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Peri Operative Management of a Patient with Sickle Cell Trait-Hematological Aspects

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Abstract

Sickle Cell Trait (SCT) person carries a single gene for Sickle Cell Disease (SCD) and can pass this gene along to their children. A rare but major concern in patients with sickle cell trait during major surgeries is the development of an intraoperative sickle cell crisis. Most people affected by the trait do not have any health problems caused by it but some do have problems if exposed to conditions causing stress or hypoxia.

Methods and findings: It is a case report of sickle cell trait patient presented with a long bone fracture that we had successfully managed at our hospital.

Keywords: Sickle cell trait; Hematological; Sickle cell crisis; Hypoxia

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Introduction

Sickle Cell Trait (SCT) person carries a single gene for Sickle Cell Disease (SCD) and can pass this gene along to their children [1]. Usually they do not have any of the symptoms of SCD and live a normal life. SCT is more common among people whose ancestors come from Africa, the Mediterranean region, Middle East, and South Asia, but anyone can have SCT [2,3]. A rare but major concern is the development of an intraoperative sickle cell crisis especially when exposed to conditions of hypoxia.

Case Report

A 25 year old male patient presented to the emergency department of our hospital with a history of road traffic accident. On examination he had pain and swelling over the right thigh, with restriction of movement. He was admitted for further evaluation and expert management in the Orthopedic ward. He had no previous medical or surgical history nor was he admitted for any illness in the past. There were no positive general examination findings. Local examination of the right lower limb showed swelling and tenderness over the right middle of thigh along with shortening, external rotation and abduction. The range of movements were restricted. Distal pulses were present. X-ray of the right thigh showed displaced fracture of middle one third shaft of right femur **(Figure 1).**

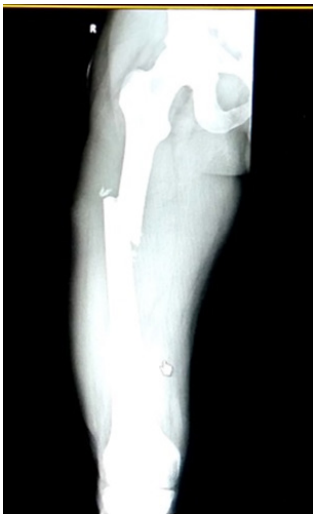

Figure 1: Fracture of middle one third shaft of right femur.

X-ray of the chest was normal. He was under close observation and his blood parameters were evaluated frequently. A decrease in hemoglobin level **(Table 1)** was observed which was also associated with a fall in oxygen saturation as per the Arterial Blood Gas (ABG) report. He developed dyspnea which required oxygen supplementation. Due to sudden development of dyspnea, occurring after fracture of long bone; a provisional diagnosis of fat embolism was made and was started on LMWH Heparin then he was shifted to Intensive care unit and the proposed surgical correction was also postponed.

Table 1: Parameters from day 1 till day 5.

Parameters	1st day	2nd day	3rd day	4th day	5th day
Hb (g/dl)	12.3	11.1	10.1	9.2	9
TC (cells/ cumm)	18,020	17,030	17,690	16,140	13,180
Platelet (cells/ cumm)	1,51,000	1,40,000	1,45,000	1,40,000	1,44,000
CRP (mg/dl)	Raised (11.1 initially)				

Role of Hematology /Transfusion Medicine

A detailed history, examination and further lab investigations were ordered. A peripheral smear examination, osmotic fragility testing and Hb electrophoresis tests were requested. The findings of the investigations are as shown below **(Table 2)**.

Table 2: Findings of the investigations.

Parameters	Results
Hb	8.4 gm/dl
Platelet	1,50,000
Reticulocyte	4.10%
Total Bilirubin	2 mg/dl
Direct Bilirubin	0.26 mg/dl
LDH	570 u/dl
Osmotic Fragility	Normal
Peripheral smear	Normocytic normochromic anemia
Mild neutrophilia, Adequate platelets	Sickling test was positive

Considering the family history of sickle cell disease and dyspnea requiring oxygen associated with the following lab findings, which includes a drop in Hb, increased reticulocytes, increased LDH, indirect hyperbilirubinemia, sickle cells in peripheral smear and a electrophoretic pattern suggestive of HbS trait, a provisional diagnosis of sickle cell trait with hemolysis was made **(Figure 2)**.

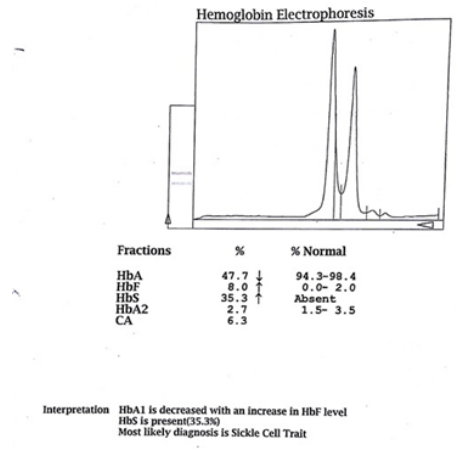

Figure 2: Hb electrophoresis.

The dyspnea and chest discomfort could have been due to the pulmonary complications due to sickle cell disease. Pulmonary complications of sickle cell anemia include reactive airway disease (pulmonary hypertension), pulmonary thromboembolism and acute chest syndrome. Pulmonary hypertension (increased pressure on the pulmonary artery), leading to strain on the right ventricle and a risk of heart failure; typical symptoms are shortness of breath, decreased exercise tolerance and episodes of syncope. This may be due in part to the depletion of nitric oxide [4]. Intravascular hemolysis, by scavenging nitric oxide and causing endothelial cell dysfunction, may play a role in the development of pulmonary hypertension [5].

Optimization of the clinical and hematology parameters was our utmost goal so as to undertake the planned surgery at the earliest. Exchange transfusion was planned so as to remove the sickle cells and give back normal red cells. Prior to the exchange transfusion the patient was pre-hydrated with intravenous fluids comprising of normal saline and ringer lactate. Venesection was done to remove 400 ml of patient's blood. 1 unit of fresh PRBC (less than 5 day old) was given to him. Hydration of the patient was escalated (atleast 50 ml/kg of RL/NS) so as to prevent any sort of dehydration which could trigger a crisis. Hydroxyurea tablet (500 mg OD) and Folic acid (5 mg) was also started. The patient tolerated these procedures without any complications.

After 2 days his Hb increased to 11 gm/dl and dyspnea had subsided. He was taken up for surgery after taking high risk consent. Closed reduction and internal fixation with interlocking nail of right femur was done. Hydration was maintained. A top up transfusion of 1 unit of PRBC (less than 5 day old) was given to him as there was a slight drop of Hb to 10 gm%.

On post operative day one, Hb was 11.2 g/dl. Post operative period was uneventful. He didn't have any further Hb drop. He had no further dyspnea or respiratory complaints. He was discharged on 8th post-operative day with an advice to continue Hydroxyurea and Folic acid for one month and to review in Hematology OPD after one month with a complete blood count.

At review he was stable, with no new complaints and an Hb of 13 gm%. No further dyspnea or breathlessness was seen. He is advised to continue Hydroxyurea and Folic acid for 3 more months and to stop it. He is also adviced to take urgent medical help in case of any surgeries or any further crisis symptoms.

Discussion

Sickle Cell Trait (SCT) is an inherited condition in which both HbA and HbS are produced. HbA=60%, HbS=40%, HbF:<2%. They act as carriers as they have a single gene for sickle cell disease (SCD) and can transmit the disease to their offsprings [4]. They usually do not have any of the symptoms of sickle cell disease **(Figure 3).**

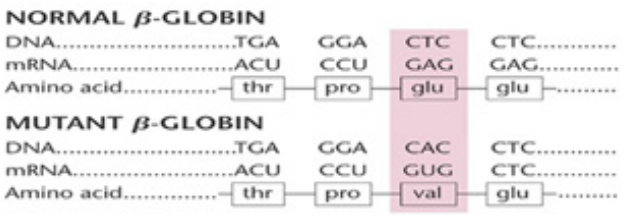

Figure 3: Composition of normal and mutated beta globin (elegant pathophysiology of sickle cell anemia in flow chart> pathophysiology of sickle cell anemia in flow chart elegant sgugenetics pathophysiology of sickle cell anemia, posted at May 24, 2018 13:25 by herigemblong in uncategorized CD and lead a normal life).

Inheritance

If one parent has sickle cell anemia and the other parent is normal, all children will have sickle cell trait. If one parent has sickle cell anemia and other has sickle cell trait there is 50% chance of either with each pregnancy [6]. Inheritance pattern in sickle cell disease is seen in **Figure 4**.

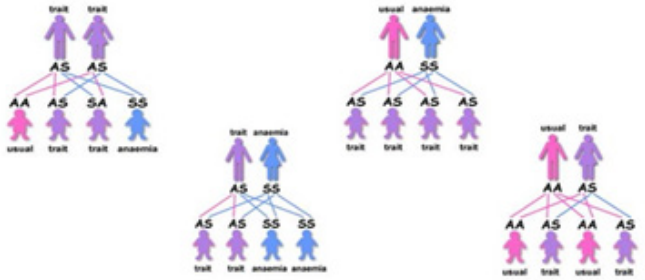

Figure 4: Inheritance pattern in sickle cell disease.

If both parents have SCT,

- 50% chance of having an SCT child.
- 25% chance of having SCD child
- 25% chance will be normal

The pathophysiology of sickle cell disease is seen in **Figure 5**.

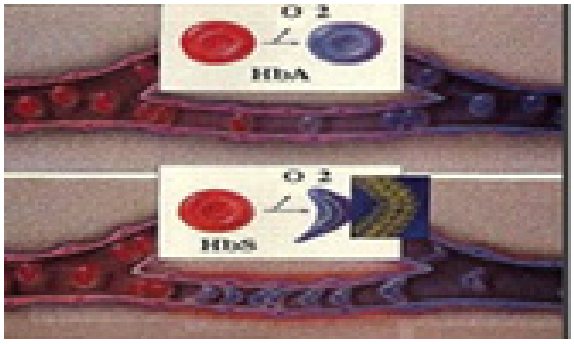

Figure 5: Pathophysiology of sickle cell disease.

Clinical features

Hb concentration is usually low compared to symptoms of anemia i.e. 6-9 g/dl. Sickle cells, target cells and anisopoikilocytosis are observed in blood films and features of splenic atrophy may be present [7,8].

Crisis

1. Vaso-occlusive
2. Haemolytic
3. Sequestration
4. Aplastic
5. Megaloblastic

Vaso-occlusive crisis: This is the most frequent and the most common clinical painful crisis which is the hallmark of SCD's during adult life. For the first 6 months of life, infants are protected by elevated levels of HbF. But as HbS replaces HbF, problem associated with sickling begins. The vaso-occlusive crises occurs when the microvasculature is obstructed by sickled RBCs, causing ischemic injury to the organ supplied and resultant pain which can affect any body part often involving the abdomen, bones, joints, and soft tissues. Main clinical pictures are pain, autosplenectomy, acute chest syndrome present as fever, chest pain, hard breathing, pulmonary infiltrate on chest x-ray.

Aplastic crisis: These occur as a result of infection with Parvovirus B-19 or from folate deficiency. This virus infects RBC progenitors in bone marrow, resulting in impaired cell division. They are characterized by a fall in reticulocytes as well as hemoglobin. Clinical presentation is as pallor, tachycardia and fatigue.

Hemolytic crisis: Characterized by a catastrophic fall in hematocrit, rise in reticulocytes, increased intensity of jaundice, and increasing reticulocyte count. It is triggered by malaria, septicemia, drugs and glucose-6 phosphate deficiency.

Most people do not have any health problems caused by sickle cell trait. Some do have problems if exposed to conditions causing stress/hypoxia. They should drink plenty of water during exercise/strenuous work.

Management of crisis

The main corner stone of management include the following:-

1. Prevention
2. Pain management
3. Blood transfusion/Exchange Transfusion
4. Drugs
5. Bone marrow transplantation.

Prevention

Avoidance of the precipitants of crises should be the main goal of therapy. The use of routine medications (folic acid, multivitamins and malarial prophylaxis). Hydration has to be supplemented. Care should be taken to promote good general hygiene and nutrition. Vaccination for Pneumococcal, Haemophilus and Meningococcal vaccinations and regular oral penicillin are effective at reducing infection rate. Hepatitis B vaccination is also given as transfusion may be needed in later life and management of clinically significant symptoms [9,10].

Crisis

1. Identify and remove the precipitating factor if possible.
2. Treat by rest and warmth.
3. Rehydration by oral fluid/normal saline.
4. Antibiotics if infection is present.
5. Blood transfusions are given only if there is severe anemia with symptoms.

Vaso-occlusion:

1. Pain control using analgesics as per pain score and ladder.

2. Regular blood transfusion.

Haemolytic crisis/Sequestration crisis: Exchange blood transfusion is a main mode of treatment in such patients. Patient must be monitored at regular intervals as attacks tend to be recurrent.

Exchange transfusion: What it does to improve the patient's condition

1. Decrease HbS
2. Increases pre-operative HbA
3. Increases haematocrit
4. Increases cellular oxygen delivery

The blood product used for exchange should be

1. Less than 5 day old product.
2. Tested for HbS and should be negative.
3. Antigentic phenotype at least for the clinically significant antigens.
4. Pre storage leuco reduced.
5. Irradiated in case of neonates.

Exchange transfusion is recommended for all patients with SS requiring high risk surgery (Grade 1C). Preoperative transfusion is recommended for patients with SC undergoing moderate risk surgery and high-risk surgery (Grade 1C)

Hydroxyurea is a newer drug which has made tremendous improvement in the management of patients with Sickle cell Disease and as well as in SCT who present with crisis. The multiple beneficial effects of hydroxyurea for patients with SCA are as follows [11,12].

1. Fetal hemoglobin induction through soluble guanylcyclase activation and altered erythroid kinetics.
2. Lower neutrophil and reticulocyte counts from ribonucleotide reductase inhibition and marrow cytotoxicity.
3. Decreased adhesiveness and improved rheology of circulating neutrophils and reticulocytes.
4. Reduced hemolysis through improved erythrocyte hydration, macrocytosis, and reduced intracellular sickling.
5. Nitric Oxide (NO) release with potential local vasodilatation and improved vascular response.

Prognosis

Sickle cell anemia has no widely available cure. However, treatments to improve the anemia and lower complications can help with the symptoms and complications of the disease in both children and adults. Blood and marrow stem cell transplants may offer a cure for a small number of people but should be done in very severe cases.

Conclusion

There is a consensus that imbalances in homoeostasis, including operative procedures can cause a critical exacerbation of sickle cell disease. Anesthesia and surgery increase sickle-related complications, particularly ACS, while transfusion reduces the risk of peri-operative complications. Meticulous attention should be paid to optimizing all aspects of peri-operative care including oxygenation, hydration and warmth, as well as anesthetic and surgical technique. Close liaison between anesthetist, surgeon and hematologist is essential to ensuring good outcomes.

The case presented here illustrates a strategy for successfully managing sickle cell disease in the perioperative period to minimize its complications by thorough pre-operative stabilization of the patient with respect to Hb and use of initial exchange transfusion.

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Conflict of Interest

Dr. Asha J, Dr. Aboobacker Mohamed Rafi and Dr. Susheela J Innah declare that they have no conflict of interest.

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