Anesthetic Management of Patients with Sickle Cell Disease Posted for Bipolar Prosthesis

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Abstract

In case of surgical procedures in sickle cell disease (SCD), patients are associated with high risk of perioperative complications like vaso-occlusive crisis, chest syndrome, post-operative infections, congestive heart failure, cerebrovascular accident and acute kidney injury. Preoperative assessment and stabilization like control of sepsis, blood transfusion, correction of hypoxia, hypothermia, dehydration and acidosis is needed to reduce peri-operative complications. Blood transfusion (Simple, manual exchange and automated exchange) remains an important therapeutic intervention in patients with SCD. The case study below shows the perioperative management of patients posted for bipolar prosthesis due to AVN (avascular necrosis). Adequate analgesia, incentive spirometry, early mobilisation and oxygen supplementation is the mainstay of post-operative management.

Keywords: *Sickle cell disease (SCD), Vaso-occlusive crisis (VOC), Avascular necrosis (AVN).*

Introduction

Sickle cell anemia (SCA) or sickle cell disease (SCD) is an autosomal recessive inherited hemoglobinopathy characterized by the presence of sickle hemoglobin (HbS), absent in healthy individuals. Homozygous individuals (HbSS) are characterized by chronic hemolytic anemia and repeated episodes of acute vascular occlusion leading to heart attacks, intense pain, and organ dysfunction, known as vaso-occlusive crisis (VOC).[1]

Different surgical procedures like hip replacement, obstetric surgery, gall bladder surgery, neuro surgery etc in SCD have been associated with relatively increased risks of peri-operative mortality, vaso-occlusive (painful) crisis, acute chest syndrome, post-operative infections, congestive heart failure, cerebrovascular accident and acute kidney injury.[2-3]

Hence preoperative assessment and stabilization like control of sepsis, blood transfusion, correction of hypoxia, hypothermia, dehydration and acidosis is needed to reduce peri-operative complications.

We are presenting a case of sickle cell disease with diagnosis of avascular nerosis (AVN) of femur posted for bipolar prosthesis.

Case Report: A 35 year old male, a known case of sickle cell disease HbSS pattern, was posted for bipolar prosthesis due to AVN. He was diagnosed to have sickle cell disease at the age of 12 year. He gave history of repeated hospitalization and blood transfusion since last 10 years. Last hospitalization was one month back and received 3 blood transfusions.

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On examination he was afebrile, pulse 100 beats per minute, blood pressure 130/90mmHg, SpO2 100%on room air. He had severe pallor. His respiratory, cardiac & neurological examination was within normal limits. Per abdomen exam revealed hepatomegaly. His lab investigation showed HB 5.6 gm %. Hematocrit was 16%, TLC- 17200/mm^3, platlet count, renal function, liver function, ECG and chest X-ray, were normal. USG abdomen showed mild hepatomegely and ABG showed mild metabolic acidosis. ESR and CRP were 112mm/hr and 78 mg/L. His urine culture was sterile while blood culture showed growth of methicillin senstive coagulase negative staphylococcus. 2Decho was normal and Hb electrophoresis showed HbS 65%. Airway and spine examination did not reveal any abnormality.

Patient was started on antibiotics according to culture sensitivity, warm intravenous fluid, tablets of sodium bicarbonate, folic acid, hydroxyurea. Oral paracetamol & tramadol were given for analgesia. Three blood transfusions were given. Patient was started on incentive spirometry. After one week his TLC count decreased to 10000, ESR & CRP normalized. Hemoglobin & hematocrit increased to 10.1 gm% & 31% respectively. ABG showed resolution of metabolic acidosis. So, patient was now posted for surgery.

Anaesthesia Management: Patient was kept NBM from midnight and allowed to take clear fluid up to 2 hours. Patients was given tablet alprazolam 0.5 mg and tablet pentaprozole 40 mg. Wide bore intravenous cannulation was done and warm IV fluid ringer lactate was started at the rate of 80 ml/hour to prevent dehydration in pre-operative period.

Spinal anaesthesia was planned for intraoperative management and femoral nerve block along with multimodal analgesia was planned for post operative period.

Pulse, blood pressure, oxygen saturation, temperature and ECG were monitored in intra-operative period.

Spinal anaesthesia was given with 25 G Quinkie needle at L4- L5 intervertebral space with 3 ml of 0.5% bupivacaine heavy with 75 mcg of buprenorphine.

Oxygen (5L/min) was given to the patient via facemask. Intravenous fluid 1.2 L of warm ringer lactate was infused along with one unit of blood transfusion. Urine output was maintained throughout the surgery at 40 to 50 ml/hour. Blood loss during surgery was approximately 400 ml. Patient's hemodynamic parameters remained stable in peri-operative period.

Femoral nerve block was given with 0.125% bupivacaine with 4 mg dexamethasone with the help of nerve stimulator in postoperative recovery room.

Patient was kept warm and oxygen supplementation was continued in postoperative period. Multimodal analgesia was supplemented in post-operative period with injection tramadol 100 mg and injection paracetamol 500mg thrice a day. IV fluid and antibiotics were continued in post-operative period. Early ambulation was encouraged.

Discussion

In patients with SCD undergoing surgery, peri-operative period is associated with increased risk of SCD related complications like increased risks of peri-operative mortality, vaso-occlusive (painful) crisis, acute chest syndrome (ACS), post-operative infections, congestive heart failure, cerebrovascular accident and acute kidney injury.[2-3] Careful preoperative assessment and management are important to reduce these risks. Peri-operative management requires a multidisciplinary approach, including surgeons, anesthesiologists, and hematologists.

Predictors of post-operative complications in patients with SCD include

1. Type of surgical procedure low, moderate or high risk.
2. Increased age- associated with disease progression
3. Frequency of recent complications-current activity of disease state
4. Hospitalization, marker of disease severity
5. Temporal clustering of ACS – progression of lung disease
6. Abnormal lung fields on radiograph – evidence of sickle chronic lung disease
7. Pregnancy –increased risk of maternal complications
8. Pre-existing infection – triggering agent for ACS
9. Haplotype – African haplotype have more severe disease than the Asian haplotype.[4]

Blood transfusion remains an important therapeutic intervention in patients with sickle cell disease (SCD), aiming to both increase the oxygen carrying capacity of blood and to reduce the complications of vaso-occlusion. Simple, manual exchange and automated exchange can be effective in reducing the acute and chronic complications of SCD.[5]

A randomized control trial compared conservative and aggressive preoperative blood transfusion in 604 operations in patients with sickle cell anemia. The conservative regimen (aiming for Hb 10 g/dL) was as effective as the aggressive regimen (aiming for Hb of 10 g/dL and HbS<30%) in preventing perioperative complications. Although alloimmunization was more common in the aggressive transfusion group. ACS was the most frequent clinical complication; seen in 10% of patients in both arms of the trial.[6] A subsequent RCT, the TAPS trial (Transfusion Alternatives PreOperatively in Sickle Cell Disease) compared preoperative transfusion with no preoperative transfusion in low- and moderate-risk surgical procedures in patients with HbSS or HbSb0 thalassemia. Patients randomized to transfusion were given a simple transfusion aiming for Hb>10 g/dL (if Hb<9 g/dL) or a partial exchange aiming for HbS<60% (if Hb>9 g/dL). Clinically important complications were significantly increased in the nontransfused group (39% vs 15% of patients, P = 0.023) and this difference was primarily explained by a marked increase in ACS in the un-transfused group (27% vs 3%).[7]

In our case patient was having initial hemoglobin 5.6gm% and HbS 65%. We did 3 simple blood transfusions to build hemoglobin to 10 gm% in preoperative period.

Urinary or respiratory tract infections are closely associated with the development of ACS.[8] Hence preoperative infection needs to be adequately controlledwith appropriate antibiotics. Chest radiograph, hemoglobin oxygen saturation, and lung function tests may delineate the degree of pulmonary pathology. Blood urea and serum creatinine concentrations and urine dipstick screening or proteinuria or occult urinary tract infection are useful and inexpensive screening tools. Other Investigations include screen - for antibodies, extended cross-match for E, C, K groups, arterial blood gas analysis, electrocardiogram, liver function tests, stool examination (mucosal ischaemia) and neurological imaging.[4]

During perioperative period, to avoid triggering of sickle cell crisis, factors like anxiety, emotional stress, infection, dehydration, acidosis, hypoxia, hypothermia, vascular stasis and increased blood viscosity should be addressed.[9]

Intracellular dehydration is a known trigger for HbS polymerization. Whenever possible, prolonged pre-operative fasting must be avoided.[9] The use of regional anaesthesia appears to be a useful means of pain control with effective blocking of nociceptive pathways and also prevent a vaso-occlusive crisis[10]. Use of tourniquet is safe to use provided optimum acid-base status and oxygenation are maintained throughout.[11-14]

Conclusion

SCD is a debilitating disease associated with increased risk of perioperative complications. Optimization of patient's laboratory parameters preoperatively is of utmost importance to prevent perioperative complications like VOC & ACS. Adequate control of infection, correction of anemia with simple or exchange transfusion can reduce the risk of complications. Adequate analgesia, incentive spirometry, early mobilisation and oxygen supplementation is the mainstay of post-operative management.

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