

Perioperative management in a case of glucose-6-phosphate dehydrogenase deficiency undergoing orthopaedic surgery

Sir,

We report a case of anaesthetic management in a glucose-6-phosphate dehydrogenase (G6PD) deficient individual undergoing orthopaedic surgery. Such a case poses an anaesthetic challenge during perioperative period due to limited choice of antibiotics and analgesics, predisposition to hemolysis with local anaesthetic lignocaine. Our case had additional issues of cerebral palsy and hypocalcaemia so general anaesthesia (GA) was administered.

An 11-year-old male child weighing 19 kg with G6PD deficiency and spastic cerebral palsy following neonatal hyperbilirubinemia (kernicterus) was posted for fracture femur surgery. Child had haemolytic jaundice due to chest infection at 4th year of life. Dye decolorization (qualitative test) for G6PD revealed complete deficiency. Haemogram, routine biochemical investigations and electrocardiography were normal. Serum calcium was low (5.1 mg/dl). Hypocalcaemia work up revealed low levels of ionized calcium (0.6 mmol/L), phosphorus (2.3 mg/dl) and Vitamin D3 (22 mg/ml) and elevated alkaline phosphatase level (862 IU/L). Calcium and Vitamin D3 supplements were given preoperatively. Premedication with injection cefoperazone and sulbactam 1 g, calcium gluconate 500 mg intravenously and salbutamol nebulisation was given. Standard endotracheal anaesthesia was administered with appropriate intravenous doses of glycopyrrolate, midazolam, fentanyl, propofol and atracurium and maintained with oxygen, air, isoflurane. Active warming measures were used to maintain temperature. Postoperative analgesia was given with injection paracetamol and tramadol. Daily haemogram was done postsurgery for early

detection of hemolysis. 3rd day child developed pneumonia requiring antibiotic therapy.

Our case needed special attention due to cerebral palsy and hypocalcaemia coexisting with G6PD deficiency; so GA was administered. Various drugs used for anaesthesia and analgesia, antibiotics, surgical stress, infection, conditions associated with generation of free radicals like perioperative ischemia, reperfusion, hypothermia and acidosis are known to cause oxidative stress and thereby hemolysis in G6PD deficient patient. [1] Peripheral limb ischemia and reperfusion is commonly encountered situation in orthopaedic surgery with tourniquet use. Young age, male gender, negative family history of G6PD deficiency, presence of fever and vomiting and high alkaline phosphatase are the predictors for severe hemolysis [2] as in this case. Laboratory findings following a hemolytic episode are anaemia, decreased serum haptoglobin and an elevated indirect bilirubin. Benzodiazepine, propofol, ketamine, fentanyl, codeine, paracetamol, inhalational agent namely isoflurane and sevoflurane are safe for perioperative use, [1,3] α_2 receptor agonist, dexmedetomidine have also been used successfully for maintenance of anaesthesia. [4] Very few case reports mentions the use of local anaesthetic, bupivacaine [5] for regional anaesthesia but lignocaine has been reported to cause hemolysis in G6PD deficiency. Epidural analgesia was the superior option in our case, but could not be implemented in view of very limited data on safety of local anaesthetics and predilection of severe hemolysis. This underlines the need of global consensus for use of local anaesthetics particularly in orthopaedic procedures. Perioperative concerns with cerebral palsy were difficulty in obtaining cooperation,

positioning, hypothermia, hypotension, seizure and muscle spasm. Epileptogenic drugs such as ketamine, etomidate, sevoflurane and enflurane were avoided in our case. Hypocalcaemia in our child was secondary to inadequate dietary intake and sun exposure. Perioperative concerns were depressed myocardial contractility, prolonged QT interval, heart block, convulsions, and laryngeal spasm. Maintenance of normocapnia helped to prevent further decrease in ionized calcium levels in our case.

Anesthetic challenges in the perioperative period in our case were prevention and monitoring of hemolysis, infections necessitating antibiotics and limited choice of analgesic drugs as it was in our case. "Do regional anesthesia and local anesthetic drugs score better than GA?" it's important to know, as they are of great clinical interest in orthopaedic procedures.

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