International Journal of Medical Studies
Available online at www.ijmsonline.in

IJMS 7(4), 35-41 (2022)
Print ISSN 2542-2766

A rare case of difficulty in diagnosing sickle cell anaemia

Ms. Ashwini Vaidya[1], Ms. Hemangi Hemant Murkute[2]

[1]*Associate Professor, Godavari College of Nursing.*

[2]*Assistant Professor, Godavari College of Nursing.*

Corresponding author: *Ms. Ashwini Vaidya*

Article history
Received 02 March 2022
Received in revised form 15 April 2022
Accepted 26 April 2022
Available online 30 April 2022

ABSTRACT

Introduction: Sickle cell anaemia is a kind of haemolytic anaemia passed down in families. It is haemolytic anaemia caused by inheriting the sickle haemoglobin 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 haemoglobin (HbS) gene. A kind of anaemia that affects both children and adults is sickle cell anaemia.

Clinical finding: For five days, patient has been experiencing generalised 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 due to the treatment. Patient no longer has generalised bodily aches, anxiety levels have decreased.

Conclusion: My patient was admitted to the Medicine ward with a history of sickle cell anaemia and complaints of nonspecific body aches and anxiousness. Patient condition improved after receiving proper therapy.

Keywords: Haemolytic Anaemia, Heterogeneous Spleen, Sickle Cell Anaemia

This article reviewed by Mr. Shoaib Khan, Dr. Ram. Edited by Dr. Pradeep J., Dr. S Gaur. Available online 30 April 2022.

INTRODUCTION

Sickle cell anaemia patients have a reduced life expectancy. On the other hand, others can go years without displaying any signs, while others do not make it through childhood. 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 characterised the majority of sufferers [1].

Bacterial infection is the most prevalent cause of mortality, followed by stroke or brain bleeding and renal, cardiac or liver failure. Later three years, the risk of bacterial infection decreases. Despite this, bacterial infections remain the leading 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 minimise further complications and save lives [2].

Surprisingly, some people 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 resistant 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. Being a sickle cell carrier (trait) may provide a selective advantage if someone lives in a malaria-prone location. The benefit that an 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 [3].

There is no such thing as a "black gene" for sickle disease. So it 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. A 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 [4].

Genetic counselling might be necessary 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 page and a 25% chance of inheriting both genes and developing sickle cell anaemia [5].

Patient Identification: A female patient age 49-year-old was brought to the Medicine ward recognised sickle cell anaemia case. She stands at the height of 160 cm and weighs 50 kilogram’s.

Present Medical History: My patient's medical history is up to date. With complaints of general body aches, anxiety, and restless sleep, a 49-year-old woman was admitted to hospital, haemoglobin level had decreased to 10.2 gm% at the time of admission due to sickle cell anaemia.

Past Medical History: A female patient was admitted for body pain and fever at the age of ten months, she was diagnosed with sickle cell anaemia.

Family history: The family consists of four people. My patient has sickle cell anaemia, and both of her 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, generalised body soreness, and a drop in haemoglobin levels (anaemia 10.2gmpercent).

Aetiology: In the normal state, RBCS and haemoglobin are generated and eliminated simultaneously. Anaemia arises when the generation of RBC and haemoglobin is reduced, and their breakdown is increased. The capacity for transporting oxygen and eliminating CO is diminished. Anaemia can have a variety of causes, but it can also be idiopathic in rare situations.

The causes of anaemia can be described as follows

Impaired RBC production: Impaired RBC production due to deficiency of hemopoietic factors in nutritional deficiency (nutritional anaemia). The most common nutritional anaemia is iron-deficiency anaemia. Other healthy deficiency conditions causing anaemia are folic acid deficiency, vitamin B12 deficiency, vitamin B deficiency and vitamin C deficiency. RBCs are being destroyed at a higher rate (haemolyticanaemia).

1. Internal causes cause homolysis's.
2. Thalassemia and LEAH sickle cell disease are both caused by abnormal haemoglobin production.
3. Glucose-6-phosphate dehydrogenase deficit is an enzyme defect.
4. RBC membrane abnormalities or structural flaws in RBC-hereditary spherocytosis.
5. Extrinsic factors cause haemolysis.
6. Malaria and kala-azar are two infections.
7. Immune reaction to Rh or ABO iso-immunization, autoimmune haemolyticanaemia, and lupus.
8. Primaquine, phenacetin, and phenytoin are some of the medications used.
9. Poisoning-lead, Burns, Splenomegaly.
10. Blood loss has increased (hemorrhagic anaemia).
11. Acute trauma, epistaxis, bleeding disorders (leukaemia, purpura, haemophilia), infant hemorrhagic illness, and scurvy are most common.
12. Hookworms, bleeding piles, chronic dysentery, and oesophageal varices are chronic conditions.

Physical examination: In a head-to-toe examination, there aren't many abnormalities. The client is frail and sedentary. She is weak, but she is not cooperative. It is discovered that the client spleen is abnormal and has grown in size.

Diagnostic evaluation: Haemoglobin 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 lac/cu mm, ALT (SGPT) was 97 U/L, AST (SGOT) was 56 U/L, total bilirubin was 5.4 mg per cent, bilirubin conjugated was 1.7 mg per cent, 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

A 49-year-old female adult was admitted to the medicine ward with complaints of generalised body soreness, anxiety, disrupted sleep, discomfort, and haemoglobin levels below the normal range. She has been diagnosed with sickle cell anaemia.

As soon as she was admitted to the hospital, an inquiry was conducted, and appropriate treatment was begun. She showed progress after treatment, and treatment was still ongoing.

Sickle cell crises, acute chest illness, haemolyticanaemia, 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 [6, 7].

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 smears 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. Her blood pressure was normal; she had hypoalbuminemia in her serum protein and albuminuria in her 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 treating sickle cell anaemia [8].

CONCLUSION

A homozygous HbS mutation causes sickle cell anaemia (HbSS). The non-covalent polarization of the haemoglobin in the low oxygen condition is encouraged by the lack of polar amino acid on 6 of the globin chain. That gives rise to crumpling the RBCs into a sickle shape and restricting their flexibility. As a result, as these complex blood cells move through tiny capillaries, they cannot 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 and the resources required to care for them, persons who are considering obtaining sickle cell anaemia should receive competent and adequate counselling.

DECLARATION OF COMPETING INTEREST

All authors declare no conflicts of interest.

REFERENCES

1. Sickle cell anaemia - Symptoms and causes - Mayo Clinic [Internet]. [cited 2021 Dec 2]. Available from: https://www.mayoclinic.org/diseases-conditions/sickle-cell-anemia/symptoms-causes/syc-20355876.

2. Wyke RJ. Bacterial infections complicate liver disease. Baillieres Clin Gastroenterol. 1989 Jan;3(1):187–210.

3. Mohandas N, An X. Malaria and Human Red Blood Cells. Med Microbiol Immunol. 2012 Nov;201(4):593–8.

4. How Science and Genetics are Reshaping the Race Debate of the 21st Century - Science in the News [Internet]. [cited 2021 Dec 2]. Available from: https://sitn.hms.harvard.edu/flash/2017/science-genetics-reshaping-race-debate-21st-century.

5. Genetic Counseling (for Parents) - Nemours KidsHealth [Internet]. [cited 2021 Dec 2]. Available from: https://kidshealth.org/en/parents/genetic-counseling.html.

6. Sickle Cell Disease: Practice Essentials, Background, Genetics [Internet]. [cited 2021 Dec 2]. Available from: https://emedicine.medscape.com/article/205926-overview.

7. Quazi1 SH, Khan2 SH, Varma3 SK, Nagpure4 S. Anaphylactic Reactions to Blood Transfusion in a Patient with Iron Deficiency Anemia: A Case Report. Indian Journal of Forensic Medicine & Toxicology. 2020 Oct 29;14(4):6416–9.

8. Hemoglobin Electrophoresis: Reference Range, Interpretation, Collection and Panels. 2021 Jun 16 [cited 2021 Dec 2]; Available from: https://emedicine.medscape.com/article/2085637-overview.