

## Case study

### CASE REPORT ON SICKLE CELL ANAEMIA (SS PATTERN)

#### Abstract

Sickle cell anemia is a kind of hemolytic anemia that is passed down in families. It is a kind of hemolytic anemia caused by inheriting the sickle hemoglobin gene. Africans, as well as individuals from the Middle East, the Mediterranean region, and India's aboriginal tribes, have a lower level of the sickle hemoglobin (HbS) gene. A kind of anemia that affects both children and adults is sickle cell anemia. **Clinical Finding:**-Since 5 days, A 25-year-old man has been experiencing generalized bodily pain and anxiety. **Examining the problem:** ALT (SGPT)- 97 U/L, AST (SGOT)- 56 U/L, total bilirubin – 5.4 mg percent, bilirubin conjugated – 1.7 mg percent, bilirubin unconjugated – 3.7 mg percent, total RBC count – 3.71 million/cu mm, total WBC count – 22100 cu mm, total platelets count – 6.46 lack/cu **Ultrasonography**– Heterogeneous spleen. **Therapeutic Intervention** -inj. Piptaz 4.5 gm TDS, inj. Levoflox 500 mg, tab. Hydroxyurea 500 mg, tab. Neurobion forte, inj. Pan 40 mg, inj. Tramadol 100 mg. **Outcome-** The client's condition has improved as a result of the treatment. He has no longer generalized bodily aches, and his anxiety levels have decreased. **Conclusion-** A 25-year-old man was admitted to AcharyaVinobaBhaveHospital's Medicine ward with a history of sickle cell anaemia and complaints of nonspecific body aches and anxiousness. His condition improved after he received proper therapy.

**Key word:**Sickle Cell Anemia

**Introduction** - Sickle cell anemia patients have a reduced life expectancy. Others, on the other hand, can go years without displaying any signs, while some do not make it through childhood. (1) Individuals can now live into their fourth decade if they receive proper treatment. Pain flare-ups, tiredness, bacterial infections, and progressive tissue and organ degeneration characterize the majority of clients. Bacterial infection is the most prevalent cause of mortality, followed by stroke or brain bleeding, as well as renal, cardiac or liver failure. Later three year, the risk of bacterial infection decreases. Despite this, bacterial infections remain the main cause of death in people of all ages. As a result, any clinical feature of infection in a sickle cell anemia patient should be evaluated by a physician to minimize further complications and save lives.

Surprisingly, in some people, they are immune to malaria thanks to the sickle cell gene. As a result, persons who carry the sickle gene have a poor chance of being immune to malaria. In addition, the regional distribution of the sickle cell gene is comparable to the geographic distribution of malaria infection. Sickle cell anemia is a life-threatening condition.(2) Being a sickle cell carrier (trait) may provide a selective advantage if a person lives in a malaria-prone location. The benefits that for individual with sickle cell trait has over someone who isn't a carrier of the gene could explain why, although being fatal, sickle cell **anemia** hasn't vanished from the planet. There is no such thing as a "black gene" for sickle disease. So it is happens to affect a disproportionate number of black people. If a black person with sickle cell disease has children with a non-black person, the sickle cell gene may be passed down to the children, regardless of race. The sickle cell gene is found in people of all races.(3)

Recent study is looking into new techniques to encourage the production of foetal hemoglobin, which delays the onset of sickle cell disease in newborns. Bone marrow transplantation is performed on patients with severe sickle cell anemia who have a sibling donor. Genetic engineering could be used in future therapeutics, potentially leading to cures. Genetic counseling might be important for family members who want to avoid sickle cell anemia. Sickle cell anemia is a disease that can be handed down through generations. Both parents must be sickle cell gene carriers for a child to have sickle cell anemia. If both parents are carriers, a child has a 50% chance of becoming a carrier and 25% chance of inheriting both genes and developing sickle cell anemia.(4)

## **CASE REPORT**

### **Patient Identification-**

A 25-year-old man was brought to the Medicine ward at **AcharyaVinobaBhave Hospital**, Sawangi (M), with a recognized case of sickle cell anemia. He stands at a height of 160 cm and weight 50 kilograms.

### **Patient Medical History-**

Client's medical history is up to date. With complaints of general body aches, anxiety, and restless sleep, a 25-year-old man was admitted to **AcharyaVinobaBhave Hospital**, Sawangi (M). His hemoglobin level had decreased to 10.2 gm% at the time of admission due to sickle cell **anemia**.

### **Past Medical History-**

When patient was admitted for bodily pain and fever at the age of ten months, he was diagnosed with sickle cell anemia.

### **Family history-**

The family of client consists of four members with parents have been diagnosed as carriers of the sickle cell trait. Nonconsanguineous marriage is a type of union. Except for patient, rest of the family is in good health.

### **Clinical Finding-**

Anxiety, generalized body soreness, and a drop in hemoglobin levels (anemia 10.2gm percent).

### **Etiology-**

In the normal state, RBCS and hemoglobin are generated and eliminated at the same time. Anemia arises when the generation of RBC and hemoglobin is reduced and their breakdown is increased. The capacity for transporting oxygen and eliminating CO is reduced. Anemia can have a variety of causes, but it can also be idiopathic in rare situations.(5)

Causes of anemia can be described as follows:

**Impaired of RBC production:** Impaired of RBC production due to deficiency of hemopoietic factors in nutritional deficiency (nutritional anemia). The most common nutritional anemia is iron deficiency anemia. Other nutritional deficiency conditions causing anemia are folic acid deficiency, vitamin B12 deficiency, vitamin B deficiency and vitamin C deficiency.(6)

RBCs are being destroyed at a higher rate (hemolytic anemia).

- Hemolysis caused by internal causes.
- Thalassemia and LEAH sickle cell disease are both caused by abnormal hemoglobin production.
- Glucose-6-phosphate dehydrogenase deficit is an enzyme defect.
- RBC membrane abnormalities or structural flaws in RBC-hereditary spherocytosis.
- Extrinsic factors cause hemolysis.
- Malaria and kala-azar are two infections.
- Immune reaction to Rh or ABO iso-immunization, autoimmune hemolytic anemia, and lupus.
- Primaquine, phenacetin, and phenytoin are some of the medications used.
- Poisoning-lead, Burns, Splenomegaly.

- Blood loss has increased (hemorrhagic anemia)
- Acute trauma, epistaxis, bleeding disorders (leukaemia, purpura, haemophilia), infant hemorrhagic illness, and scurvy are some of the most common.
- Hookworms, bleeding piles, chronic dysentery, and esophageal varices are all chronic conditions.(7)

### **Physical examination-**

In a head to toe examination, there isn't much abnormalities. The client is frail and sedentary. He is frail, but he is not cooperative. It is discovered that the client's spleen is abnormal and has grown in size.

### **Diagnostic evaluation-**

Hemoglobin percent was 10.2 gm%, total RBC count was 3.71 million/cu mm, total WBC count was 22100 cu mm, total platelets count was 6.46 lack/cu mm, ALT (SGPT) was 97 U/L, AST (SGOT) was 56 U/L, total bilirubin was 5.4 mg percent, bilirubin conjugated was 1.7 mg percent, and bilirubin unconjugated was 3.7 mg percent.

### **Therapeutic intervention -**

Inj. Piptaz 4.5 gm TDS, inj. Levoflox 500 mg, tab. Hydroxyurea 500 mg, tab. Neurobion forte, inj. Pan 40 mg, inj. Tramadol 100 mg.

### **Discussion-**

Sickle cell disease a genetic disorder of the red cell but frequently associated with multiple end organ complications if not diagnosed at birth and managed appropriately.(8)

It is estimated that over 2 million Americans are genetic carriers of SCD and that 70-80,000 Americans have sickle cell disease. A common misperception is that SCD affects only people of African ancestry; however, SCD can affect persons of any race or ethnicity. Genes for SCD are common in persons of African, Mediterranean, Middle Eastern, and Indian ancestry and persons from the Caribbean and parts of Central and South America. SCD occurs in approximately 1 in 350 African-Americans.(9)

A 28-year-old female, a known case of sickle cell anemia was admitted for fever and bony pains for 3 days. She developed sudden chest pain, breathlessness and died. She had previous history of jaundice during the ninth month of gestation and found to be sickle cell anemia on investigations.(10)

Sudden death due to exertion has been associated with athletes, police, and military recruits. According to Harmon KG and colleagues, there was a 37 times higher risk of exertional death in Division I football players with sickle cell trait in their database study. As a result of this complication found in athletes, a mandatory policy of the NCAA sickle cell screening program was proposed. Tarini BA et al estimated that over 2000 athletes can be identified with this screening program. These identified individuals can be prevented from having a sudden death if proper intervention is made. Studies have also shown that sickle cell is associated with exertional rhabdomyolysis. Rhabdomyolysis is the breakdown of skeletal muscle cells during physical exertion causing myoglobinuria. There is a 54% higher rate of rhabdomyolysis during physical exertion in the presence of sickle cell trait. It is said to be the cause of the sudden death of a 19-year-old college athlete during intense football training. The death of this college freshman led to the screening policy implemented by the NCAA.<sup>11</sup>

#### **Conclusion-**

A homozygous HbS mutation causes sickle cell anemia (HbSS) The non-covalent polarization of the hemoglobin in the low oxygen condition is encouraged by the lack of polar amino acid on 6 of the globin chain. That gives rise to crumple the RBC's into a sickle shape and restricting their pliability. As a result, as these hard blood cells move through small capillaries, they are unable to soften, resulting in artery occlusion and Ischemia. Having **holistic** care for those who are suffering will be vital. To avoid the misery and crisis that come with the burden of sickle cell anemia, as well as the resources required to care for them, persons who are considering obtaining **treatment for** sickle cell anemia should receive competent and adequate counseling.

**Ethical Clearance:** Taken from institutional ethics committee.

**Sources of Funding:** Self.

**Conflicts of interest:** Nil.

#### **Consent Disclaimer:**

As per international standard or university standard, patient's consent has been collected and preserved by the authors.

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