Case report

Anaesthesia management in a patient with sickle cell trait and β thalassemia trait for open reduction of fracture radius and ulna”

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ABSTRACT

Sickle cell disease is associated with sickling, microvascular plugging in presence of hypoxia, acidosis, hypothermia and hypovolemia leading to destructive events in vital organs. High HbS, general anaesthesia and surgical trauma are also responsible for above conditions. Patients with sickle cell disease (SCD) may present to the anesthetist in different clinical settings like perioperative care, management of acute painful crises and intensive therapy for acute respiratory failure. We present a case of a 19-year old patient with sickle cell trait and β thalassemia trait with fracture radius and ulna posted for open reduction under regional anaesthesia. The importance of preoperative stabilization and careful anesthetic strategy is emphasized. Regional techniques are not contraindicated in sickle cell disease and are useful for post-operative analgesia. We describe the anaesthetic management and possible peri, intra, and postoperative problems for these patients while reviewing the recent literature.

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Introduction

Sickle cell disease (SCD) is a complex clinical entity characterized by an inherited chronic hemolytic anemia associated with variable number of acute painful vaso-occlusive episodes[1] Polymerization of hemoglobin-S (HbS) after deoxygenation is the fundamental molecular event that underlies the protean clinical manifestation of SCD [2] It is transmitted via autosomal recessive way. Its acute and chronic symptoms are within the interests of anaesthesiologists [3] There are at least 40 variants of S haemoglobinopathy. Patients with SA haemoglobin (sickle cell trait) usually have a normal life expectancy and few complications. Haemoglobin levels are normal and sickling occurs only under extreme physiologic conditions. Haemoglobin S may be combined with thalassemia trait resulting in haemoglobin S- β thalassemia trait [4] Sickle cell crises is usually triggered by factors such as infection, cold weather, dehydration or other distressed conditions. Increased perioperative complications may result from vaso-occlusion after transient hypoxia, hypothermia, dehydration or acidosis. Inadequate post-operative pain of incision may reduce respiratory effort, leading to poor pulmonary function and relative hypoxia. We planned anaesthesia and analgesia of the patient to avoid vaso-occlusive episodes and prevent complications.

CASE REPORT

A 19-year young male, a known case of sickle cell trait and β thalassemia trait was scheduled for open reduction surgery for fracture radius-ulna.

On general examination, he was icteric and had stable vital parameters with regular pulse rate 90/min and blood pressure (BP) 110/86 mmHg and room air spo2 of 98%. Per abdomen no organomegaly, respiratory and cardiovascular examination was normal. Investigations showed Hb 7.6 gm%; HbS 67.7%; baseline coagulation parameters, liver function tests, renal function tests, blood sugar, electrocardiogram, X-ray chest, 2D echocardiogram and arterial blood gas (ABG) were within normal limits. He received 2 units packed cell volume and Hb increased to 9.1gms%. Since his HbS was 67.7%, partial exchange transfusion given, after which his HbS dropped to 34.4%. Packed cell volume (PCV), total count and platelets were normal.

Consent was taken, adequate starvation confirmed, all monitors attached, O2 started at rate of 5 litre per min through Hudson’s mask. In travenous access was secured with 18 G cannula and IV Ringer Lactate was started to prevent dehydration. Regional anaesthesia with supraclavicular block was the choice of anaesthesia. All the necessary equipments and drugs needed for...
administration of general anaesthesia and for resuscitation was kept ready in order to manage the case of failed block or toxic reactions occurring during the procedure.

Premedication with IV glycopyrrolate 0.2 mg, midazolam 1 mg, fentanyl 1 µg/kg, ranitidine 50 mg and ondansetron 4 mg was given.

Under aseptic precautions supraclavicular brachial plexus block was performed with 12ml of 2% lignocaine, 15ml of 0.5% bupivacaine, 90 µg buprenorphine and 1 ml of NaHCO₃ using paraesthesia technique. No tourniquet was used. Care was taken to avoid hypoxia, hypovolema and hypothermia. Surgery lasted for 2 1/2 hours and vital parameters were stable with minimal blood loss. ABG was done to rule out hypoxia and acidosis. No blood transfusion was required and he was shifted to the intensive care unit for monitoring and observation.

DISCUSSION

Sickle cell disease (SCD) is part of a heterogeneous group of inherited disorders of the β-haemoglobin chain, either in homozygous or heterozygous states or in combination with other haemoglobin β chain abnormality. Homozygous SCD is always associated with increased risk of sickle cell crisis. In the heterozygous state sickling is uncommon as the HbS concentration is <50%. Vaso-occlusive phenomena and hemolysis are the clinical hallmarks of SCD. Vaso-occlusion results in recurrent painful episodes and a variety of serious organ system complications that can lead to disabilities and early death. Haemolysis and other agents have been used to increase the production of HbF, inhibiting HbS polymerization [5] Alkalization using magnesium glutamate, to increase oxygen affinity of Hb in the RBC, has been tried. Oral magnesium supplement reduces erythrocyte dehydration, reducing the cellular concentration of HbS in SCD patients [6] Hemolysis of altered erythrocytes with release of free haemoglobin causes oxidant injury to tissues and reduces nitric oxide level [4] Inhaled nitric oxide and other new investigational drugs have shown promise in being able to reduce the sickling process and eventually uncure sickles [7].

Pertoperative concerns of SCD includes anaemia, chronic pulmonary disease, pulmonary hypertension, cardiomegaly and heart failure, renal failure, haemolytic transfusion reaction resulting from alloimmunisation and extreme vulnerability to dehydration, acidosis, hypoxia and hypothermia [8] Optimal preoperative preparation is required. Prophylaxis against pneumococcal sepsis was given to our patient. The definitive diagnosis of SCD is made with haemoglobin electrophoresis[4] Patients should be hydrated, infections controlled and hemoglobin levels should be within acceptable limits. Preoperative need for transfusion should be determined regarding general condition of the patient and surgical procedure he/she will undergo. Our patient received 2 units of packed cells which increased his Hb from 7.6 to 9.1gm%. Partial blood exchange is generally recommended before major surgical interventions in order to minimize sickling by reducing the circulating HbS concentration below 30% [7, 9, 10] Blood exchange decreases blood viscosity unlike simple transfusions. Moreover, it increases oxygen carrying capacity and decreases tendency to sickle. A clear risk factor for sickling is a low PO2 [9] The aim of these transfusions generally is to obtain a hematocrit level of 35 to 40% with a normal hemoglobin (HbA) component of 40 to 50%. Exchange transfusion may be used with an aim to keep the haematocrit at 30% and reduce the concentration of HbS to 30 to 40% [4] Although the benefit of blood exchange in patients receiving anaesthesia is not clearly proven, it certainly is helpful for patients experiencing sickling crises. Our patient had a partial exchange transfusion after which his HbS dropped to 34.4% from 67.7%. Modern exchange transfusions reduces and maintains HbS within acceptable limits (<40%). It corrects chronic anaemia but suppresses erythropoiesis. High Hb > 11gm% increases viscosity. Crises that may be seen in SCD are: splenic sequestration, aplastic anaemia, right upper quadrant syndrome and acute chest syndrome[4] Factors that precipitate sickle crises such as dehydration, hypoxia, acidosis, infection, hypothermia and circulatory stasis should be prevented [8] Hence goals of anaesthesia was to avoid tourniquet, hypoxia, dehydration, hypovolaemia, acidosis and hypothermia so as to prevent vaso-occlusive crisis and sickling. Hazards introduced by the use of tourniquet include localized stasis of blood flow, acidosis and hypoxaemia with the subsequent formation of sickle cells [9] Regional anaesthesia provides effective analgesia, comfort, reduces complications associated with GA. Regional anaesthesia has been successfully employed for surgery, labor and delivery and pain management [4] Postoperative oxygen therapy, liberal hydration and normothermia is to be maintained for minimum of 24 hrs, because crisis may occur suddenly postoperatively.

CONCLUSION

Surgery in patients with sickle cell trait, β thalassemia trait and SCD carry risk due to the nature of the disease. Prevention of conditions that favour sickling is the basis for recommendation regarding perioperative management. Meticulous anesthetic management in the form of avoiding acidosis, hypoxia, hypothermia, hypovolema, maintaining normocarbia, good intraoperative and postoperative pain relief using regional techniques, postoperative thromboprophylaxis, oxygen therapy with inspired concentration up to 40%, chest physiotherapy, nebulization, incentive spirometry with early mobilization and regular ABG monitoring plays an important role in improving the patient outcome. Pain relief is vital in avoiding pulmonary complications and preventing acute chest syndrome. We successfully managed our patient using regional block, avoiding tourniquet, hypoxia, hypovolema, hypothermia and effective analgesia with buprenorphine.

REFERENCE

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