



Splenic Sequestration Crisis in Sickle Cell Anemia: A Case Report

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ABSTRACT

Sickle cell anemia is one of the most common genetic disorders worldwide. It is an inherited disease of public health importance especially in sub-Saharan Africa. Acute painful crisis is the hallmark of the disease with major impact on the psychosocial wellbeing of both the patient and caregiver. Here, we report a case of 8yr old female with sickle cell disease (SCD). She presented to the emergency unit with complaint of recurrent abdominal pain of 4 weeks duration and whitening of the palms and soles of the feet of 3 days duration. Laboratory investigations revealed anemia with packed cell volume (PCV) of 16% and ultrasound scan with impression of splenomegaly. This case shows that splenectomy is one of the mainstays in the treatment of sequestration crisis. It equally demonstrates that ultrasound scan is beneficial, especially when contrasted computed tomography (CT) is not available as is found in many resource-poor settings.

Keywords: Sickle cell anemia, splenic sequestration crisis, ultrasound scan.

INTRODUCTION

Sickle cell anemia is an inherited autosomal recessive disorder (Graham, 1985; Quinn and Miler, 2004). Splenic sequestration is one of the feared complications of sickle cell anemia (SCA) that primarily affects young children (Salako et al, 2020). Sickle cell disease (SCD) is characterized by a point mutation that results in the substitution of glutamic acid with valine on the sixth amino acid of β -globin. The mutant Hb molecules, hemoglobin S (HbS), polymerize upon deoxygenation, causing the erythrocytes to assume a sickle shape and reduce the fluidity of the cell membrane (Jang et al., 2021).

Under condition of hypoxic stress, HbS-containing RBCs undergo shape transformation to the characteristic sickle form. The deformed RBCs exhibit increased stickiness, causing abnormal adherence to the endothelium (Osunkwo et al., 2020). The principal cause of vaso-occlusive crisis (VOCs) is microvascular occlusion, leading to increased inflammation and tissue ischemia-reperfusion injury (Frenette, 2004; Ballas et al., 2012). Complications of SCD include unpredictable, recurrent acute pain, as well as significant multiorgan dysfunction and premature mortality (Yawn et al., 2014; Azar and Wong, 2017). The hallmark of SCA worldwide remains the acute painful episode also known as the vaso-occlusive crisis (VOC) which is an acute drop in hemoglobin of 2g/dl from the patient's baseline accompanied by splenomegaly (Ilan et al, 2023). A study by Brousseau et al, (2011) showed that of the close to 40, 000 visits to the emergency department for VOC, about 40% of the treat and release visits had either an inpatient hospitalization or had another treat and release at a different emergency department within 14 days of the index visit.

CASE PRESENTATION

We reported a case of an 8 year old female who is a known sickle cell anemic patient. She presented to the emergency unit on account of recurrent abdominal pain of 4 weeks duration and whitening of the palms and soles of the feet of 3 days duration. She has had 3 episodes since onset with similar characteristics of about 4 day's interval. The current episode started 68 hours prior to presentation mainly in the left upper part of the abdomen. It was stabbing in nature, severe enough to prevent her from her daily routine activities, there was no relieving or aggravating factors and the pain does not radiate. There was no associated loss of appetite, vomiting, passage of loose stool, no history of abdominal trauma to the abdomen prior to onset of symptoms. Patient's caregiver (mother) noticed associated whitening of the palms and soles of the feet 3 days prior to presentation, which has been progressing until presentation. There was

associated dizziness, but no bleeding from any of the orifices, passage of bloody stool, passage of worm in the stool, yellowish discoloration of the eyes, and no hematuria. She is a known SCA patient diagnosed 6 years ago at Chukwuemeka Odumegwu Ojukwu University Teaching Hospital, Awka. Her steady state packed cell volume (PCV) is 16% (0.16L/L). She was regular on her clinic visits and routine medications since diagnosis. She has had several hospital admissions and the last one was 2 months prior to this current admission. She has had several blood transfusions due to crises (vaso-occlusive crisis).

ON EXAMINATION

Patient was in obvious painful distress, pale, febrile to touch, not cyanosed, anicteric, well hydrated with no peripheral edema and palpable significant lymphadenopathy. Pulse rate was 142 b/m (elevated for age), SPO₂ 99%, Temperature 38°C, respiratory rate 48 c/m (elevated for age).

Anthropometry: weight 23kg (90.2% of expected), height 110 cm (90.2% of expected).

Digestive system Examination: good oral hygiene, appropriate dentition.

Tongue, mucous membrane and palate were all normal, swallowing reflexes were intact.

Abdomen was asymmetrical, distended, and moved with respiration. Tenderness was noted on the left hypochondrium. Spleen was palpable and measured about 13cm below the left costal margin. It was tender with smooth surface, firm in consistency and splenic notch was felt. The liver was not palpable and there was no demonstrable ascites. Bowel sound was present and normoactive.

DIAGNOSIS

A diagnosis of anemic crisis — sequestration crisis precipitated by (? acute) uncomplicated malaria

Differential diagnosis: mixed crisis (vaso-occlusive crisis- mesenteric crisis)

TREATMENT PLAN

Urgent full blood count with differentials and ultrasound scan were requested. Patient was transfused with 2 units of blood as packed cell volume (PCV) was 16% and eventually worked up for partial splenectomy after result of the ultrasound revealed marked splenomegaly which measured 17.11cm in size. After partial splenectomy, patients' visits to emergency department have reduced.



Fig. 1: Ultrasound picture of the spleen



Fig. 2: Ultrasound picture of the spleen



Fig: 3 The Spleen after splenectomy

DISCUSSION

Clinical assessment accompanied by a thorough history and focused physical examination is often used in making the diagnosis of acute splenic sequestration crisis (Brousseau et al, 2014). The severity of the anemia is often assessed with full blood count and its differentials. Reticulocyte count is important to rule out other possible causes of acute anemia (Ilan et al, 2023). Provided that the splenomegaly is appreciable, imaging of the spleen is mostly not needed for the diagnosis to be made. Patient's discomfort is often the major limitation of ultrasound imaging (Ilan et al, 2023). If the diagnosis is being queried, a contrasted computed tomography (CT) of the abdomen is also considered Sheth et al, (2000). A study done by Owusu et al (2017) compared the treatment options: splenectomy versus conservative management for acute sequestration crises in people with SCA. It was concluded that Splenectomy would prevent further sequestration and if partial, may reduce the recurrence of acute splenic sequestration crises. However, there is a lack of evidence from trials showing that splenectomy improves survival and decreases morbidity in people with sickle cell disease. Hence, there is a need for a well-designed, adequately-powered, randomized controlled trial to assess the benefits and risks of splenectomy compared to transfusion programs, as a means of improving survival and decreasing

mortality from acute splenic sequestration in people with sickle cell disease. Study by Al Salem et al, (2006) showed that splenectomy with good perioperative management in children with SCA is not only safe, but also beneficial in treating sickle cell, hence eliminating the risks of acute splenic sequestration crisis, reducing patients' transfusion requirements, and eliminating the discomfort/ mechanical pressure of the enlarged spleen.

A study done by Vick et al (2009) revealed that partial splenectomy prevents splenic sequestration crisis in sickle cell disease. They also concluded that acute splenic sequestration is a fatal complication in sickle cell disease. Total splenectomy in younger patients may predispose them to a greater risk of infections, whereas partial splenectomy may maintain immuno-competence.

CONCLUSION

Partial splenectomy decreases the risk of sequestration crisis in SCD and reduces the need for blood transfusions. Infection rates did not increase after the procedure during the follow-up period. Partial splenectomy should be considered for patients who experience multiple acute SS crises or have long-term transfusion requirements.

REFERENCES

- Al-Salem AH. (2006). Indications and complications of splenectomy for children with sickle cell disease. *J Pediatr Surg.* 41(11):1909-15.
- Azar S and Wong TE (2017). Sickle cell disease: a brief update. *Med Clin North Am* 101: 375-393.
- Ballas SK, Gupta K and Adams-Graves P (2012). Sickle cell pain: critical reappraisal. *Blood* 120: 3647-3656.
- Brousseau DC, Steiner CA, Owens P, Mosso A, Panepinto JA. (2011). Emergency department treat-and-release visits for sickle cell disease: a sign of acute events to come. *Blood.* Pp118:169.
- Brousseau V, Buffet P, Rees D (2014). The spleen and sickle cell disease: the sick(led) spleen. *Br J Haematol.* 166 (2): 165- 76.
- Frenette PS (2004). Sickle cell vasoocclusion: heterotypic, multicellular aggregations driven by leukocyte adhesion. *Microcirculation* 11: 167-177.
- Graham RS, Sickle cell disease, In Graham RS. *Sickle cell disease (1st Edn.)*, USA: Oxford University Press; 1985: 1-475.
- Ian K, Abhishek K, Elftheria A, Shivaraj N (2023). Splenic Sequestration Crisis. National Library of Medicine.
- Jang T, Poplawska M, Cimpeanu E, Mo G, Dutta D and Lim SH (2021). Vaso-occlusive crisis in sickle cell disease: a vicious cycle of secondary events. *J Transl Med*, 19: 397. <https://doi.org/10.1186/s12967-021-03074-z>.
- Osunkwo I, Manwani D, and Kanter J (2020). Current and novel therapies for the prevention of vaso-occlusive crisis in sickle cell disease. *Ther Adv Hematol* 11: 1-16.
- Owusu-Ofori S and Remington T. (2017). Splenectomy versus conservative management for acute sequestration crises in people with sickle cell disease. *Cochrane Database Syst Rev.*11(11):CD003425.
- Quinn CT, Miller ST (2004) Risk factor and prediction of outcome in children and adolescent with Sickle cell anaemia. *Hematol Oncol Clin N Am* 18(6): 1339-1354.
- Salako A, Aworanti O, Okoli C, Odubela O, Ogundeji S, and Temiye E (2020). Pattern and severity of Vaso Occlusive Crisis in Pediatric sickle cell Anemia Patients. *Academic Journal of Pediatrics and Neonatology.* 9 (5): 0060- 0064.
- Sheth S, Ruzal- Shapiro C, Piomelli S, Berdon WE (2000). CT imaging of splenic sequestration in sickle cell disease. *Pediatr Radiol.* 30 (12): 830- 3.
- Vick LR, Gosche JR, Islam S. (2009). Partial splenectomy prevents splenic sequestration crises in sickle cell disease. *J Pediatr Surg.* 44(11):2088-91.
- Yawn BP, Buchanan GR, Afenyi-Annan AN (2014). Management of sickle cell disease: summary of the 2014 evidence-based report by expert panel members. *JAMA*, 312: 1033-1048.

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