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Hip replacement in a patient of sickle cell disease: an anaesthetic challenge

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Abstract

Majority of surgical procedures in patients with sickle cell disease are a result of the disease process itself and require intervention for either treatment or prevention of certain complications. Avascular necrosis of femoral head is a known complication of sickle cell disease. Hereby we describe the successful anaesthetic management of a 45 year old male patient with sickle cell disease with avascular necrosis of femoral head who underwent total hip replacement surgery under central neuraxial blockade. Proper understanding of the disease process and its anaesthetic implications are essential for successful perioperative management of sickle cell disease patient.

Keywords: Avascular necrosis of femoral head, Sickle cell disease, Central neuraxial blockade

Introduction

Sickle cell disease (SCD) is a group of autosomal recessive disorders of the β-haemoglobin chain. It is characterised by chronic haemolytic anaemia, intermittent painful vaso-occlusive episodes, pulmonary infections and multi-organ damage. A single amino acid substitution, valine for glutamate, at position 6 on the β chain is responsible for the condition. The single amino acid substitution creates an allosteric abnormality of the haemoglobin molecule rendering it unstable as well as less soluble when deoxygenated. The unstable structure of haemoglobin causes accelerated breakdown and haemolytic anaemia while the reduced solubility of haemoglobin permits haemoglobin tetramers to form polymers forming long helical bands causing distorted red cells and hence the name sickle cell disease.

Avascular necrosis of femoral head is a known complication of sickle cell disease following vaso-occlusive event. About 7% of all deaths among patients with sickle cell anaemia are related to surgery.(1) Perioperative management in these patients focuses on prevention of vaso-occlusive event which may get precipitated by hypoxia, hypothermia, hypotension, hyperthermia, hypovolemia and acidosis.(3)

Case Report

45 year old, averagely built male patient, presented with complain of excruciating pain and limited range of movements of right hip. On radiological examination, patient was diagnosed with avascular necrosis of femoral head and hence posted for total hip replacement (THR). Patient was found to be anaemic (Hb-7.5gm%), and work up for cause of anaemia resulted in patient being diagnosed as a case of sickle cell disease. Patient did not give any recent or previous history suggestive of respiratory infection or complications, chest pain or renal symptoms. His preliminary investigations were as follows: Hb-7.5 gm%: HbA-12.6%, HbA2-5.5%, HbF-13.1%, HbS-68.8%. Chest x-ray showed fibrosis of left lower zone suggestive of old pulmonary pathology (probably infarct). Renal and liver function test, electrocardiogram, coagulation profile, blood sugar and arterial blood gas analysis were normal. In consultation with the haematologist, patient was started on tablet hydroxyurea 500 mg BD. Patient was followed up and six weeks later repeat haemoglobin electrophoresis showed Hb-7.8%; HbS- 42.8% with TLC and platelet count within normal limit. Subsequently the patient was transfused two units of packed cell volume 2 days prior to surgery and repeat haemoglobin electrophoresis was done. After the transfusion, Hb improved to 9.8gm%, HbS decreased to 33.6% with total leucocyte count 10,200/cu.mm and platelet count 2,70,000/cu.mm.

Preoperatively patient was started on chest physiotherapy and incentive spirometry. Injection ceftriaxone was started prophylactically. Patient was kept nil per oral as per the standard ASA guidelines. Overnight hydration was maintained with Ringer Lactate at 100ml/hour IV. Tablet alprazolam 0.5 mg was given as premedication on the night before surgery. Patient was taken up for surgery under ASA physical status II. Blood grouping and cross matching, with adequate blood availability was ensured prior to surgery.

The plan of anaesthesia was regional anaesthesia using combined spinal epidural technique. After shifting the patient inside the operation theatre, standard ASA monitors were attached (ECG, NIBP, pulse oximeter). Large bore 18G cannula was secured over right forearm and Ringer Lactate infusion via fluid warmer was started. Operation theatre temperature was regulated to keep it between 26-28^{0}C. Vasopressors and inotropes were kept ready. Patient was given left lateral position and under all aseptic precautions combined spinal epidural anaesthesia was given at L2-3 space with 2.2 ml of Injection 0.5% heavy Bupivacaine

intrathecally, subsequently the epidural catheter was fixed at 8cm mark. Patient was turned supine. Care was taken to avoid hypotension following spinal anaesthesia. After achieving adequate sensory level, the patient was given left lateral position for surgery. Adequate padding of the pressure points was done and axillary temperature probe was placed. Patient was wrapped with cotton and covered using warming blanket. Oxygen was administered via venturi mask at 0.35 fiO_2 and nasal $EtCO_2$ was also monitored. To allay the anxiety, patient was given Injection Midazolam 1 mg IV. Injection Paracetamol infusion 1 gm IV was given for multimodal analgesia. Intraoperatively, patient was haemodymanically stable with pulse rate 70-80/min and mean BP between 65-75 mmHg. Intraoperative blood loss was 800 ml which was replaced with 500ml of hydroxyethyl starch and 1 unit of packed cell volume. Intraoperative ABG was within normal limit. Surgery lasted for one and half hours and after the completion of surgery, patient was catheterised to monitor the urine output for assessing the fluid status postoperatively. For postoperative pain relief, injection morphine 1.5 mg (30µg/kg) was given via epidural catheter after ensuring negative aspiration and was repeated as required based on verbal rating scale(VRS≥3). In PACU, patient was given oxygen via venturi mask at 0.35 fiO_2, pain assessment was done using verbal rating scale and ringer lactate continued at 100ml/hr IV till the patient started to take orally. Patient was started on low molecular weight heparin as thromboprophylaxis. The epidural catheter was followed for 2 days and was removed following ASRA guidelines. Postoperatively the patient's stay was uneventful and was discharged on the 5th postoperative day.

Discussion

Due to the vast array of complications of SCD, persons with these disorders often require surgical intervention for treatment or prevention of certain complications. Anaesthetic considerations require a thorough understanding of the implications involved in this already compromised patient. There are few reports where anaesthetic management of sickle cell disease patient for posted for common bile duct exploration[8] and splenectomy[7] is discussed with respect to considerations for general anaesthesia, unlike our case where regional anaesthesia was safely administered to the patient with favourable outcome. Also the previous case reports have discussed about exchange blood transfusion and partial exchange transfusion but we opted for medical therapy with hydroxyurea as we do not have this facility in our set up and also as was an elective surgery where time was not a constraint. With the help of medical therapy, we succeeded in overcoming the complications and cost of exchange transfusion.

Vaso-occlusive phenomena and chronic haemolytic anaemia are the clinical hallmarks of SCD, with multiorgan dysfunction resulting in high morbidity as well as mortality. The basis of anaesthetic management has traditionally been to avoid the factors known to increase erythrocyte sickling and precipitate the vicious cycle of vaso-occlusive episodes, such as volume depletion, hypoxemia, infection, acidosis, hypothermia, hyperthermia, decrease in 2,3-DPG.[2] The risk intrinsic to the type of surgery should be considered. Among orthopaedic procedures, hip surgery and hip replacement are associated with a high risk of complications.[3] Pre-operative examination should aim to find the risk of SCD-related organ dysfunction.

Proper optimization of the patient before taking up for the surgery can help reduce the complications associated with the disease. History of episodes of painful crises must be sought.[6] Preoperative thorough examination and relevant investigations should be done as SCD involves multiple organ systems. It is advised that the HbS should ideally be below 30% so as to avoid sickle cell crisis during the perioperative period. Hydroxyurea has been used to increase the production of HbF, inhibiting HbS polymerization.[4] Wherever facilities are available, exchange transfusion helps in decreasing the proportion of HbS in the blood.

In our patient, preoperative pharmacotherapy with hydroxyurea and blood transfusion before the surgery reduced the HbS level from 68.8% to 33.6% which greatly improved the patient outcome. It is also advisable to start these patients on broad-spectrum antibiotics in the preoperative period as functional hyposplenism makes them susceptible to infection.

The choice of anaesthesia in these patients should be based on the type of surgery and patient's general condition. Regional anaesthesia helps in avoiding airway manipulation, multidrug regimen and postoperative chest infection. In addition there is decreased blood loss, lower incidence of deep vein thrombosis and effective pain management in the postoperative period. Hypotension associated with regional anaesthesia can be prevented and managed with adequate hydration, proper titration of drug used, and the availability of vasopressors and inotropes.

As dehydration precipitates RBC sickling and occlusion of microvasculature at a level of precapillary sphincters, perioperative hydration must be ensured.[5,6] Many of these patients have impaired kidney function due to renal medulla infarction which may interfere with the ability to maintain fluid and electrolyte balance during periods of stress.[8] Consideration for deep vein thrombosis should be given to patients who may be immobilised for prolonged period of time perioperatively. It is adviced to start low molecular weight heparin in these patients in postoperative period.[8] Early mobilization combined with incentive spirometry and chest physiotherapy may reduce pulmonary complications. Operating room temperature

should be set at a minimum 24^0C or maximum achievable based on patient age and underlying medical condition. Active warming devices are helpful in maintaining normothermia. Postoperative pain relief is an essential part of perioperative management in SCD patients as it may precipitate as well as create dilemma in the diagnosis of sickle cell crisis.

In conclusion, most surgical procedures in the sickle cell anaemia patient are a result of the disease process itself. However, this is a disease which rapidly can turn the most routine operation into a challenging situation. Most of the anaesthetic implications focus on the prevention of undesirable outcome. The anaesthetic protocol should be developed using techniques that will least compromise the patient. Preoperative optimization, tailored anaesthesia technique, intensive perioperative monitoring and timely intervention in these patients are essential for a favourable outcome.

References

1. Vinchinsky EP, Lubin BH. Sickle cell anaemia and related haemoglobinopathies. Pediatr Clin North Am 1980;27:429-47.
2. Bharati S, Das S, Majee P, Mandal S. Anesthetic management of a patient with sickle β thalassemia. Saudi J Anaesth 2011;5:98-100.
3. Christine S Rinder. Hematologic Disorders. In: Paul AK, Editor. Stoeltings's Anaesthesia and Co-existing Disease. 5th ed. New Delhi: Elsevier; 2012.p.448-79.
4. Goldberg MA, Brugnara C, Dover GJ, Schapira L, Lacroix L, Bunn HF. Treatment of sickle cell anaemia with hydroxyurea and erythropoietin. N Engl J Med 1990;323:366-72.
5. Marchant WA, Walker I. Anaesthetic management of the child with sickle cell disease. Paediatr Anaesth 2003;13:473-89.
6. Firth PG. Anaesthesia for peculiar cells – A century of sickle cell disease. Br J Anaesth 2005;95:287-99.
7. Bala I, Sahni N, Mitharwal SM. Anaesthetic challenges in a child with sickle-cell disease and congenital heart block. Indian J Anaesth 2016;60:294-5.
8. Ingle SS, Ubale P. Anesthetic management of a patient with sickle cell disease for common bile duct exploration. J Anaesth Clin Pharmacol 2011;27:547-9.