Case study

CASE REPORT ON SICKLE CELL ANAEMIA (SS PATTERN)

Abstract

Sickle cell anaemia is a kind of hemolytic anaemia that is passed down in families. It is a kind of hemolytic anaemia 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 anaemia that affects both children and adults is sickle cell anaemia. **Clinical Finding:**-Since 5 days, A 25-year-old man have 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 AVBR Hospital'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 anaemia patients have a reduced life expectancy. Others, on the other hand, can go years without displaying any signs, while others 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 characterise the majority of sufferers. 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 anaemia patient

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should be evaluated by a physician to minimise 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 anaemia 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 anaemia 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 haemoglobin, which delays the onset of sickle cell disease in newborns. Bone marrow transplantation is performed on patients with severe sickle cell anaemia who have a sibling donor. Genetic engineering could be used in future therapeutics, potentially leading to cures.Genetic counselling might be important for family members who want to avoid sickle cell anaemia. Sickle cell anaemia 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 anaemia. 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 anaemia.(4)

CASE REPORT

Patient Identification-

A 25-year-old man was brought to the Medicine ward at A.V.B.R.H. Sawangi (M), with a recognized case of sickle cell anaemia. He stands at a height of 160 cm and weighs 50 kilograms.

Patient Medical History-

My patient'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 AVBRH Sawangi (M). His haemoglobin level had decreased to 10.2 gm% at the time of admission due to sickle cell anaemia.

Past Medical History-

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

Family history-

The family consists of four people. My patient has sickle cell anaemia, and both of his parents have been diagnosed as carriers of the sickle cell trait. Non consanguineous marriage is a type of union. Except for my patient, the rest of the family is in good health.

Clinical Finding-

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

Etiology-

In the normal state, RBCS and haemoglobin are generated and eliminated at the same time. Anemia arises when the generation of RBC and haemoglobin 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 haemoglobin 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 anaemia, 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.

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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 25-year-old male adult customer from Adilabad was admitted to the AVBRH medical ward with complaints of generalized body soreness, anxiety, disrupted sleep, and discomfort, as well as haemoglobin levels below the normal range. He has been diagnosed with sickle cell anaemia. As soon as he was admitted to the hospital, an inquiry was conducted and appropriate treatment was begun. He showed progress after treatment and treatment was still ongoing.

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Sickle cell crises, acute chest illness, hemolytic anaemia, and nephrotic syndrome are all symptoms of sickle cell disease. With acute chest syndrome, anaemia is present, but all other symptoms are absent in this patient.

A convincing diagnosis requires haemoglobin electrophoresis, which must reveal the absence of Hb A, 2-20 percent Hb F, and the presence of Hb S. In this situation, haemoglobin electrophoresis revealed the presence of Hb F (10%) and Hb S. (57.1 percent). This patient's peripheral blood smear shows sickle cells, which account for 5 to 50% of red cells. Sickle cell disease patients are more susceptible to bacterial infection, particularly pneumococcal infection. This patient was treated with blood transfusions, antibiotics, steroids, diuretics, and oxygen inhalation. His blood pressure was normal; he had hypoalbuminemia in his serum protein, and albuminuria in his urine. The patient was given steroid injections and intravenous albumin. The patient's oedema was treated with intravenous albumin and blood transfusions. Although it should be used with caution, the introduction of hydroxyurea was the most significant advancement in the treatment of sickle cell anaemia.

A 28-year-old female, a known case of sickle cell anaemia 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 anaemia on investigations.(10)

Conclusion-

A homozygous HbS mutation causes sickle cell anaemia (HbSS) The non-covalant 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 who listic care for those who are suffering will be vital. To avoid the misery and crisis that come with the burden of sickle cell anaemia, as well as the resources required to care for them, persons who are considering obtaining sickle cell anaemia should receive competent and adequate counseling.(11)

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COMPETING INTERESTS DISCLAIMER:

Authors have declared that no competing interests exist. The products used for this research are commonly and predominantly use products in our area of research and country. There is absolutely no conflict of interest between the authors and producers of the products because we do not intend to use these products as an avenue for any litigation but for the advancement of knowledge. Also, the research was not funded by the producing company rather it was funded by personal efforts of the authors.

References-

1. Sickle cell anemia - Symptoms and causes - Mayo Clinic [Internet]. [cited 2021 Oct 23]. Available from: https://www.mayoclinic.org/diseases-conditions/sickle-cell-anemia/symptoms-causes/syc-20355876
2. Rees DC, Williams TN, Gladwin MT. Sickle-cell disease. Lancet Lond Engl. 2010 Dec 11;376(9757):2018–31.
3. Luzzatto L. Sickle Cell Anaemia and Malaria. Mediterr J Hematol Infect Dis. 2012 Oct 3;4(1):e2012065.
4. Salinas Cisneros G, Thein SL. Recent Advances in the Treatment of Sickle Cell Disease. Front Physiol. 2020 May 20;11:435.

5. Anemia: Symptoms, Types, Causes, Risks, Treatment & Management [Internet]. Cleveland Clinic. [cited 2021 Oct 23]. Available from: https://my.clevelandclinic.org/health/diseases/3929-anemia

6. Chaparro CM, Suchdev PS. Anemia epidemiology, pathophysiology, and etiology in low- and middle-income countries. Ann N Y Acad Sci. 2019 Aug;1450(1):15–31.

7. Francis RO, Jhang JS, Pham HP, Hod EA, Zimring JC, Spitalnik SL. Glucose-6-Phosphate Dehydrogenase-Deficiency in Transfusion Medicine: The Unknown Risks. Vox Sang. 2013 Nov;105(4):271–82.

8. Case report on multiple pathologies in an adolescent sickle cell disease patient in Ghana. Int Clin Pathol J [Internet]. 2016 Apr 13 [cited 2021 Oct 23];Volume 2(Issue 2). Available from: https://medcraveonline.com/ICPJL/ICPJL-02-00036.pdf

9. Sawke GK, Dangi CBS. Sickle Cell Disease: Case Study with Clinico-Pathological Aspect. Biomed Pharmacol J. 2015 Feb 16;2(2):463–5.

10. Niraimathi M, Kar R, Jacob SE, Basu D. Sudden Death in Sickle Cell Anaemia: Report of Three Cases with Brief Review of Literature. Indian J Hematol Blood Transfus. 2016 Jun;32(Suppl 1):258–61.

11. Sickle cell disease: MedlinePlus Genetics [Internet]. [cited 2021 Oct 23]. Available from: https://medlineplus.gov/genetics/condition/sickle-cell-disease/