Case Report

Anaesthetic Management Of A Patient In Sickle Cell Crisis With Empyema Gall Bladder

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Abstract:

Patients with sickle cell disease (SCD) may present to the anaesthesiologist for peri-operative care for an elective or emergency surgery, management of an acute painful crisis or intensive treatment for acute respiratory failure. We describe the successful management of a young male patient with SCD, who presented in crisis, posted for laparoscopic cholecystectomy under general anaesthesia. Preoperative optimization and careful anaesthetic planning is of prime importance.

Keywords: sickle cell crisis, cholecystectomy, exchange transfusion, analgesia

Introduction

Sickle cell disease (SCD) is a complex clinical entity characterized by an inherited chronic haemolytic anaemia associated with variable number of acute painful vaso-occlusive episodes [1]. Over 30 million people worldwide have SCD with the high concentration of the disease among persons of African, Middle Eastern and central Indian ancestry [2-3]. Polymerization of haemoglobin S (HbS) after de-oxygenation is the fundamental molecular event that underlies the protean clinical manifestation of SCD [4]. Polymerised HbS confers a characteristic sickle shape to the RBC, in association with other cellular alterations; furthermore, these RBCs are more likely to rupture, releasing damaging cell-free haemoglobin (Hb) into the circulation (haemolysis), with significant consequences that include vascular nitric oxide (NO) consumption and oxidative stress [5]. Process of polymerization is triggered and/or enhanced by hypoxia, vascular stasis, infection, inflammation, increased blood viscosity, vasoconstriction, dehydration, hypotension, stress, cold temperature, acidosis and decreased flow [6,7]. Cholelithiasis is the well recognized complication of chronic haemolysis. About 7% of all deaths among patients with sickle cell anaemia are related to surgery [8]. Surgical procedures in sickle cell patients have been associated with relatively increased risks of peri-operative mortality, vaso-occlusive crisis, acute chest syndrome, post operative infections and congestive heart failure [9,10]. We had
planned anaesthesia and analgesia of this patient to avoid vaso-occlusive episodes and prevent complications.

Case report

A young male patient, who was diagnosed to have sickle cell anemia with homozygous trait two and a half decades back, presented to the hospital with complaints of generalised weakness, abdominal pain and yellowish discoloration of eyes since one week.

The abdominal pain was over the right upper quadrant which was cramping in nature, it was intermittent and lasting for few hours, radiating to upper back between the shoulder with no apparent aggravating or relieving factors and no relation to food intake. It was associated with nausea, vomiting and heart burn. He had 3 episodes of vomiting which was bilious in nature. He had progressive yellowish discoloration of eyes and skin since past one week. It was not associated with pruritis or loose stools. He also complained of high coloured urine since the past two weeks. He had high grade fever on admission which started 2 days back. It was intermittent in nature not associated with chills or rigors. He was prescribed oral hydroxyurea, folic acid and antibiotic prophylaxis with intravenous cefotaxim, metronidazole. Patient also received IV Tramadol 100 mg b.d and inj Fentanyl bolus 100 mcg for analgesia. Ultrasound abdomen showed distended gall bladder with sludge a large 17 mm stone present with gall bladder wall thickening and edema noted. No pericholecystic collection, No CBD stone. He was hence posted for laparoscopic cholecystectomy.

Patient gave a history of sickle cell disease which was diagnosed at the age of 10 years after evaluation for frequent episodes of fever and weakness. Patient was advised to start medication (Tab. Hydroxyurea) but he was noncompliant to medication since the past 5 years. He has had 2 hospital admissions in the past 5 years after complaints of acute back pain, each admission lasting for 5 days of hospital stay. He also gives a history of blood transfusions in the past admissions.

Patient did not give any history of any known co-morbidities. He did not give any recent history suggestive of respiratory infection or joint pain, chest pain suggestive of cardiac involvement. He had moderate effort tolerance (METS >4) prior to admission. He has not undergone any surgeries in the past.

Patient gave a history of the sickle cell disease in younger brother who passed away at the age of 10 years. He also has a daughter who was diagnosed to have sickle cell disease at the age of 5 years and is currently on Tab. Hydroxyurea.

On general examination, patient was moderately built and nourished, he was icteric and had stable vital parameters with regular pulse rate of 90 beats/min and blood pressure of 110/80 mmHg, right arm sitting position. Airway assessment showed normal mouth opening, with a Mallampatti grade 2 and adequate neck movements with sternomental distance of 12.5 cm. Per abdomen examination revealed tenderness in right hypochondrium, murphy’s sign was positive. No visceromegaly. All other systems were within normal limits.

His baseline investigations were as follows: Hb: 9.4 gm%, Total count 20,800 cells/mm$^3$, neutrophil 95%, platelet 3.6 lakh/mm$^3$, packed cell volume 28.3%, retic count 7.8%, creatinine 0.8mg/dl, total bilirubin 6.6 mg/dl direct bilirubin 1.3mg/dl, indirect 15.0 mg/dl, LDH 52 U/L, peripheral smear showed sickle cell anemia in haemolytic crisis with neutrophilia, Hb electrophoresis was done which showed HbS 75.3%, HbF 11%,Hb A0 6.1% Hb A1c 3.8% Hb A2 of 3.4%. Baseline electrocardiogram and chest X-ray were within normal limits. Since the HbS was 75.3%, after discussion with the surgeon it was decided that owing to the risks involved it is better to postpone the surgery until the level of HbS had
reduced. Hence, he underwent four partial exchange transfusions. His antibiotics were stepped up to Inj. Meropenem 1g 8th hourly. There was a serial reduction in the values of HbS which came down to 55.3% and HbF of 7.7% after the first partial exchange transfusion, then a HbS of 48.2% and HbF of 6.2%, followed by HbS of 41.9% and HbF of 5.1% and finally his HbS was 36.7%, HbA0 47.4%, HbF 4.4% HbA1c 5.1% Hb A2 3.1% prior to surgery. Hb was 11.2gm%, Total count 12300cells/mm3, neutrophil 80%, platelet 2.19 lakh/mm3, packed cell volume 32.5%, sodium 136 mEq/L, potassium 3.6 mEq/L, total bilirubin 31.6mg/dl, direct bilirubin 20.7 mg/dl.

Patient was started on incentive spirometry pre operatively. He was kept fasting after 12 midnight and started on ringer lactate 100ml/hour to avoid dehydration.

Patient received intravenous Midazolam 1mg, Ondansetron 4 mg, Dexamethasone 8 mg and Glycopyrrrolate 0.2 mg as pre medication half an hour prior to surgery. On arrival to the operation theatre for the scheduled laparoscopic cholecystectomy, all ASA standard monitors were attached. Pulse oximeter showed room air saturation of 97%. Oxygen was administered via Hudson’s mask. Patient was draped in a warming blanket. He was pre loaded with 1 litre of warm intravenous ringer lactate. He was induced with inj fentanyl 200 mcg (titrated doses), propofol 100 mg, lignocaine 60mg, precurarisation with atracurium and succinyl choline 100mg. Rapid sequence induction was performed as per the present COVID 19 protocol. Intubation was done with Karl Storz videolaryngoscope. Airway secured with 8 mm cuffed polyvinyl endotracheal tube fixed at 21 cm lip level after confirming bilateral equal air entry. Following which 50 mg Atracurium was given intravenously. Endotracheal cuff pressure was 24 cmH20. Patient's stomach was deflated using a suction catheter of size 14. Patient was mechanically ventilated using closed circuit and ET CO2 monitoring (end tidal carbon dioxide) with volume control ventilation (VCV) (tidal volume: 550 ml, respiratory rate 14/minute, PEEP 5 cmH20, FiO2 of 45%). Utmost care was taken during induction to avoid hypotension and hypoxia. ABG was done which was normal. An invasive arterial line was introduced into the left radial artery. A temperature probe was introduced for monitoring which was maintained at 36°C to 37°C. Pneumatic compression stockings were placed on the patient. He was maintained on good depth using oxygen, air, sevoflurane (MAC 0.6-1.2). Fentanyl infusion was administered at 25mcg/hr. A Fentanyl patch of 25mcg was put on the right arm of the patient. Patient was maintained on warm intravenous crystalloids such as balanced salt solution and ringer lactate. 1g MgSO4 was administered as infusion in 100 ml saline. Throughout surgery patient remained hemodynamically stable with pulse rate of 84-96/min and systolic BP of 110-126 mmHg. Urine output was adequate. Intraoperatively a thickened edematous gall bladder with dense omental adhesions filled multiple pigment calculi and pus was noted which was dissected and removed. On completion of surgery, (duration of 2 hours) neuromuscular block was reversed with intravenous neostigmine 2.5 mg and glycopyrrrolate 0.4mg. Patient was extubated on table and shifted to post operative ICU for monitoring with stable hemodynamics and no pain. He received oxygen supplementation 4-6L/min. Repeated ABG analysis ruled out hypoxia and acidosis. Patient was hemodynamically stable throughout and urine output was 100-120ml/hour. Postop period otherwise uneventful patient kept in propped up and incentive spirometry re initiated by post op day 2. He was shifted to ward, discharged on postop day 4.

Discussion

Sickle cell disease must be appreciated as a multi system disease that affects almost all organs [11]. Vaso-occlusive phenomena and hemolysis are the clinical hallmarks of SCD, an inherited disorder due to homozygosity for the abnormal haemoglobin, that is, HbS. The sickle haemoglobin mutation results from a single amino acid substitution of valine for

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glutamic acid in the 6th position of the beta globin chain [12]. Homozygous inheritance of sickle cell gene or co inheritance of the sickle cell gene with another mutated haemoglobin variant results in SCD [2,3,12-15]. Vaso-occlusion results in recurrent painful episodes and a variety of serious organ system complications which can lead to disabilities and early death.

Hydroxyurea and other agents have been used to increase the production of HbF, inhibiting HbS polymerisation [16]. The hemoglobin (Hb) F molecule does not interact with HbS during sickling [17]. Thus, the presence of high concentrations of HbF within the HbS-containing erythrocyte protects this cell from the distortion, rigidity, and injury incurred during the sickling process. A result of this is that HbS cells with high levels of HbF have a survival advantage when compared to HbS cells containing little HbF [18]. Alkalisation 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 [19].

Acute chest syndrome is one of the most serious complications of SCD. Excessive use of IV fluids as well as respiratory sedation from the use of opioid medications and adjuvants potentiate this risk [20]. By definition, acute chest syndrome is the presence of new pulmonary infiltrates with chest pain, tachypnoea, hypoxia, dyspnoea, cough, fever or leukocytosis [11,20,21]. Progressive fibrosis has been detected in children with multiple episodes of acute chest crisis [22]. It is difficult to differentiate respiratory symptoms due to bacterial infection from acute chest syndrome. So, it is advisable to start patients on broad spectrum antibiotics in perioperative period. Functional hyposplenism makes these patients susceptible to streptococcal infection. To prevent pulmonary complications, prophylactic continuous positive airway pressures and incentive spirometry should be started [23].

Proper planning and optimal perioperative preparation is a key to successful management of SCD patients. Adequate hydration to decrease the viscosity of blood, control of infections and getting the haemoglobin levels normal and PCV between 30% and 35% is essential [1]. Normal saline is acidic (9g/dl of sodium chloride contains 154 milliequivalents of sodium, pH of 5.5, osmolarity of 308 milliosmoles per litre) and it increases the viscosity of blood [24]. Hypotonic fluids, in theory, decrease RBC sickling and are preferred [25-27]. Many of these patients have impaired kidney function due to renal medulla infarction which may interfere with their ability to maintain fluid and electrolyte balance during periods of stress. Preoperative need for exchange transfusion depends on the general condition of the patient and the type of surgical procedure. Exchange transfusion is generally recommended before major surgical interventions in order to minimise sickling and reduce the circulating HbS below 30% [28]. In our patient the exchange transfusions done before surgery reduced the HbS level from 75.3% to 36.7% which drastically improved perioperative outcome.

Sickle cell disease is a hypercoagulable state [29,30]. Current evidence suggests increased platelet and coagulation activation, even at patients basal state. SCD patients have low circulating levels of anticoagulant proteins C and S, moderate thrombocytosis, decreased platelet thrombospondin 1 content and increased levels of markers of platelet activation [29,30]. Adequate DVT prophylaxis must be instituted after all major surgeries until patients are sufficiently ambulatory [31].

Conclusion

Meticulous anaesthetic management in the form of avoiding acidosis, hypoxia, hyperthermia, hypovolemia, maintaining normocarbia, good intraoperative and postoperative pain relief, postoperative thromboprophylaxis, postoperative oxygen therapy with inspired concentration upto 40%, chest physiotherapy, nebulisation, incentive spirometry with early mobilization and regular ABG monitoring played an important role in improving outcome. Post operative
monitoring and pain relief play an important role in avoiding pulmonary complications.

**Key Points**

- Meticulous anaesthetic management is of key importance.
- This is done by avoiding acidosis, hypoxia, hypothermia and hypovolemia.
- Normal HbF level less than 0.6%.
- Maintaining normocarbia.
- Good intraoperative and postoperative pain relief.
- Postoperative thromboprophylaxis.
- Postoperative oxygen therapy (4L/min).
- Incentive spirometry with early mobilization and regular ABG monitoring played an important role in improving outcome.
- Post operative monitoring and pain relief play an important role in avoiding pulmonary complications.

**References**


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