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Case Report

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ANAESTHETIC MANAGEMENT OF A PATIENT WITH G-6-PD DEFICIENCY: AN OUNCE OF PREVENTION IS WORTH A POUND OF CURE

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ABSTRACT

Glucose-6-Phosphate dehydrogenase (G-6-PD) deficiency is a clinical condition characterized by red blood cell enzymatic defect resulting in hemolysis following exposure to oxidative stress from medications and infections.[1] The management of a patient with G-6-PD deficiency, involves careful preoperative assessment, including a thorough work-up, optimisation of haematocrit, and being prepared for transfusion of blood. Herein we discuss a case of 6 year old male child diagnosed case of G-6-PD deficiency with septic arthritis managed successfully with arthrotomy under subarachnoid block. The focus of anesthetic management in this child was avoiding drugs which may cause hemolysis in G-6-PD deficiency, reducing surgical stress with adequate analgesia, and adequate preparations to manage hemolysis should it occur.

KEYWORDS: Glucose-6-Phosphate dehydrogenase deficiency, Oxidative stress, Hemolysis.

INTRODUCTION

Glucose-6-phosphate dehydrogenase (G-6-PD) deficiency is an X-linked disorder affecting red blood cells (RBCs) and is one of the most prevalent enzyme deficiencies.[2] G-6-PD is a key enzyme in the rate limiting step of the Pentose Phosphate Pathway (PPP), in which glucose is converted to a 5-carbon form ribose-5-phosphate. As this takes place nicotinamide adenine dinucleotide phosphate (NADP+) is converted to its reduced form NADPH. This then becomes the reducing power of the cell, by maintaining Glutathione in its reduced form. Reduced Glutathione has potent anti-oxidant activity, and protects the cell from free radical and oxidative damage.[3] Whilst other cells have multiple metabolic pathways to generate the reductive agent NADPH, for red blood cells the PPP is the only modus.[3] Hence when disturbed, the red cells are relatively unprotected, and are prone to hemolysis when challenged with oxidative stress.

Here we report a case of 6 year old male child, diagnosed case of G-6-PD deficiency with septic arthritis managed successfully with arthrotomy under subarachnoid block. We also discuss the different types of medication to be used in such a patient based on review of literature.

Case presentation

A 6 year old male child weighing 20 kg presented to emergency department with complaint of fever for 3 days, swelling right knee with difficulty in walking for 2 days. Fever was insidious in onset, documented as 102^0 F-103^0 F which got relieved after taking medication. The swelling was also insidious in onset and associated with difficulty in walking. Patient has no history of trauma. X ray right knee joint showed joint effusion and ultrasound showed inflammatory changes in knee joint. Patient was taken for arthrotomy of the knee joint. The patient was a known case of G-6-PD deficiency, diagnosed since his neonatal period. He was delivered by lower segment caesarean section at POG 38 weeks 2 days with a birth weight of 2.6 kg. He had history of neonatal jaundice and received phototherapy. After discharge at the age of 22 days, the mother was counseled to avoid trigger factors for G6PD deficiency hemolysis. After routine investigations and pre anaesthetic check up patient was taken for surgery. The anaesthetic technique was discussed with parents highlighting the risks of general anaesthesia compared to regional anaesthesia. An informed consent for surgery and anaesthesia was obtained from the parents. Perioperative management should focus on avoiding the triggers including pain, anxiety and drugs resulted in hemolysis.

Following pre-anaesthetic machine checklist, the child was positioned appropriately and standard monitoring applied using the multiparameter monitor, the initial vital signs were recorded as: heart rate 92 beats/minute, arterial oxygen saturation (SpO2)- 99% on room air, respiratory rate – 20 breaths/minute and normal electrocardiographic measurements. Using a 22- gauge

cannula intravenous fluid started, child was pre-medicated with intravenous glycopyrrolate 0.1mg. A sleeping dose of ketamine (1mg/kg) was administered intravenously and 100% Oxygen was administered at the rate of 4Litre/minute via a nasal cannula. Following an aseptic protocol, the subarachnoid block was given after free flow of cerebrospinal fluid using 0.4mg/kg body weight of 0.5% hyperbaric bupivacaine at L4/L5 inter-spinal space, using 26-Gauge Quincke spinal needle in lateral position. The child was gently re-positioned supine, in horizontal position and vital signs were monitored closely. The level of sensory block was assessed to be T10 before incision. Oxygen inhalation @ 4L/min via nasal cannula maintained throughout surgery. The air-conditioning system in the operating room was switched off and exposed parts of patient were covered with warm gauze padding. Hydration was done peri-operatively with intravenous fluids. The total amount of crystalloid administered was 650 ml while the estimated blood loss was less than 25mls. The child was kept warm and transferred to the postoperative anaesthesia recovery unit without pain or other complains, for continuous monitoring of vital signs and oxygen supplementation. He was kept there for one hour. Pain management was done with inj paracetamol 300 mg 6 hourly during postoperative period in ward. The child was orally allowed after 6 hours of surgery. He was discharged following day without any complications. The parents of the child were taught to look out for signs and symptoms of hemolysis.

DISCUSSION

G-6-PD deficiency is the most commonly occurring enzyme deficit, affecting approximately 400 million people worldwide.[1] The prevalence in India is 3%.[2] The geographical distribution correlates with areas endemic to Malaria, as G-6-PD deficiency imparts increased resistance to plasmodium falciparum, which cannot tolerate the increased levels of oxidative stress.[2]

During surgery, anesthetic management should focus on minimizing oxidative stress, monitoring and treating hemolysis. The main causes of oxidative stress are the consumption of fava beans, certain drugs, infections and metabolic conditions such as diabetic ketoacidosis metabolic acidosis, hyperglycemia, hypoglycemia and hypothermia.[1,4] 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.[6] The mechanism of hemolytic anemia is due to membrane damage by oxidized hemoglobin. At the same time, anesthetic drugs such as diclofenac, metoclopramide, lidocaine, methylene blue, and prilocaine are contraindicated in patients with G-6-PD deficiency.[7] 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.[7] However, there are anesthetic agents still in controversy such as sevoflurane and midazolam.[7] Hence, for patients with G-6-PD deficiency, perioperative medications use should be minimized to avoid acute hemolytic crisis.

As anaesthesiologists, we always need to focus on anaesthetic methods and anaesthetics when we meet a patient with favism. Local infiltration anaesthesia, peripheral nerve block and spinal anaesthesia are much safer options in these patients.

Sub arachnoid block (SAB) with heavy bupivacaine was the preferred anaesthetic technique in the child discussed in present case report because the procedure was carried out below the umbilical level. It also allowed for minimal use of drugs (0.5% hyperbaric bupivacaine) unlike in general anaesthesia, where poly-pharmacy may be required to achieve effective anaesthesia. Hemolysis can also be caused by surgical stress, which has been reported in the literature.[8] Therefore, perioperative adequate analgesia and optimum temperature worked effectively to decrease the level of stress related to the surgery. We provided adequate analgesia throughout the intraoperative and postoperative interval.[8] Patient was discharged without complications on the following day.

Table 1: List of safe and unsafe drugs in Glucose-6-phosphate deficiency.

Unsafe for class I,II,III	Safe for class II and III
Acetanilid	Acetaminophen
Dapsone	Aminopyrine
Methylene blue	Ascorbic acid
Nalidixic acid	Aspirin
Nitrofurantoin	Chloremphenicol
Niridazole	chloroquine
Primaquine	Colchicine
Toulidine Blue	Diphenhydramine
Vitamin K	isoniazide
Sulfacetamide	L-Dopa
sulfamethoxazole	probenecid
Furazolidone	procainamide
Isoflurane	Pyrimethamine
sevoflurane	Quinidine
Diazepam	Paraminobenzoic acid
phenazoyridine	Phenacetin
trinitrotoulene	Quinine
thiazosulfone	sulfamethoxypyridazine
	streptomycin
	sulfisoxazole
	trimrthoprim

CONCLUSION

G-6-PD Deficiency is a rare disease that presents as a spectrum, and inattentive management is associated with largely preventable complications that significantly worsen post up outcome, hence a high degree of

vigilance is required when these patients require the administration of anesthesia. An optimal pre-operative workup, and meticulous planning of anesthetic medications, along with a detailed discussion with the patient and family will ensure that the best possible care is given.

CONFLICT OF INTEREST

All authors declare they have no conflict of interest

FINANCIAL DISCLOSURE

Nil

REFERENCES

1. Cappellini MD, Fiorelli G. Glucose-6-phosphate dehydrogenase deficiency. Lancet, 2008; 371(9606): 64-74.
2. Panich V. Glucose-6-phosphate dehydrogenase deficiency. Part 2. Tropical Asia.
3. Petz LD, Garratty G. 2nd ed. Philadelphia: Churchill Livingstone; Immune haemolytic anemias, 2004; 261–317.
4. Carvalho CG, Castro SM, Santin AP, Zaleski C, Carvalho FG, Giugliani R. Glucose-6-phosphate-dehydrogenase deficiency and its correlation with other risk factors in jaundiced newborns in Southern Brazil. Asian Pac J Trop Biomed, 2011; 1(2): 110-3.
5. Beutler E. Glucose- 6 -phosphate dehydrogenase deficiency: a historical perspective. Blood, 2008; 111(1): 16-24.
6. Hou L, Lin S, Meng Z, Zhang L, Liu Z, Luo X, Liang L. Young type 1 diabetes mellitus (T1DM) patient with glucose-6-phosphate dehydrogenase deficiency occurring hemolysis: A case report. Biomedical Research, 2017; 28(16): 6994-6.
7. Sharma V, Verma S, Bhatia PK, Sethi P. Propofol infusion in an infant with glucose-6-phosphate dehydrogenase deficiency. J Anaesthesiol Clin Pharmacol, 2018; 34(1): 136-7.
8. Valiaveedan S, Mahajan C, Rath GP, Bindra A, Marda MK. Anaesthetic management in patients with glucose-6-phosphate dehydrogenase deficiency undergoing neurosurgical procedures. Indian J Anaesth, 2011; 55(1): 68-70.