenase Deficiency Undergoing Coblation Adenoidectomy With Septoplasty and Turbinectomy

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Abstract

Glucose-6-phosphate dehydrogenase (G-6-PD) is the major enzyme in the pentose phosphate pathway (PPP). The end products of this pathway are ribose-5-phosphate and nicotinamide adenine dinucleotide phosphate hydrogen (NADPH). G-6-PD deficiency is known to be the most common enzymatic deficiency in red blood cells (RBCs). Genetically, the mode of inheritance is an X-linked recessive disease. The exposure to oxidative stressors will result in hemolytic anemia including fava beans, infections, metabolic conditions such as diabetic ketoacidosis, metabolic acidosis, hyperglycemia, hypoglycemia, and hypothermia. Moreover, surgical stress and certain types of medication are known to lead to hemolytic anemia. Acute hemolytic crisis is a life-threatening situation in patients with G-6-PD deficiency. Therefore, it is extremely important to monitor the patient perioperatively. The authors present this case of successful anesthetic management in a 23-year-old lady with G-6-PD deficiency and a previous history of acute hemolytic anemia undergoing coblation adenoidectomy with septoplasty and turbinectomy.

Keywords: Glucose-6-phosphate dehydrogenase; Deficiency; Hemolytic crisis; Medications; Hemolysis

Introduction

Glucose-6-phosphate dehydrogenase (G-6-PD) deficiency is an enzymopathy that leads to red blood cells (RBCs) hemolysis [1, 2]. It is the most common enzymopathic disorder of RBCs [3, 4]. It is a genetic disorder localized on the X chromosome which is inherited by an X-linked recessive pattern. Thus, it is transmitted and occurs primarily in males. Worldwide, it has been reported that more than 400 million people have been diagnosed with this disease [3-5]. The rate of new cases is associated with areas endemic for malaria. However, it is conventional in other countries [6]. A huge body of evidence considered that the G-6-PD is the most important factor in the production of antioxidants, which protecting the cells from oxidative damage [6]. Accordingly, patients with G-6-PD deficiency are highly susceptible to oxidative stressors [6]. In the previous reported cases, successful anesthetic management of a patient with G-6-PD deficiency involved the decreasing of possible oxidative stressors, and this is dependent on the type of surgery [5-7]. In our report, we present this case of successful anesthetic management in a 23-year-old female patient with G-6-PD deficiency undergoing coblation adenoidectomy with septoplasty and turbinectomy. We also discuss the different types of medication to be used in such a patient based on reviewing the literature and our experience.

Case Report

A 23-year-old female patient (height: 150 cm, weight: 48 kg) was diagnosed as G-6-PD deficiency at the age of 7 years after a hemolytic episode, and she received a blood transfusion at that time; no other episodes were reported. She presented to the Otolaryngology-Head and Neck Surgery Clinic with symptoms of nasal obstruction, nasal tone of voice, and mouth breathing. A routine laboratory investigation was revealed: hemoglobin of 12.3 g/dL, white blood cell counts of 4.63×10^9/L, RBC counts of 4.09×10^{12}/L, and platelet of 158×10^9/L; all are within normal limits. Computerized tomography (CT) of paranasal sinuses showed deviated nasal septum, minimal mucosal thickening seen in the maxillary sinuses, left upper molar radicular cyst, and asymmetrical thickening of the nasopharyngeal mucosa.

The planned surgery was for coblation adenoidectomy with septoplasty and turbinectomy. Intraoperatively, a 20-gauge intravenous cannula was established, and the patient was moni-

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tored by electrocardiogram (ECG), bispectral index sensor (BIS) and the measurement of oxygen saturation (SpO_2), non-invasive blood pressure (NIBP), temperature, and followed by preoxygenation. Pre-induction vital signs revealed blood pressure of 118/81 mm Hg, heart rate of 82 beats per minute (bpm), SpO_2 of 100%, and a temperature of 36.4 °C. Thereafter, general anesthesia induced with fentanyl (100 µg), propofol (150 mg), rocuronium (30 mg), and dexamethasone (16 mg). The patient was intubated by a direct laryngoscope with oral endotracheal Ring-Adair-Elwyn (RAE) tube (internal diameter (ID): 6.5 mm) with an inflatable cuff and connected to the circuit of the anesthesia machine. Anesthesia has been maintained by the inhalational anesthetic agent sevoflurane (2.0%) with O_2/N_2O mixture (1.0/1.0 L/min). The patient was positioned supine for the surgery. Before the incision was made, cefazolin (2 g) was given as a prophylactic antibiotic. At 40 min into the surgery, we administered paracetamol (1 g) followed by oxycodone (4 mg). Also, she received isotonic, crystalloid fluid (Ringer's lactate) with a total infusion of 650 mL. The estimated blood loss was 50 mL. Intraoperatively, the patient was vitally stable. The duration of the surgery was approximately 90 min. At the end of the surgery, we reversed the residual neuromuscular medication with glycopyrrolate (0.4 mg) plus neostigmine (2.5 mg), and when the patient was fully awake, the trachea was extubated successfully. Postoperatively, oxycodone has been used as a postoperative analgesia. Moreover, we monitored the patient for any signs or symptoms of an acute hemolytic crisis. The patient was discharged on the second postoperative day (POD) after assuring the result of complete blood count (CBC) is normal.

Discussion

In the pentose phosphate pathway (PPP) which known also as phosphogluconate pathway and hexose monophosphate (HMP) shunt, the major enzyme is G-6-PD [5, 6]. It is the first step in PPP resulting in converting glucose to ribose-5-phosphate. In our body, the only source for generating the nicotinamide adenine dinucleotide phosphate hydrogen (NADPH) is PPP [6]. In contrast, the generation of NADPH will lead to reducing cell energy by controlling the reduced glutathione within the cell [2]. Oxidative damage will be bypassed by glutathione reduction as this mechanism results in cell protection [2]. RBCs do not have another metabolic backup system as other human cells, which work to generate the intracellular NADPH [2, 8]. Therefore, any exposure to oxidative stress will contribute to hemolytic anemia [2, 8]. Subsequently, G-6-PD deficiency is lethal in RBCs [2, 8]. However, RBCs are vulnerable to oxidative stress, leading to hemolytic anemia [6].

Hemolysis can be happening after exposure to many factors such as fava beans, infections, metabolic conditions such as diabetic ketoacidosis, metabolic acidosis, hyperglycemia, hypoglycemia and hypothermia, and have been all over reported previously [1, 9-11]. Furthermore, certain types of medication such as antimicrobials (sulfonamides, nitrofurantoin, and chloramphenicol), non-steroidal anti-inflammatory drugs (NSAIDs), anticonvulsants, diuretics containing sulfonamide, insulin, oral hypoglycemic agents, and ranitidine, are known to lead to hemolytic anemia [5, 6, 9-11]. The mechanism of hemolytic anemia is due to membrane damage by oxidized hemoglobin [5, 6]. At the same time, anesthetic drugs such as diclofenac, metoclopramide, lidocaine, methylene blue, and prilocaine are contraindicated in patients with G-6-PD deficiency [11, 12]. While glycopyrrolate, fentanyl, sufentanil, tramadol, ketamine, propofol, thiopental, halothane, nitrous oxide, rocuronium, succinylcholine, neostigmine, bupivacaine, and heparin are reported previously as it can be administrated safely in patients with G-6-PD deficiency [1, 4, 6, 11]. However, there are anesthetic agents still controversy such as sevoflurane and midazolam [6]. In our case, we administrated the inhalational agent sevoflurane, and to our experience it is safe in patients with G-6-PD deficiency. Overall, there is insufficient evidence of medications to be used in G-6-PD deficiency patients, and some of them are still controversy [6]. Hence, for patients with G-6-PD deficiency, perioperative medications must be known as much as possible to avoid acute hemolytic crisis. All the administrated medications in our case were safe for a patient with G-6-PD deficiency.

Also, hemolysis can be caused by surgical stress, which has been reported in the literature [5]. Therefore, perioperative adequate analgesia will work effectively to decrease the level of stress related to the surgery [5]. We provided adequate analgesia throughout the intraoperative and postoperative interval because this surgery is painful. In such a case, our aim is to avoid acute hemolytic crisis [5]. However, acute hemolytic crisis is self-limiting and in severe rare cases granting blood transfusion [6]. After 24 to 72 h, the hemolysis can become demonstrable in patients with risk factors [2, 6]. Intraoperatively, reducing oxidative stress is extremely important besides monitoring and treating hemolysis [6]. During surgery, hematuria is an indicator of an acute hemolytic crisis [5]. Reaching to the diagnosis is substantial, and once that made, we have to cease the triggering agent and maintaining the urine output by infusing crystalloids as well as administration of non-sulfonamide diuretics [5, 13]. Unfortunately, a Foley catheter was not inserted in our case because the surgical duration was short; but we recommend inserting a Foley catheter in such a patient to monitor the urine output regardless of the surgical duration. Postoperatively, the symptoms of hemolysis will be appearing clearly by 7 days after the hemolysis, which include headache, fatigue, tachycardia, cyanosis, dyspnea, lethargy, lumbar pain, abdominal pain, splenomegaly, hemoglobinuria and scleral icterus [3, 6]. To monitor the need for blood transfusion, CBC must be done daily after an episode of acute hemolytic crisis [5]. Microscopic examination of the peripheral blood smear will show RBC fragments such as reticulocytes, schistocytes, and Heinz bodies. Other lab investigations that can be done are lactate dehydrogenase and unconjugated bilirubin which will be increased, whereas haptoglobin levels will decrease [2, 6]. Additionally, urobilinogen and hemosiderin can be detected in the urine [2, 6]. Also, we can use the direct Coombs test to rule out an immunological reaction [2, 6].

Conclusions

From an anesthetic point of view, acute hemolytic crisis is correlated with different triggers as we previously mentioned. Out

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of these triggers, there are particular types of medication we have to avoid as much as possible in patients with G-6-PD deficiency. Moreover, monitoring the patient perioperatively for such a crisis is extremely important, which includes surgical stress, because acute hemolytic crisis is a life-threatening situation in these patients.

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Conflict of Interest

None to declare.

Informed Consent

Obtained.

Author Contributions

Abdulrhman Saleh Dairi is the first author involved in the literature review and preparation of the manuscript (initial and revised versions). Moheyeldin A. Farghaly and Ghida Dairi are involved in the critical revision of all versions of the manuscript.

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