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Case Report

Anaesthetic management of a patient with sickle cell disease: Case report and review of literature

Syama G M[1], Rajesh M C[1,*], Anu Mary Thomas[1], Jithin T N[1]

[1] *Dept. of Anaesthesia, Baby Memorial Hospital, Calicut, Kerala, India*

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ABSTRACT

Sickle cell anaemia is a haemoglobinopathy with an autosomal recessive inheritance. With the advent of advanced medical care, longevity of patients with sickle-cell disease has improved greatly. Our aim is to highlight the perioperative strategies to avoid complications due to the disease itself which can get exacerbated by moderate-high risk surgeries, hypoxia, dehydration, hypothermia, acidosis, vascular stasis and infection.

1. Introduction

Sickle cell disease (SCD) is a congenital haemoglobinopathy resulting in formation of deformed red blood cells, episodic attacks of pain and pulmonary compromise, widespread organ damage, and early death.[1] Highest occurrence of SCD has been reported worldwide among sub-Saharan Africa, the Mediterranean, India and Middle east.[2] India is estimated to have the second highest burden of the disease.[3] India has 8.6% tribal population which is 67.8 million across Indian states. SCD is one of the 10 special health problem in tribals. Wayanad has the largest density of tribal population in Kerala and is the neighbouring district of Kozhikode. We often come across patients with SCD getting admitted for various health issues and it is important to have an understanding of the implications of the illness.

The anaesthetist being experts in pain management and critical care, is commonly involved in the management of acute complications of SCD and in the perioperative care.[4] So, it is essential to know the preoperative optimisation, anaesthetic management and postoperative care of these patients.

2. Case Report

A 31-year-old female, known case of SCD presented to hospital with complaints of right sided abdominal pain, fever and vomiting since last one week. On further evaluation, ultrasonography showed distended gall bladder with a calculus, thickened and oedematous wall with thin pericholecystic fluid, heteroechoic lesion with cystic and tubular structures in the pouch of douglas and splenomegaly. She was also positive for urine pregnancy test. Hence, she was posted for laparoscopic cholecystectomy and salpingectomy.

Patient was diagnosed with SCD at 8 years of age. She had a history of previous hip replacement surgery 8 years back and caesarean section (LSCS) 10 months back under subarachnoid block. Patient also gives history of pulmonary embolism during the postoperative period following LSCS. Now she is on tab Hydroxyurea 500mg and Tab folic acid 5mg once daily along with Tab Warfarin 3 mg 5 days a week and 4 mg for 2 days.

There was no history of any other comorbidities. No history of recent fever or respiratory tract infections. She

* *Corresponding author*.
E-mail address: rithraj2@yahoo.co.in (Rajesh M C).

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had moderate effort tolerance with METS > 4.

On general physical examination, patient weighed 45 kg, was poorly built and nourished, had icterus and vitals were pulse rate of 94 beats/min, regular and blood pressure of 100/65 mm Hg in the right arm in sitting position. On assessment of the airway, mouth opening was normal with adequate neck movements and Mallampati grade was 1. Per abdominal examination revealed tenderness in the right hypochondrium. Other system examination were normal.

Her blood investigations were - Hb 9.5gm%, platelet count – 1.98 lakhs/mm3, total count 5500 cells/mm3, packed cell volume 29%, serum creatinine 0.52mg/dl, serum sodium 133meq/L, serum potassium 3.9meq/L, total bilirubin 9.97mg/dl, direct bilirubin 1.08mg/dl, AST 30U/L, ALT 52U/L, ALP 155U/L. Hb electrophoresis showed HbS 54.5%, HbF 16.4% HbA0 21.5%, HbA1C 4.9%, HbA2 1.8%. Chest x-ray and electrocardiogram taken were normal.

As her HbS was 54.5%, after discussing with surgeon and haematologist it was planned to transfuse one unit packed red cells and to do a repeat Hb electrophoresis. After one unit of blood transfusion HbS was 40.2%, HbF 13.2% and therefore another unit packed cells were transfused. Her HbS was 32.8% and HbF 10% – before taking for surgery.

Patient was started with incentive spirometry preoperatively. She received inj. cefotaxime 1g, inj. metronidazole and inj. paracetamol 1g for analgesia. Warfarin was stopped and bridged to inj. heparin 5000 units s/c 6^{th} hourly. She was kept fasting 8 hours before planned procedure and started on ringer lactate 125ml/hour. She was advised carboload in 250ml of water night before and morning 2 hours before surgery.

In the premedication room, she received injection midazolam 1mg, 0.2mg glycopyrrolate, 8mg dexamethasone and 4mg ondansetron and 1g N-acetyl cysteine intravenously 30 minutes before planned procedure. In operation theatre, all standard ASA monitors – 5 lead ECG, non-invasive blood pressure and pulse oximeter were attached. Patient was immediately covered with warming blanket. Room air saturation was 97%. Oxygen was initiated via Hudson's face mask at a flow rate of 6L/min. Preloading was done with 500ml of warm ringer lactate.

She was given fentanyl 80mcg, priming done with atracurium and induced with propofol 75 mg, atracurium 25 mg and intubated after a dose of lignocaine 50mg with 7mm id cuffed PVC endotracheal tube with cuff pressure of 25cmH2O and fixed it at 20cm at lip level after bilateral air entry was confirmed. Maintenance was using oxygen, air, sevoflurane ensuring adequate depth. A transdermal fentanyl patch releasing 25microgram/hour was placed after induction on the right side of chest. 1g MgSO4 in 1g paracetamol was infused over 15 minutes.

A suction catheter of size 14 was inserted to deflate the stomach. Patient was mechanically ventilated with PCV-VG mode using closed circuit and end-tidal carbon dioxide monitored. The ventilatory settings were adjusted as - tidal volume: 350ml, respiratory rate 14/minute, PEEP 5 cmH20, FiO2 of 55%. An arterial line was inserted in the left radial artery. The temperature was monitored using a probe and maintained at 35°C to 36°C. Intermittent pneumatic compression device for thromboprophylaxis was placed. Induction was performed with care to avoid hypotension and hypoxia. ABG done following induction was normal. It showed Pa02 263mmhg, PaCO2 42mmhg and Hb 11.2g%.

Intraoperative fluid requirements were met with warm balanced crystalloid solutions such as ringer lactate. Intraoperatively patient was hemodynamically stable with pulse rate of 90-96/min and systolic BP of 110-140mmHg. Urine output of 50-60ml/hour was adequate.

Surgery took 2 hours due to presence of multiple adhesions. Neuromuscular blockade was reversed with 2mg of neostigmine and 0.4mg glycopyrrolate intravenously. She was extubated and then shifted to post operative ICU for further monitoring. Patient was kept in propped up position and oxygen was supplemented at 4-6L/min via Hudson facemask. Vitals were stable and she did not complain of pain. ABG was repeated to rule out hypoxia and acidosis. Her urine output was 50-60 ml /hour. Postoperative period was uneventful. Oral fluids were initiated after 4 hours and incentive spirometry was continued in the postoperative period. On day 2 warfarin was restarted and she was discharged on postop day 4.

3. Discussion

SCD is an inherited haemoglobinopathy. Normal adult haemoglobin (HbA) has 2α and 2β subunits. The α and β subunits are coded on chromosome 16 and 11 respectively. Normal adult has 95% HbA ($\alpha_2\beta_2$); 2.5% HbA_2 ($\alpha_2\delta_2$) and 2.5% of HbF.[5]

SCD results from mutation occurring on chromosome 11. This results in substitution of glutamic acid by valine in the sixth position and formation of mutant sickle cell hemoglobin (HbS).[4] Homozygous inheritance or co inheritance of the sickle cell gene with another mutated haemoglobin variant leads to SCD.[6] Heterozygous HbAS causes the sickle cell trait.[7]

Desaturation causes polymerization of haemoglobin molecules leading to sickling of RBC and formation of large aggregates called tactoids.[5]This activates the 'vicious cycle' of venous stasis, capillary congestion, hypoxia which favours further sickling. The sickle cells are fragile and have a reduced life span of 12 days when compared with 120 days, and is rapidly destroyed leading to chronic haemolytic anaemia.[4]

Acute complications of SCD include[8]

1. Acute chest syndrome (ACS)
2. Acute anaemia
3. Acute cholecystitis
4. Stroke
5. Hepatic crisis
6. Priapism
7. Chronic complications include[8]
8. Chronic kidney disease (CKD)
9. Thrombosis
10. Cholelithiasis
11. Avascular necrosis
12. Pulmonary hypertension

Hydroxyurea is used to increase the production of Hb F, as it does not interact with Hb S during sickling, inhibiting Hb S polymerisation. The Hb F within Hb S cells protects from injuries such as distortion and rigidity that occur during sickling process. These Hb S cells with higher Hb F have better survival. Magnesium supplementation decreases cellular concentration of Hb S in SCD by reducing cellular dehydration.[6]

Perioperative recommendations are aggressive hydration, pre-emptive erythrocyte transfusion and adequate analgesia along with utmost care to avoid hypoxia, hypothermia, acidosis.[1]

SCD patients are prone for dehydration as they have impaired urinary concentrating ability. Dehydration is associated with increased blood viscosity and it must be avoided as it can cause sickling.[9] Therefore it is essential to ensure adequate perioperative hydration and minimise prolonged fasting.

In Hb SS disease, preoperative transfusion depends on the health status of the patient, baseline Hb and risk of bleeding with the planned procedure. Optimisation of Hb to 10g/dl is preferred prior to low to moderate risk surgery under general anaesthesia. For high-risk surgeries like cardiac or major neurosurgical procedures and in patients with high perioperative risk a Hb S value <30% is preferred. Indication for exchange transfusion**.**

1. Severe acute chest syndrome not responding to simple transfusion.
2. Acute complications with Hb ≥9g/dL
3. Reduction in Hb S percentage is required such as in stroke or stroke prevention.[10]

A life-threatening complication of SCD is acute chest syndrome (ACS) defined as 'an acute illness characterised by fever, respiratory symptoms, or both, accompanied by new pulmonary infiltrate on chest X-ray'.[10] Differentiating if the respiratory symptoms are due to ACS or bacterial infection is often difficult. SCD patients are also prone to streptococcal infection due to functional hyposplenism.So, it is essential to maintain strict aseptic precautions and also to start broad spectrum antibiotics.[6]

Oxidative stress plays central role in the pathophysiology of SCD. The antioxidant N -acetylcysteine (NAC) is an important precursor of cysteine, has intracellular antioxidant effects.[11]

Ensure adequate hydration by using balanced salt solutions till patient starts drinking well. Maintain SpO_2>94%, if needed supplement oxygen and early incentive spirometry is beneficial. Multimodal analgesia involving regional techniques is advised to maximise analgesia and minimise opioid use, and has the advantage of producing less sedation and PONV. Pain is associated with risk of atelectasis and subsequently ACS.[12]

SCD is a hypercoagulable state due to haemostatic alterations such as increased activation of platelets and clotting cascade, intravascular haemolysis and impaired fibrinolysis. Therefore, hospitalized patients with SCD must receive venous thromboembolism prophylaxis if there is no contraindication since it is associated with increased mortality.[13]

Key points

1. Meticulous aseptic precautions.
2. Optimise Hb S < 30%
3. Avoid prolonged fasting and dehydration
4. Avoid hypoxia, hypovolemia, hypothermia and acidosis.
5. Maintain normocarbia
6. Good perioperative analgesia
7. Postoperative oxygen supplementation and incentive spirometry
8. Early mobilisation and thromboprophylaxis

Table 1:

Preoperative	**Intraoperative**	**Postopeartive**
Improve Hb F	Normothermia	Analgesia
Optimise Hb S	Normocarbia	Oxygen supplementation
Avoid prolonged fasting	Oxygenation	Normothermia
Antibiotics	Hydration	Mobilisation
Analgesia	Analgesia	Thromboprophylaxis
	Mechanical thromboprophylaxis	Early oral intake

4. Conclusion

With increasing life expectancy of patients with SCD, a good percentage of these patients may come to anaesthesiologists with a variety of surgical problems. It is crucial for the anaesthesiologists to avoid any situations which will precipitate sickling crisis. Optimal Hb S level should be below 30 though in emergency situations achieving this target may be difficult. Hb F is protective to sickling crisis.

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6. Conflict of Interest

None.

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Author biography

Syama G M, Resident

Rajesh M C, Senior Consultant

Anu Mary Thomas, Senior Consultant

Jithin T N, Senior Consultant

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