Case Report

Splenomegaly and Splenic Pseudocyst in a Female Teenage Patient with Sickle Cell Anemia–A Case Report

AK Orolu[1], SF Ahmad[2], OT Oluwaseun[1]

[1]Department of Heamatology, Alimosho General Hospital, [2]Department of Surgery, Division of General Surgery, Alimosho General Hospital, Igando, Lagos State, Nigeria

Received: 13-Nov-2022;
Revision: 12-Feb-2023;
Accepted: 19-May-2023;
Published: 14-Jul-2023

Abstract

Splenomegaly is one of the complications of sickle cell disease (SCD) occurring in early childhood. This risk is reduced by the age of five years as the spleen undergoes auto splenectomy as a consequence of recurrent vasooclusion and splenic infarction. However, in some variants of SCD, the persistence of the spleen occurs. This can be complicated rarely by the formation of a splenic pseudocyst. We report a 17-year-old teenager with sickle cell anemia who presented with an 8-year history of gradually increasing left-sided abdominal swelling and a 2-month history of recurrent left upper abdominal pain. A computerized tomography scan revealed splenomegaly and multiple splenic cysts, not responsive to opioid analgesics which necessitated a total splenectomy. The histology report found an absence of epithelial lining confirming splenic pseudocysts. SCD patients with splenomegaly have underlying splenic infarction, which is a predisposition to splenic pseudocyst formation, though a rare occurrence. Many patients with splenic pseudocyst are usually asymptomatic and do not need intervention. However, the index patient developed serious symptoms indicating a need for a total splenectomy. A splenic pseudocyst is rare and found in <1% of splenectomies.

Keywords: *Case report, sickle cell anemia, splenic pseudocyst, splenomegaly, total splenectomy*

Introduction

Sickle cell disease (SCD) is commonly associated with splenomegaly in early childhood, however, as the child grows the spleen undergoes auto splenectomy due to repeated red cell sickling and splenic infarction.[1] In some older patients, though uncommon, the spleen persists till adulthood[2] giving rise to some complications like splenic sequestration, splenic infarction, and rarely splenic pseudocyst. Splenic complications in SCD have been associated with increased mortality and morbidity which can be addressed by splenectomy. We present a 17-year-old female teenager with a history of splenomegaly and recurrent splenic pain who was found to have multiple splenic cysts and splenomegaly on computerized tomography (CT) scan and ultrasound scan imaging and was treated with open splenectomy. A splenic pseudocyst is rare and found in <1% of splenectomies.[3]

Access this article online

Quick Response Code:

Website: www.njcponline.com

DOI: 10.4103/njcp.njcp_793_22

Case Report

A 17-year-old female sickle cell anemia (SCA) patient presented to the hematology outpatient clinic with a recent history of recurrent left upper abdominal pain. The pain was dull, intermittent, and mild to moderate in severity. The pain was initially relieved after taking non-steroidal anti-inflammatory drugs. The pain was associated with left upper abdominal swelling. No history of fever, vomiting, or diarrhea. She first noticed left upper abdominal swelling at nine years of age which gradually increased over the years. She has a history of frequent painful vaso-occlusive episodes (six per year). No previous history of surgery, blood transfusions, trauma to the abdomen, or hydroxyurea use.

Address for correspondence: Dr. AK Orolu, Department of Heamatology, Alimosho General Hospital, Igando, Lagos State, Nigeria.
E-mail: bukrolly@yahoo.com

How to cite this article: Orolu AK, Ahmad SF, Oluwaseun OT. Splenomegaly and splenic pseudocyst in a female teenage patient with sickle cell anemia–A case report. Niger J Clin Pract 2023;26:844-6.

Physical examination revealed a young girl who was pale and icteric. Abdominal examination revealed a smooth, firm, and mildly tender spleen in the left hypochondrium, about 6 cm below the left coastal margin.

The hematological parameters were stable. Hemoglobin (Hb) quantification done using high-performance liquid chromatography reported a Hb S fraction of 93.3%, fetal Hb (HbF) fraction of 2.8%, and Hb A2 fraction of 3.5%, suggestive of HbSS and borderline HbF and HbA2 levels. Abdominal ultrasound scan (USS) revealed multiple micro well-defined cystic lesions with heterogeneous conduct within the parenchyma, the largest 3.02 × 2.56 cm. This USS finding necessitated a CT scan. The CT scan showed an enlarged hyperdense spleen, with multiple non-enhancing, well-defined, round isodense cysts of varying sizes demonstrated within [Figure 1a and b].

A diagnosis of splenomegaly with multiple splenic intraparenchymal cysts in an HbSS patient was made. Patient symptoms of recurrent bouts of severe left upper quadrant pain from cystic splenic enlargement responded poorly to opioid analgesics, necessitating the plan for an open splenectomy. She was immunized with pneumococcal, meningococcal, and H. influenza vaccines two weeks before surgery. She also had her hemoglobin optimized to 10 g/dl. Intra-operatively, a grossly enlarged spleen with multiple surface bosselations, about 16 cm in the polar axis, and weighing about 350 grams was found [Figure 2]. A total splenectomy was done, and the specimen was sent for histopathology. Postoperatively, she was placed on parenteral broad-spectrum antibiotics and analgesics. Post-operative monitoring, assessment of hemograms, and blood biochemistry were done. She was discharged home on oral analgesics and antibiotics after achieving a clinically stable state for seven days post-op. The patient reported compliance with penicillin prophylaxis during the follow-up visit. She also consented to commence hydroxyurea. Histopathology results from the removed spleen didn't show the presence of epithelial lining confirming the presence of splenic pseudocysts.

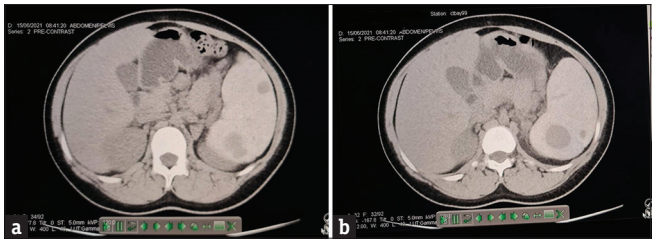

Figure 1: (a) CT scan of the abdomen showing enlarged hyperdense spleen with multiple intraparenchymal cysts. (b) CT scan of the abdomen showing enlarged hyperdense spleen with multiple intraparenchymal cysts

Discussion

In patients with SCD, there is a risk of disease-related complications involving the spleen. In early childhood, trapping sickled red cells in the spleen during vasoocclusion may cause moderate splenomegaly.[4] However, as the child grows, repeated splenic vasooclusion occurs, resulting in infarction and fibrosis. By the age of five years, most patients experience autosplenectomy.[1] In some patients, the spleen persists till adulthood possibly due to the presence of Hb SC disease,[2] co-inheritance of alpha or beta thalassemia,[2,5] high HbF levels,[6] or use of hydroxyurea.[7] The index patient presented with splenomegaly and was not taking hydroxyurea, but she however had borderline Hb F and HbA2 levels.

The persistence of splenomegaly into adulthood in SCD may lead to life-threatening complications like acute splenic sequestration, hypersplenism, splenic abscess, splenic infarction, and rarely splenic pseudocyst. Splenic cysts can be classified as parasitic or non-parasitic. Non-parasitic cysts are subdivided into primary (true-epidermoid cyst, epithelial or congenital) or secondary (pseudo) cysts. Splenic pseudocysts are most often preceded by splenic trauma or splenic infarction[8,9] and they can be differentiated from a true cyst by the absence of an epithelial lining on histology findings.[10] Splenic infarction is also common in young female adults less than 40 years with hematological disease.[11,12] The index patient who is an SCA teenager did not have a history of trauma but had a history of frequent (>6) episodes of painful vasooclusion per year before presentation at our facility. She did not have any infectious syndrome; thus, a parasitic cyst was ruled out. Furthermore, though the patient did not have the funds for tumor marker level (CA19-9), the CT scan findings did

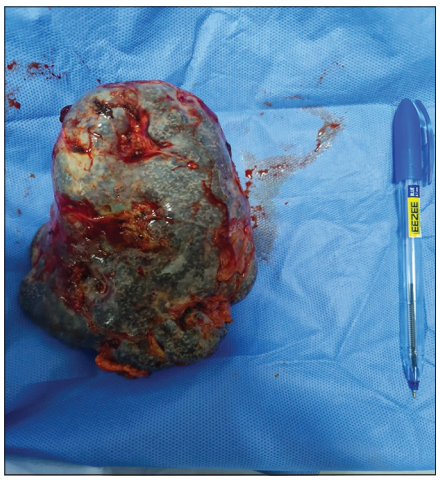

Figure 2: Surgically excised spleen specimen

not suggest a malignant cyst. In the index patient's histopathology report, there were no features of malignancy and no epithelial lining seen suggesting a diagnosis of a splenic pseudocyst.

Most patients with splenic pseudocysts are asymptomatic and thus do not require treatment except when symptoms develop. The presence of clinical symptoms like abdominal pain or early satiety and the presence of some peculiar characteristics of pseudocysts such as giant cysts, multiple cysts, and intraparenchymal cysts requires surgical management (partial or complete splenectomy). Splenic pseudocysts may rarely be complicated by infection or splenic rupture.[13] The index patient was presented with symptoms of pain unresponsive to analgesia and had multiple intraparenchymal cysts on a CT scan which necessitated surgical intervention by complete splenectomy.

Conclusion

Splenomegaly, although uncommon among older children with SCA, can in rare cases be associated with splenic pseudocyst precipitated by underlying infarction. The presence of a symptomatic splenic pseudocyst and the nature (size and position) of the cyst necessitates surgical intervention.

Patient perspective

The teenager was glad that her long-standing left abdominal swelling and pain had been resolved. She hopes for better days ahead.

Acknowledgment

We acknowledge the contribution of the nurses in the hematology department towards writing this report as well as the patient.

Informed consent

The patient gave informed consent to the case report and was happy to share her clinical data.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Brousse V, Elie C, Benkerrou M, Odièvre MH, Lesprit E, Bernaudin F, *et al*. Acute splenic sequestration crisis in sickle cell disease: Cohort study of 190 paediatric patients. Br J Haematol 2012;156:643-8.
2. Naymagon L, Pendurti G, Billett HH. Acute splenic sequestration srisis in adult sickle cell disease: A report of 16 Cases. Hemoglobin 2015;39:375-9.
3. Verma A, Yadav A, Sharma S, Saini D, Om P, Khoja H, *et al*. A rare splenic pseudocyst. J Surg Case Rep 2013;2013:rjt086. doi: 10.1093/jscr/rjt086.
4. Brousse V, Buffet P, Rees D. The spleen and sickle cell disease: The sick (led) spleen. Br J Haematol 2014;166:165-76.
5. Higgs DR, Aldridge BE, Lamb J, Clegg JB, Weatherall DJ, Hayes RJ, *et al*. The interaction of alpha-thalassemia and homozygous sickle-cell disease. N Engl J Med 1982;306:1441-6.
6. Al-Salem AH, Al-Aithan S, Bhamidipati P, Al-Jam'a A, Al Dabbous I. Sonographic assessment of spleen size in Saudi patients with sickle cell disease. Ann Saudi Med 1998;18:217-20.
7. Brousse V, Elie C, Benkerrou M, Odièvre MH, Lesprit E, Bernaudin F, *et al*. Acute splenic sequestration crisis in sickle cell disease: Cohort study of 190 paediatric patients. Br J Haematol 2012;156:643-8.
8. Chapman J, Helm TA, Kahwaji CI. Splenic infarcts. In: StatPearls. Treasure Island (FL): StatPearls Publishing; 2021. Available from: https://www.ncbi.nlm.nih.gov/books/NBK430902/. [Last assessed on 2022 Nov 10].
9. Daga G, Mittal V, Singh R J, Sood N. Epithelial cyst of the spleen. J Indian Assoc Pediatr Surg 2011;16:18-20.
10. Fowler RH. Nonparasitic benign cystic tumors of the spleen. Int Abstr Surg 1953;96:209-27.
11. Smoot RL, Truty MJ, Nagorney DM. Splenectomy for conditions other than trauma. In: Yeo CJ. editors. Shackelford's Surgery of the Alimentary Tract, 2 Volume Set. 8th ed. Philadelphia: Elsevier; 2019. p. 1635-53.
12. Pointer DT, Slakey DP. Cysts and tumors of the spleen. In: Yeo CJ, editors. Shackelford's Surgery of the Alimentary Tract, 2 Volume Set. 8th ed. Philadelphia: Elsevier; 2019. p. 1654-9.
13. Geraghty M, Khan IZ, Conlon KC. Large primary splenic cyst: A laparoscopic technique. J Minim Access Surg 2009;5:14-6.