Case Report

Acute splenic sequestration crisis in an adult with sickle cell disease – A case report

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Abstract

Acute splenic sequestration crisis (ASSC) is a life threatening complication of sickle cell anemia (SCA) that primarily affects infants and children. Acute splenic sequestration crisis is rare in adults. Splenomegaly is rarely seen in adults with sickle cell anemia, however, in patients with splenomegaly the persistence of high Hb F levels and concomitant presence of alpha thalassemia may be involved in predisposing the patients to acute splenic sequestration crisis (ASSC). We report a case of 30 years old male patient who presented with complaints of fever with chills and rigors with jaundice of one day duration, chest pain left lower thorax of one day duration. There were similar complaints four times in the past with fever and jaundice. He developed tender splenomegaly with sudden drop in hemoglobin levels. Sickling Test was positive and his hemoglobin electrophoresis showed HbA-26.5%, HbS-56.4%, HbA2-2.5%, HbF-14.6%. This was a rare case of Acute Splenic Sequestration Crisis (ASSC) in an adult with sickle cell anemia.

Key words

Acute Splenic Sequestration Crisis (ASSC), Sickle cell anemia (SCA), Hemoglobin electrophoresis.

Introduction

Acute splenic sequestration crisis (ASSC) is a life threatening complication of sickle cell anemia (SCA) which affects primarily infants and children. Acute splenic sequestration crisis is rare in adults. Splenomegaly is rarely seen in adults with sickle cell anemia, however in patients with splenomegaly the persistence of high Hb F levels and concomitant presence of alpha thalassemia may be involved in predisposing the patients to acute splenic sequestration crisis (ASSC).

ASSC is defined as acute splenic enlargement with a drop in the hemoglobin level of at least 2

gm/dl and a normal basal reticulocyte count [1]. The pathogenesis behind ASSC is the trapping of RBCs in spleen. Acute venous obstruction of spleen leads to trapping of entire arterial output in the spleen. This leads to hypovolemic shock unless the volume is restored promptly, by blood transfusions [2]. A precipitating factor such as infection of upper respiratory tract or gastrointestinal tract may trigger or amplify red blood cell sickling in spleen leading to obstruction [3].

The life-long prevalence of acute splenic sequestration ranges from 7 to 30% [1] and 50-75% of patients experience more than one episode. Since recurrences are common, an early detection and parental education have a major impact on acute splenic sequestration crisis related mortality [4]. Patients with multiple episodes require surgical splenectomy, a temporary transfusion program or both.

Materials and methods

Day 1

30 years old male patient presented with complaints of fever with chills and rigors with jaundice of one day duration, chest pain left lower thorax of one day duration. There was history of similar complaints 4 times in the past with fever and jaundice. There was no history of cough, breathlessness, palpitations, pedal edema, any indigenous drug intake. There was no history of blood transfusions.

On examination, pallor ++, icteric ++ was present, and there was no clubbing, no koilonychia, and no enlarged lymph nodes. Temperature was normal, PR 100/min, BP 110/80 mmHg, cardiovascular system and respiratory system examination was normal. On palpation of the abdomen, tender splenomegaly ++ was present without any splenic rub. Clinically we thought of Clinical malaria and treated with anti-malarials.

Day 2

On investigations, complete blood picture showed Hb - 10.4%, RBC count – 3.9 mill/cumm, WBC count - 10500 cells/cumm, with Neutrophils - 59%, Lymphocytes - 36% Eosinophils - 02%, Basophils - 0%, Monocytes - 3%, Platelet count normal, Reticulocyte count 0.2%, Smear for Malarial Parasite was negative, Widal Test O-1:20, H-1:20, para A-1:20, paraH-1:20, Dengue serology non-reactive, HIV I and II, HbsAg and HCV - non reactive, RBS – 90 mg/dl, Blood Urea - 40 mg/dl, Serum Creatinine - 0.8 mg/dl, Serum Sodium - 144 meq/l, Serum Potassium - 3.5 meq/l, Serum Chloride - 104 meq/l, Liver Function Tests, Serum bilirubin - 4.8 mg/l, AST – 59 IU/L, ALP - 429 IU/L, ALT - 50 IU/L, Serum LDH - 2248 IU/L. His ECG was normal and X- ray Chest PA view was normal. Bone marrow was not done. Blood group was B Positive.

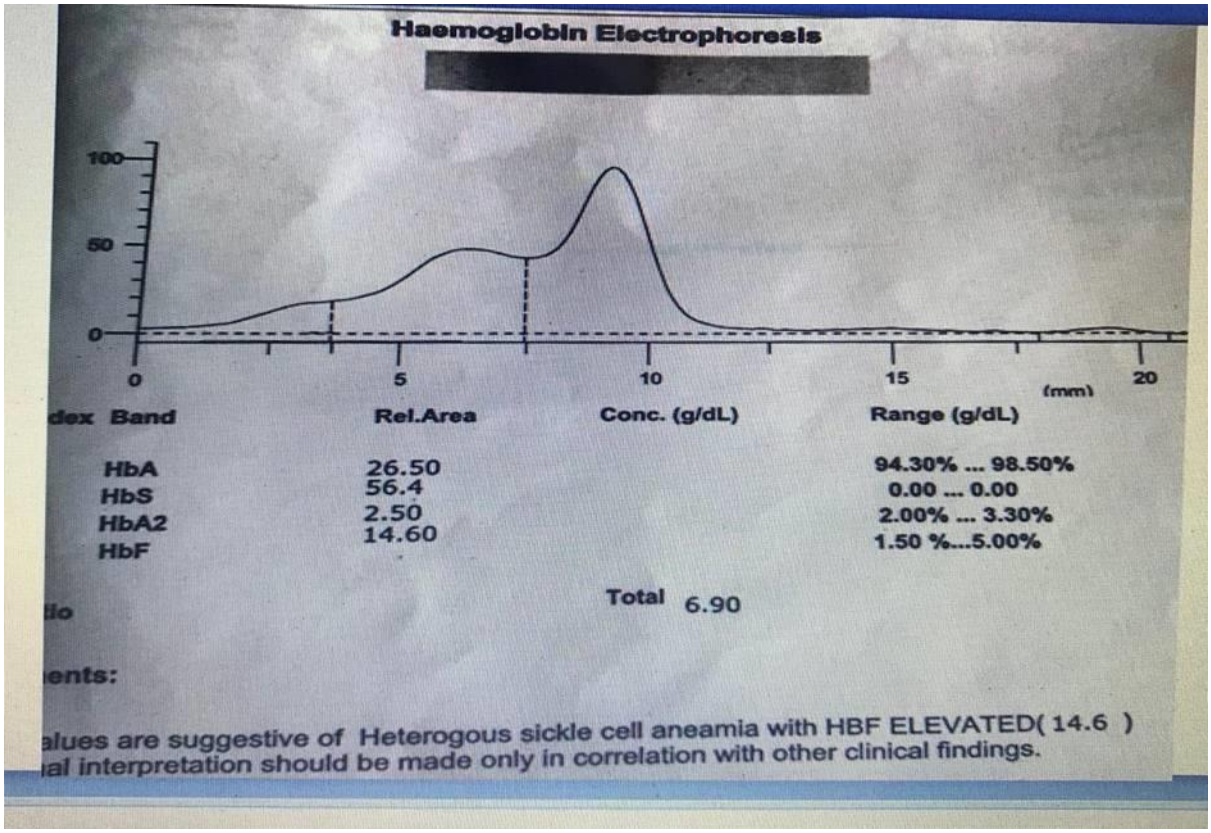
Day 3

On investigations Hb – 6.9 gm/ dl, RBC Count – 2.81 millions/cumm, MCV – 74.4 fl, MCH - 24.7 pg , MCHC – 33.2 G/dl, RDW - 15.7%. Sickling test was positive. His hemoglobin electrophoresis showed HbA 26.5%, HbS 56.4%, HbA2 2.5%, HbF 14.6% (**Figure - 1**).

USG abdomen showed liver 16.7 cm, mildly enlarged in size and shows normal echotexture, no e/o focal lesions and portal veins were normal. Gall bladder distended, no calculi/ sludge, and walls were normal. Pancreas was normal size and echotexture, no e/o duct dilatation. Spleen measured 15.3 cm mildly enlarged and showed normal echotexture, e/o three hypoechoic areas, largest measuring 3.4 into 1.9 cm. On Doppler, no e/o of colour uptake. Both kidneys were normal in size and echotexture, no calculi/hydronephrosis. Urinary bladder-distended no e/o calculi/sludge/wall thickening was seen. Prostate was normal in size and echotexture.

The patient was diagnosed as sickle cell anemia with Acute Splenic Sequestration Crisis, as there was sudden drop in Hb from 10.4 to 6.9 and splenomegaly.

Figure – 1: Hemoglobin Electrophoresis.



Day 4

Patient was given 2 units of whole blood transfusion, after which splenic size regressed to 12 cm. Following transfusion of two units of whole blood transfusion, his hemoglobin was 8.2 g/d.

Day 6

Transfusion of two more units of whole blood transfusion resulted in clinical improvement and a decrease in splenomegaly to 10 cm over the following 4 days. Patient was suggested splenectomy during the follow up.

Discussion

Splenomegaly in Sickle Cell Anemia

Spleen is one of the early and common organs injured in Sickle Cell Anemia. Repeated vaso-occlusions in splenic vessels lead to infarction resulting in progressive atrophy of spleen. However, splenomegaly is also seen in Sickle Cell Anemia and can persist till adulthood. The presence of splenomegaly may predispose for the complications like acute splenic sequestration crisis, hypersplenism, massive splenic infarction, and splenic abscess which may necessitate splenectomy [5-11].

It is seen in infancy and also in some persons in whom it persists till adulthood. The spleen was palpable in 93% of infants by the age of 1 year decreasing to 16% at 10 years [4]. The prevalence of splenomegaly in Sickle Cell Anemia is difficult to interpret because of various interfering genetic and infectious factors. Persistent high level of fetal hemoglobin (HbF) is associated with massive splenomegaly. Concomitant presence of alpha thalassemia is also associated with persistence of splenomegaly and its function [12, 13]. In one study only 24 (6·6%) patients had autosplenectomy i.e., no visible spleen by ultrasound. HbF levels were higher in patients with massive splenomegaly than in those with autosplenectomy. Estimated splenic volume increased with age until about 40 years and then gradually decreased [14].

Infectious causes like malaria is also associated with increased spleen size in sickle cell anemia patients as seen in malaria endemic countries. In Kenya, for instance, in a total of 124 SCA children, splenomegaly was present in 41 (33%) subjects at a median age of 6·3 years [15].

Sickle Cell Anemia and Malaria

The prevalence of malarial parasitaemia were lower in children with sickle cell anemia than in patients without sickle cell anemia with a trend toward a lower incidence of severe forms [16]. However, this protection is not complete. SCA induced hyposplenism may explain this incomplete protection. In Kenya, out of 124 SCA children, splenomegaly was present in 41(31%) subject at median age of 6.3 years. Falciparum carriage was significantly associated with severe malarial anemia and death in SCA patients in Kenya [17] and Tanzania [18].

In a study conducted in Saudi patients, there were 6.6% patients who had autosplenectomy and 11.8% patients had massive splenomegaly, out of 363 patients with mean age 16 years [14]. Malaria chemoprophylaxis is thus recommended in SCA patients living in malaria endemic regions [19].

Interestingly, the prevalence of parasitaemia is generally identical or slightly greater in HbSS than in HbAS subjects, when a stronger protection of HbSS subjects might be expected. Higher concentration of HbS should lead to stronger protection through impaired parasite growth, cytoadherence or enhanced adaptive response. SCA-induced hyposplenism may explain why protection is weaker than expected [20].

Acute Splenic Sequestration Crisis (ASSC)

ASSC is a life-threatening complication of Sickle Cell Anemia (SCA) that is primarily a disease of children. ASSC is rare in adults with SCA and lead to life threatening situation [21]. ASSC is characterized by sudden onset of anemia, splenomegaly, evidence of active bone marrow, and the splenic size regress after blood transfusion.

Depending on severity of attacks ASSC is divided into minor and major attacks.

In minor attacks, there is moderate increase in the size of spleen with a sudden drop of hemoglobin of 2-3g/dl.

In major attacks, there is a significant increase in spleen size with a greater drop of hemoglobin sometimes decreasing to reach as low as 2-3g/dl and hypovolemia.

There are two types of splenic sequestration in sickle cell disease.
1)Acute and
2)Chronic.

1).Acute splenic sequestration is a sudden enlargement of the spleen that can be life-threatening. In sickle cell disease, acute splenic sequestration can happen at any age, but normally it occurs in infants and young children. Acute splenic sequestration happens when sickled red blood cells get trapped in the spleen, causing the spleen to enlarge .The body does not get enough oxygen because of the sickling. If not treated, acute splenic sequestration can cause the body to go into shock. Acute splenic sequestration is a medical emergency.

2).Chronic splenic sequestration may not cause problems and the doctor may choose to record the size of the spleen at each visit to make sure it is not getting any larger. Chronic splenic sequestration usually occurs in older children and adults with sickle cell disease.

The diagnosis is based on clinical signs. A precipitating event, such as fever or infection may trigger or amplify red cell sickling in the splenic red pulp. Upon random accumulation of sickle cells in a zone close to a draining vein, mechanical obstruction of blood flow would induce a drop in oxygen concentration leading to amplification and extension of sickling. This

acute event may be self-limited and transient or lead to extensive irreversible infarction. The symptoms of acute splenic sequestration in a person with sickle cell disease patient will become anemic quickly. Signs of severe anemia include pale skin, weakness, breathlessness, and tachycardia. The spleen will become hard and enlarged. Splenic sequestration can be very painful. Young infants will not have energy to play and will appear extremely drowsy or lack energy and will be hard to awaken. Acute splenic sequestration is a medical emergency.

Recurrence of acute splenic sequestration crisis

Relapse of acute splenic sequestration crisis is frequent with 50-75% of patients experiencing more than one episode [22, 23]. The age at the first episode was the only factor predicting recurrence: the risk was lower when first episode occurred after 2 years than before 1 year.

Differential diagnosis of splenomegaly in sickle cell anemia

1. Primary Lymphoma Of Spleen
2. Hodgkin's Lymphoma
3. Acute Splenic Sequestration Syndrome (ASSC)
4. Splenic Infarction
5. Splenic Abscess

Magnetic resonance (MR) imaging techniques have increased role in detection and characterization of splenic disease and is a excellent tool for diagnosis and evaluation of focal lesions and pathologic conditions of the spleen [24].

Management of ASSC

Treatment for acute splenic sequestration is immediate treatment with red blood cell transfusion. This provides the body with much needed oxygen to the cells and releases the sickled red blood cells trapped in the spleen. The spleen reduces in size and the anemia is corrected. Also includes treating an associated infectious cause. Blood transfusion should be done cautiously to avoid a post-transfusional hematocrit above 35%.

Patient and parental education: A concern following a first episode is preventing the risk of recurrence. Educating patients and their family on how to palpate spleen and identify acute pallor is very important.

Recurrent episodes are managed based on patients' age, severity and parental environment. The strategies include watchful waiting, chronic transfusions and splenectomy. Algorithm for management of recurrent acute splenic sequestration crisis is given in **Figure - 2**.

Splenectomy in SCA and perioperative management

Splenectomy is indicated in SCA when there is evidence of hypersplenism is present or in life-threatening episodes of acute splenic sequestration crisis. Perioperative mortality can be reduced by following steps:-

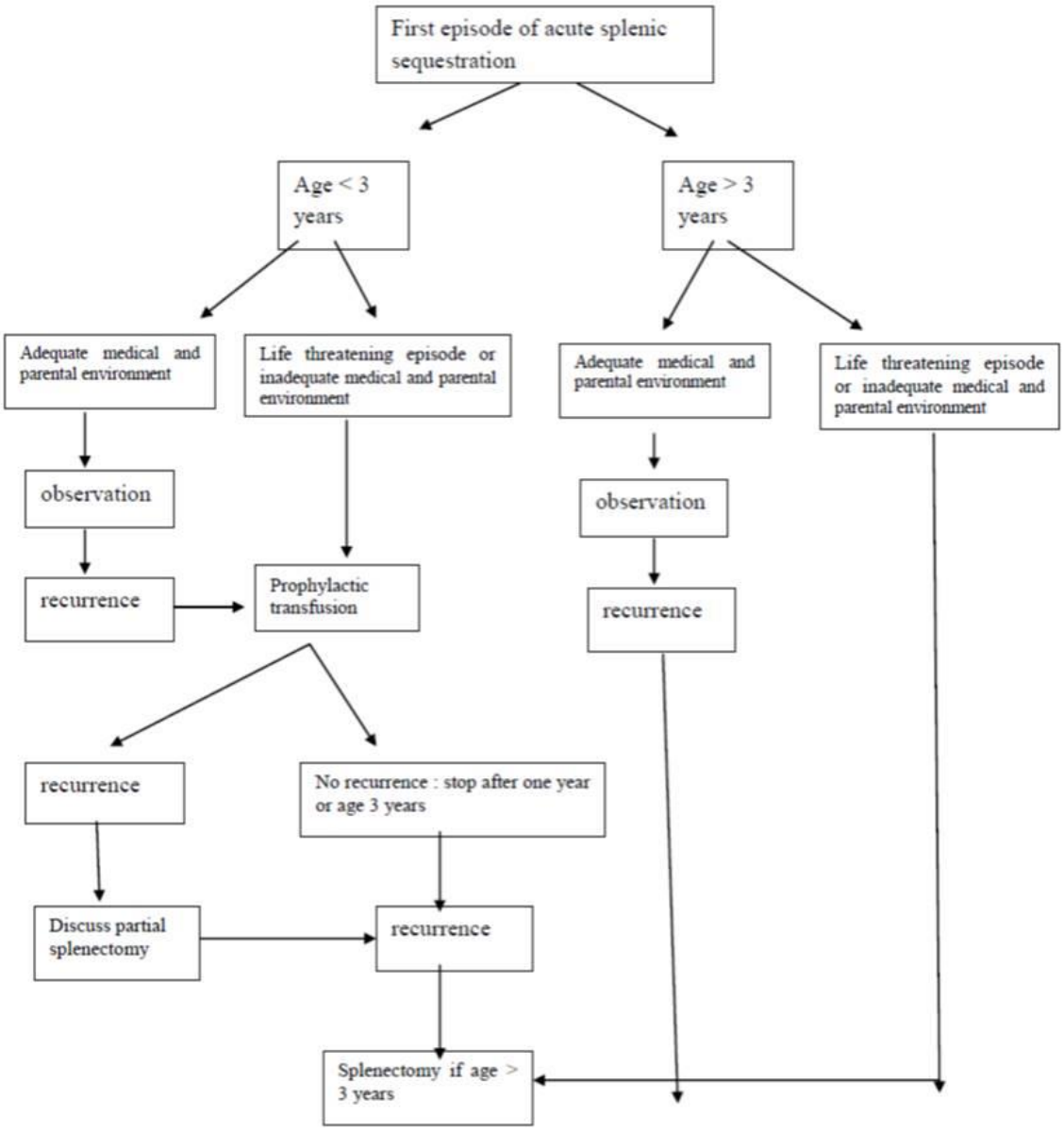
- Adequate preoperative and postoperative hydration.
- Preoperative blood transfusion to increase hemoglobin level to 10-11 g/dl,
- Immunization with Pneumococcal, Meningococcal, H. Influenza Vaccines at least 10-14 days preoperatively. This should be given as soon as possible postoperatively in case of emergency operation.
- Penicillin prophylaxis should be initiated if required.
- Avoid intraoperative hypoxia, acidosis, hypothermia and hypercarbia.
- Use adequate painkillers postoperatively and
- Early mobilization and chest physiotherapy.

Laparoscopic splenectomy should be preferred over open splenectomy because of its superior cosmetic appearance, shorter hospital stay, early postoperative recovery, and less postoperative complications.

Conclusion

It is commonly thought that there is no splenomegaly in Sickle Cell Anemia. But it is not so. Splenomegaly is seen in Sickle Cell Anemia both in children and also in adults. Earlier it was thought that splenomegaly is not seen in adults with sickle cell anemia because of progressive atrophy but this is not true in every case. Acute splenic sequestration crisis (ASSC) is a rare complication of sickle cell anemia (SCA) which is seen in children, but is also seen in adults. Acute splenic sequestration crisis (ASSC) should be suspected when a patient with SCA presents with sudden onset of anemia and splenomegaly. It is a medical emergency and should be treated accordingly with blood transfusions and with splenectomy.

Figure – 2: An algorithm for management of acute splenic sequestration [3].



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