

Case study

CASE REPORT ON SICKLE CELL ANEMIA IN CHILDREN

Abstract

Introduction: - Sickle cell anemia is a kind of severe hemolytic anemia caused by the sickle haemoglobin (HbS) gene, which results in a faulty haemoglobin molecule. The term "sickle cell disease" refers to a set of genetic illnesses affecting red blood cells. People with sickle cell disease have irregularly formed red blood cells, which might be troublesome since they don't stay as long as healthy blood cells once they reach the state of blood arteries.

Clinical findings:-

- Progressive Anemia
- Mild jaundice
- Fever
- Headache
- Growth retardation
- Superadded bacterial infection
- Enlarge heart
- Non healing ulcer
- Organ failure
- Abdominal pain with distension
- Joint pain

Diagnostic Evaluation: - Abdominal pain, fever, Hb (8.5 gm%), Abdomen swelling, Increase abdominal girth, pain in joint and irritability

Ultrasonography: - Abdominal USG

Therapeutic Investigation: - Inj. Ceftriaxone 1mg BD, Inj pantoprazole 20mg OD, Inj. Neomole 30ml SOS, Inj. Tramadol 0.4mg TD S, Cap. Hydra 500mg OD, syr. syncoria 5ml OD, Tab. Folic Acid 5mg OD.

Outcome: - All the investigation of the patient is done and the patient is diagnosed as sickle cell anemia.

He showed significant progress after receiving therapy, and the treatment was kept ongoing until my last date of care.

Conclusion: - A male child of 8 yrs was brought to AVBRH on by his parents with a complaint of abdomen pain with distension, Pain in joints, irritability and he was admitted to pediatric ward. All the investigation of the patient is done and the patient is diagnosed as sickle cell anemia. He improved dramatically after receiving therapy, and the treatment was continued until my final date of care.

Key words: - sickle cell anemia, children

Introduction

The term "sickle cell disease" refers to a set of genetic illnesses affecting red blood cells. Red blood cells in people with sickle cell disease are irregularly formed, which can be problematic since they don't last as long as healthy blood cells once they reach the condition of blood arteries.¹

Sickle cell anemia is a hereditary disease in which a malfunctioning haemoglobin produces chronic anemia, which can lead to a variety of significant problems. Under low oxygen tension, these HBs form crescent-shaped crystals and produce sickle-shaped RBCs. These RBCs tend to impact in capillaries and cause hemolysis with local anorexia. It leads to further sickling. Blockage of capillaries cause infraction in various tissue and organ (spleen, liver, heart, lungs, brain, bones, GIT, urinary tract, muscles.)

The sudden and sever hemolysis in sickle cell anemia is termed as hemolytic crisis the sudden onset of symptoms lead to occlusion causing vaso-occlusive crisis .¹

Definition: An hereditary condition characterised by a crescent-shaped red blood cell. This kind of blood cell can clog tiny blood veins and does not persist as long as regular blood cells, resulting in sickle cell anaemia.

Types of sickle cell anemia:

- Hemoglobin SS disease
- Hemoglobin SC disease
- Hemoglobin SBT(beta) thalassemia
- Hemoglobin SBO (beta zero) thalassemia

Patients Identification: On 07/02/2021, an 8-year-old boy with sickle anemia was admitted to paediatric unit no. 22 at AVBRH. He weighs 10 kilogrammes and stands 112 centimetres tall.

Present medical history: The parents of an 8-year-old boy brought him to AVBRH with complaints of abdominal discomfort with distension, joint pain, and agitation, and he was hospitalised to the paediatric department. He is a know case of sickle cell anemia and his Hb level 8.5 gm% at the time of admission. The child was weak and inactive on admission.

Past medical history: My patient was diagnosed as sickle anemia at when she was admitted to hospital due to abdominal pain with distension, Pain in joints, irritability and tenderness till then ,he was admitted to hospital .he do not any past history of any disease condition.

Family history: There are 4 members in his family. All others members of the family were not having any complaints in their health. In his family no other member is having any problem of hypertension, diabetes, or any other disease condition.

Past investigation and Outcome: All the investigation of the patient is done and the patient is diagnosed as sickle cell anemia.

He showed significant progress after receiving therapy, and the treatment was kept ongoing until my last date of care.

Clinical findings: Abdominal pain, fever, Hb (8.5 gm%) , Abdomen swelling ,Increase abdominal girth, pain in joint and irritability.

Etiology: Sickle cell bug is a hereditary illness triggered by a mutation in the beta globins gene, which results in haemoglobin is, a defective haemoglobin protein. Hemoglobin s transforms a flexible red body fluid cell into a sickle-shaped stiff cell. Sickle cells can obstruct blood flow, causing discomfort and organ damage.

Other members of the child's family have sickle cell trait or other haemoglobin genes that are faulty, such as beta thalassemia. That sickle-cell patient has haemoglobin C, haemoglobin D, and haemoglobin E in his blood.³

Physical examination: There is not much abnormality found in head to foot examination, the child is lean and shrill and having dull look and not active. He is weak and not cooperative. Though it is found that the child is having abdominal ultrasonography.

Diagnostic assessment: Hb%-8.5gm% ,RBC-3.0millions/cu.mm ,WBC-3000/cu.mm ,platelets-4.05lacs/cu.mm ,MCV-76.7n ,MCH25.7pico-gm ,MCHC-33.6% ,Monocytes-03% , Granulocytes-75% , Lymphocytes-20% , Eosinocytes-02% , Biasophiles-00%.

Therapeutic Intervention: Inj. Ceftriaxone 1mg BD, Inj. pantoprazole 20mg OD, Inj. Neomole 30ml SOS, Inj. Tramadol 0.4mg TD S, Cap. Hydra 500mg OD, syr. syncoria 5ml OD, Tab. Folic Acid 5mg OD.

DISCUSSION

Dactylitis, defined as a steady-state haemoglobin side by side of a smaller amount than 7 g per decilitre, was found in this observational study of children with sickle cell anaemia, and leucocytosis in the absence of infection were all linked to poorer outcomes later in childhood. A spike in the percentage of pocked red cells in the blood, which indicates early loss of splenic function, might be a red flag. Though the key laboratory tests for determining prognosis — haemoglobin levels and white-cell count — are generally obtainable, steady-state standards must be gained during a hospital visit when no important issues or additional disorders are present.⁴

A mutation in the HBB gene causes sickle cell anaemia (SCA), which impairs the generation of normal haemoglobin, the body's major oxygen transport protein. SCA is a hereditary disease that runs in families. which means that both parents must pass on two copies of the defective gene for the disease to manifest in the offspring. To put it another way, if both parentages are transporters of the faulty gene, their children have a 25% risk of acquiring the active illness and a 50% chance of becoming carriers. The sickle cell mutation is more common in particular ethnic groups, and it's assumed to be linked to sickled cells' ability to defend them from Malaria. African Americans, Sub-Saharan Africans, Latinos, Indians, Mediterranean descendants, and Caribbean residents are the ethnic groups most likely to be affected. The haemoglobin protein is made up of two alpha-globin and two beta-globin subunits. Carriers of sickle cell disease will have a mutation in one of their beta-globin units, resulting in no clinical symptoms. As demonstrated in Martin's case, these people live regular lives and are mostly impacted by the mutation. People with active illness will have mutations in both beta-globin subunits, causing their red blood cells to sickle. Blood cells that have been sickled become less flexible in their manoeuvring through the vasculature, resulting in a restriction of blood flow to numerous bodily areas. Sickle cell disease is a haemoglobin production disorder caused by a genetic mutation that affects millions of individuals throughout the world. Vaso-occlusive, aplastic, and sequestration crises are common in sickle cell patients. One of the leading causes of death in children with sickle cell anaemia is acute splenic sequestration crisis.. Children with splenic sequestration illness who have not yet had an auto splenectomy, as well as older individuals with sickle cell disease or S-beta thalassemia, may experience a sudden, huge expansion of the spleen with a significant amount of red cell mass trapped. There may be significant hypotension on physical examination, as well as cardiac decompensation and massive splenomegaly. Haemoglobin levels are at least 2 g/dL lower than normal, and a rapid reticulocytotic with enlarged nucleated red cells and moderate to severe thrombocytopenia are seen. ACS is a lung injury-related acute illness marked by chest discomfort, fever, or respiratory symptoms, as well as a new pulmonary infiltrate on a chest radiograph. In children under the age of four, higher WBC counts, dactylitis, and anaemia indicated more severe subsequent outcomes. Dactylitis, often known as hand-foot syndrome, is the most common kind of pain in children with sickle cell anaemia, affecting 50 percent of children by the age of two.⁵

CONCLUSION

A male child of 8 yrs old was brought to AVBRH on by his parents with a complaint of abdomen pain with distension, Pain in joints, irritability and he was admitted to paediatric ward

All the investigation of the patient is done and the patient is diagnosed as sickle cell anemia.

He showed significant progress after receiving therapy, and the treatment was kept ongoing until my last date of care.

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