

## Splenomegaly and Hypersplenism in a Child with Sickle Cell Anemia: Case Report

Haloom Abdel Salam Elhashmi<sup>1</sup>, Hanan Elsalhein<sup>2</sup>, Huda Kutrani<sup>3</sup>,  
Asma Ali Abdulwahed<sup>4</sup>

<sup>1</sup>(Pediatric Department, Faculty of Medicine, University of Benghazi, Libya)

<sup>2</sup>(Pediatric hematology Department, Faculty of Medicine, University of Benghazi, Libya)

<sup>3</sup>(Health Informatics Department, Faculty of Public Health, University of Benghazi, Libya)

<sup>4</sup>(Hematology Unit, Pediatric Department, Benghazi Medical Center, Libya)

Corresponding Author: Haloom Abdel Salam Elhashmi

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**Abstract:** Persistent Splenomegaly in pediatric patients with homozygous sickle cell anemia (HbSS) is uncommon. And Hypersplenism is also uncommon. This case report describes a patient with HbSS who, at the age of six, began to experience hypersplenism as manifested by; progressive splenomegaly; Persistent thrombocytopenia; increasing severity of anaemia with the need for repeated blood transfusion, Leukopenia, High reticulocyte count and Circulating nucleated red blood cells. These episodes occurred with sufficient frequency and severity to warrant splenectomy. He has improved after splenectomy, with a resolution of the thrombocytopenia and leukopenia; improvement in the degree of anemia, decrease in the reticulocytes and disappearance of circulating nucleated red blood cells. This case is presented to emphasize; although, uncommon, splenomegaly can persist in children with homozygous HbSS and can be associated with hypersplenism and even life-threatening infection; bleeding and severe anemia make splenectomy as a mandatory option for saving a life.

**Keywords:** Sickle cell anemia, persistent Splenomegaly, Hypersplenism

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### I. Introduction

Sickle cell anemia is one of the common hemoglobinopathies in the world. It is inherited as an autosomal recessive. It is Widespread throughout Africa, the Middle East, parts of India and the Mediterranean [1]. It causes a wide range of severe and even life-threatening consequences. Sickle-cell anemia (SCA) was demonstrated as a molecular disease occurs due to an abnormality in the hemoglobin [2]. It is now well known that SCA results from a single change caused by the amino acid substitution in the  $\beta$ -globin gene which is located on the short arm of chromosome 11. As a result, valine is produced instead of glutamic acid at the sixth position among the 146 amino acids of the hemoglobin beta chain causing the production of sickle hemoglobin (HbS) [1,3-4]. This causes red cell sickling during deoxygenation, leading to increase rigidity and aggregation in the microcirculation. These changes resulting in the following clinical manifestations of sickle cell anemia as painful vaso-occlusive crises, haemolytic anaemia and end organ damage caused by vasculopathy and tissue ischaemia. Complications may be of sudden onset, known as a sickle cell crisis, that leads to long-term organ damage [5,6].

It can affect any part of the body and one of the most common and an early organ to be affected in SCA is the spleen [7]. It is commonly enlarged during the first decade of life but then undergoes progressive atrophy leading to autosplenectomy [6-12]. However, this is the most observed scenario occurred in SCA; sometimes splenomegaly progressively enlarged make the splenectomy as a mandatory treatment option for different causes like hypersplenism, splenic sequestration crisis, splenic abscess. And rare unique cause as massive splenic infarction [7].

The splenectomy in SCA patients with hypersplenism has a great benefit in making the transfusion need less and eliminating the discomfort that result from the enlarged spleen. postsplenectomy laboratory data show a significant increase in the platelets, white blood cells, hemoglobin, accompanied with a significant decline in the reticulocytes [13].

## **II. Case Presentation**

This case report illustrates the splenomegaly with hypersplenism experienced by a sickle cell anemia patient. Although he enjoyed fairly good health till age 4; he subsequently suffered recurrent episodes of severe anemia. It usually dominates the clinical course with HB around 5 g/dl which necessitate blood transfusion and recurrent infections associated with neutropenia. The infection spectrum ranged from sepsis, chest infection and gastroenteritis managed as in patient with parenteral antibiotics. He eventually became underweight, pale, jaundiced with hugely distended abdomen and severely enlarged spleen (12 cm<sub>up</sub> to umbilicus). Also, the patient complained from abdominal distension, pain, nausea, and vomiting after having milk and fatty meals.

Wide investigation showed pancytopenia with reticulocytosis and lab. Evidence of hemolysis including increased LDH, SGOT, and indirect bilirubin usually dominates the clinical course.

Abdominal USS show splenomegaly, cholecystitis and multiple cholelithesis (gallbladder stones). Thus, diagnosis of hypersplenism was made based on the following criteria; the first criterion chronic hypersplenism was arbitrarily defined as a spleen of 4 cm or more below the left costal margin; the second criterion is hemoglobin concentration <60 g/l. The third criterion is reticulocyte counts >15%, and the fourth criterion is platelet counts <200X10<sup>9</sup>/l, all criteria were recorded.

Laparoscopic cholecystectomy and splenectomy were performed under special vaccination cover. Subsequently, the patient completely recovered and not received blood for a long time.

## **III. Discussion**

Our patient showed progressive splenomegaly, recurrent episodes of severe anemia, thrombocytopenia, severe infection warrant for urgent blood transfusion, and antibiotic starting. The peripheral blood smear is characteristically abnormal with irreversibly sickled cells comprising 5-50% of red cells. Moreover, the nucleated RBC, reticulocytosis with pancytopenia Peripheral blood smear from this patient showed sickle cells. This patient was treated with blood transfusion and antibiotics. USS abdomen showed huge splenomegaly and Cholithesis Chronic hypersplenism was arbitrarily defined as a spleen of 4 cm or more below the left costal margin, hemoglobin concentration <60 g/l, reticulocyte counts >15%, and platelet counts <200X10<sup>9</sup>/l, all were recorded.

The patient recovered from his illness after laparoscopic splenectomy (under coverage of Pneumovax 23, meningococcal vaccine and H. influenza vaccine) and cholecystectomy. Although our patient developed postsurgical severe acute chest syndrome, hydroxyurea start was considered after patient post-surgical recovery. With such treatment, the patient recovered well and discharged. Thus, the patient's follow up was scheduled after ten days in the OPD. The follow up revealed that the patient was healthy. Hence the post-splenectomy showed recovery of the patient clinical as well as laboratory results.

## **IV. Conclusion**

It is possible that hypersplenism may occur more commonly than previously recognized in sickle cell disease and is missed or misdiagnosed. Although in our patient's case there were clear findings suggestive of hypersplenism, some presentations could be more subtle. However, many clinicians especially if not hematologist may not think of the hypersplenism when a patient with sickle cell presents with splenomegaly, pancytopenia and reticulocytosis. The clinicians may think that all SCA patient finally ended in autosplenectomy manifested as non-palpable spleen clinically. They may think this case is sickle thalassemia rather than SCA with persistent splenomegaly and eventually ended in hypersplenism. We hope that this report may lead others who care for people with sickle cell disease to be vigilant to the possibility of persistent splenomegaly and hypersplenism.

### ***Patient Consent***

Written informed consent was obtained from the patient's parent for publication of this manuscript and any accompanying

### **Home massages**

- Not all SCA patient ended by autosplenectomy.
- Persistent and progressive splenomegaly which may eventually result in hypersplenism can happen in sickle cell anemia.
- Hypersplenism is uncommon in children with SCA but more cases are being reported.
- Any patient with SCA or any hematological diseases manifested with odd or strange presentation should be referred to hematologist to clarify the problem and managed properly.
- Any hematological cases should be managed with hematologist.
- Hypersplenism in a setting of SCA can develop with the time and require splenectomy.

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