Revista Colombiana de
Hematología y Oncología
Enero - Junio de 2025 • Volumen 12 - Número 1

CASO CLINICO

Acute splenic sequestration in an adult patient with sickle cell trait and beta-thalassemia

Secuestro esplénico agudo en un paciente adulto con rasgo falciforme y beta-talasemia

»Leonardo José Enciso Olivera [1]
»Daniel Martin Arsanios [2]
»Elías Quintero-Muñoz [2]
»Federico González Mahecha [2]
»Viviana Poveda-Hurtado [3]
»María Camila Delgado Cañaveral [2]
»Laura Perdomo [2]

[1] Hospital Universitario la Samaritana, Bogotá, Colombia.
[2] Universidad de La Sabana, Chía, Colombia.
[3] Universidad Tecnológica de Pereira, Pereira, Colombia.

Recibido el 26 de marzo de 2025; aceptado el 30 de abril de 2025

https//doi.org/10.51643/22562915.760

Abstract

Introduction: Splenic sequestration (SE) crisis with sickle cell disease is a rare disease in adults. Its occurrence in patients with sickle cell trait and beta-thalassemia is unusual, particularly during pregnancy. This report aims to describe the case of an adult patient with a mixed hemoglobinopathy (HbS/β-thalassemia) who presented with SE crisis as the first manifestation of the disease. **Clinical case:** A descriptive study case report of a 35-year-old female patient with a history of anemia during her second pregnancy, presented two years later referring fatigue, weight loss, and dyspnea. During hospital stay, laboratory tests, including hemoglobin electrophoresis, and imaging studies revealed arregenarative anemia secondary to hemolysis, a mixed phenotype for sickle cell disease with B thalassemia and hepatosplenomegaly. Despite transfusional support, hemolysis persisted, requiring emergency splenectomy which improved symptoms and decreased hemolytic activity.

*** Autor para correspondencia:** Leonardo Enciso. Hematólogo, Hospital Universitario La Samaritana.
Correo electrónico: leonardo.joenol@gmail.com
Doi: https//doi.org/10.51643/22562915.760

Histopathological analysis from splenic biopsy showed red pulp expansion with diminished white pulp. **Conclusions:** Splenic sequestration is a life-threatening complication primarily observed in children with sickle cell disease, but its occurrence in adults is exceptionally rare, particularly in patients with mixed hemoglobinopathy. In this case, splenic function was preserved due to high percentage of circulating HbA, which explained late presentation of the disease. In adult patients with severe anemia refractory to treatment, splenectomy should be considered to prevent severe complications and improve patient outcomes.

Keywords: beta-thalassemia; sickle cell disease; splenectomy; hemolytic anemia; hypersplenism.

Resumen

Introducción: La crisis de secuestro esplénico (SE) en la enfermedad de células falciformes es una complicación poco frecuente en adultos, su aparición en pacientes con rasgo falciforme y betatalasemia es inusual, particularmente durante el embarazo. Este estudio describe el caso de una paciente adulta con hemoglobinopatía mixta (HbS/β-talasemia) que presentó una crisis de SE como primera manifestación de la enfermedad. **Caso clínico:** Paciente femenina de 35 años con antecedente de anemia durante su segundo embarazo, quien acudió dos años después refiriendo fatiga, pérdida de peso y disnea. Durante su hospitalización, los exámenes de laboratorio, incluyendo electroforesis de hemoglobina, e imágenes revelaron anemia arregenerativa secundaria a hemólisis, fenotipo mixto de enfermedad de células falciformes con B talasemia y hepatoesplenomegalia. A pesar del soporte transfusional, la hemólisis persistió, requiriendo esplenectomía urgente, cuya histopatología mostró expansión de pulpa roja con pulpa blanca disminuida. Tras la esplenectomía, hubo disminución significativa de la actividad hemolítica con mejoría de los síntomas y posterior alta hospitalaria. **Conclusiones:** El secuestro esplénico es una complicación potencialmente mortal reportada principalmente en niños con enfermedad de células falciformes, pero su aparición en adultos es excepcionalmente rara, en particular en pacientes con hemoglobinopatía mixta. En este caso, la función esplénica se conservó debido al alto porcentaje de HbA circulante, que provocó la presentación tardía de la enfermedad. En pacientes adultos con anemia grave refractaria al tratamiento, se debe considerar la esplenectomía para prevenir complicaciones graves.

Palabras clave: beta-talasemia; enfermedad de células falciformes; esplenectomía; anemia hemolítica; hiperesplenismo.

Introduction

Acute splenic sequestration crisis typically occurs in the pediatric population with sickle cell disease and less frequently in other types of hemoglobinopathies [1]. Adults with hemoglobin SS (HbSS) rarely have enough splenic function and distensibility to precipitate an acute splenic sequestration crisis, as in early life the spleen loses functionality and decreases in size due to recurrent episodes of infarction and scarring [2-4]. We present the case of a 35-year-old female patient with sickle cell trait (HbS 23.7%) and beta-thalassemia (HbA2 3.4%), who presented with an anemic syndrome associated with splenic sequestration during the first trimester of her second pregnancy at the age of 32. This is

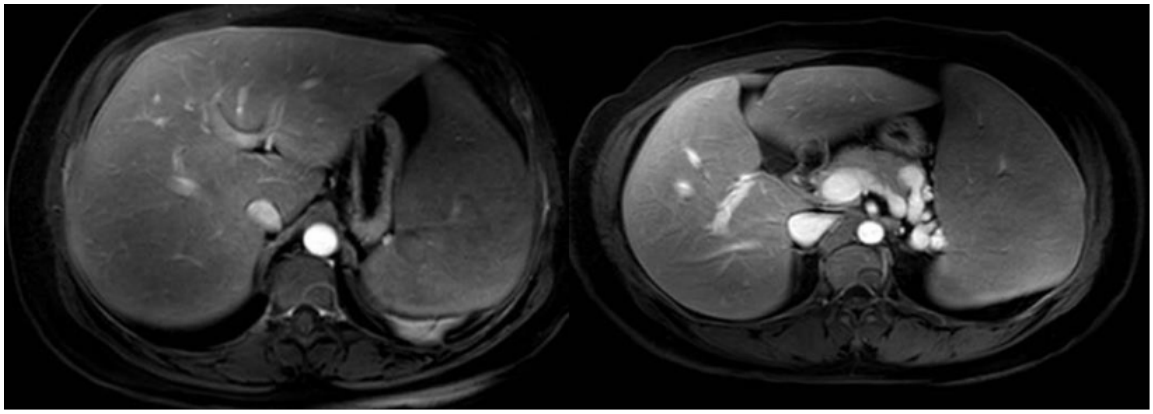
the 20th case—and the third pregnancy-related case—reported in the literature [2, 5].

Clinical case

A 35-year-old female patient with no relevant family or medical history was diagnosed during the first trimester of her second pregnancy with anemia associated with HbSB+ (beta-thalassemia trait combined with sickle cell trait). As she exhibited no significant clinical or paraclinical manifestations at the time, no specific treatment was required. Two years later, she developed asthenia, adynamia, hyporexia, weight loss (5 kg) and painless dyspnea on slight exertion. Upon presentation to the emergency department, her vital signs included a blood pressure of 114/79 mmHg and a heart rate of 98 bpm. Physical examination was notable for mucocutaneous pallor, scleral icterus, and hepatosplenomegaly; Paraclinical findings showed a hemoglobin level of 8.6 g/dL, hematocrit of 27%, MCV of 88.9 fL, and MCH of 28.5 pg. Lactate dehydrogenase (LDH) was elevated at 512 U/L. Indirect hyperbilirubinemia was present, with a total bilirubin of 3.68 mg/dL and an indirect fraction of 3.23 mg/dL. Reticulocyte count and reticulocyte production index indicated an aregenerative pattern (2.1% and <2, respectively). Haptoglobin was decreased (<4.96 mg/dL), while the deficiency profile—including serum iron, ferritin, vitamin B12, and folic acid—was within normal limits. Acute and chronic infectious processes were ruled out. Abdominal ultrasound, contrasted CT and abdominal MRI were performed, the latter reported in Figure 1. As complementary studies, flow cytometry and bone marrow biopsy were taken revealing erythroid hyperplasia. A new hemoglobin electrophoresis with HbA 71.7%, HbF 1.2%, HbS 23.7% and HbA2 3.4% confirmed a mixed phenotype for sickle cell disease and B thalassemia.

Figure 1.

Liver significantly enlarged with a length of 24 cm. The hepatic contours are well defined, no focal lesions are observed. The spleen is enlarged, measuring 20 x 16 x 9 cm, with an approximate volume of 500 cc. There is a diffuse decrease in the signal intensity of the hepatic and splenic parenchyma compared to other structures.



Given the high transfusion requirements—16 units of red blood cells within a month—ambulatory splenectomy was considered for symptom control and hemoglobin level regulation. However, the patient experienced persistent hemolysis, requiring massive transfusion within 24 hours and emergency laparoscopic splenectomy. The excised spleen measured 14 × 10 × 8 cm and weighed 1,370 g. Microscopic examination showed marked expansion of the red pulp, with the presence of erythroid and myeloid lineage cells, as well as occasional cells resembling megakaryocytes. The white pulp was diminished and composed predominantly of reactive lymphocytes. No areas of necrosis, granulomas or metastatic lesions were identified. After splenectomy, there was a very significant decrease in hemolytic activity, an increase in hemoglobin levels and symptom improvement, leading to hospital discharge 13 days after the procedure without new transfusion requirements.

Discussion

Splenic sequestration involves the abrupt drop in hemoglobin levels due to entrapment in the spleen and is associated with high morbidity and mortality in children with sickle cell disease. The characteristics of the splenic microcirculation promote the polymerization of hemoglobin S (HbS) as it passes through this organ, leading to the thrombotic and ischemic complications observed in these patients [6].

Also, splenic sequestration is characterized by an acute increase in splenic volume, accompanied by a decrease in hemoglobin and often platelet levels, followed by abdominal pain and signs of hypovolemia. This clinical presentation has typically been described in patients with sickle cell disease (HbSS and HbSC)[7]. However, only 19 cases of splenic sequestration associated with sickle cell disease and beta thalassemia (Hb S-β thalassemia) have been reported.

The occurrence of splenic sequestration in adults with sickle cell disease is unusual, as functional splenic tissue is typically lost due to fibrosis developed during childhood [4]. In individuals with Hb S-β^0 thalassemia, a similar process occurs. Due to the low percentage of HbA, the clinical presentation resembles that of patients with HbSS sickle cell anemia, who typically lack a functional spleen by adulthood as a result of autosplenectomy [8].

This could explain the absence of case reports of splenic sequestration in older individuals with in this population. One of the pathophysiological factors that allow the late presentation of splenic sequestration in patients with Hb S-β thalassemia is the high percentage of circulating HbA [1]. In this condition, the percentage of HbA ranges between 20% and 30%, so splenic damage during infancy is less pronounced, allowing late presentation of [1]. In our patient's scenario, HbA percentage was 71.7%, which is higher than the average reported and may explain the absence of symptoms until adulthood (Table 1).

Table 1.

Reported cases of patients with HbS / β + -thalassemia and acute splenic sequestration crisis.

Author, year	N°	Sex	Age	% HbA	Comorbidity	Spleen weight	Splenic infarction	Splenectomy	Outcome
Current case	20	F	35	71.7	None	1370 g	No	Yes	Alive
Esterson, 2021[14]	19	M	32	NR	None	NR	NR	Yes	Alive
Edo-Osagie, 2019[1]	18	F	20	48.0	None	NR	Yes	Yes	Alive
Di Vincenzo, 2018[16]	17	M	53	NR	Cholelithiasis	NR	NR	Yes	Alive
Naymagon, 2015[4]	16	M	24	NR	NR	NA	NA	No	Alive
	15	F	51	NR	NR	NA	Yes	No	Alive
	14	M	34	NR	NR	NR	Yes	Yes	Alive
	13	F	19	NR	NR	NR	Yes	Yes	Alive
	12	F	26	NR	NR	NR	Yes	Yes	Alive
Noreldeen, 2008[5]	11	F	23	NR	None	NA	Yes	No	Alive
Tsikrikas, 2008[13]	10	M	21	NR	None	NA	Yes	Yes	Alive
Koduri, 2006[7]	9	M	22	29.0	None	795 g	No	Yes	Alive
Aslam, 2005[8]	8	F	34	NR	Cholelithiasis	NA	No	No	Dead
Hutchins, 2001[15]	7	M	29	----	----	NA	----	No	Dead
Sheth, 2000[6]	6	NR	31	NR	NR	NA	NR	Unknown	NR
Van Rhee, 19913	5	M	32	19.0	None	750 g	No	No	Dead
Berry, 1991[9]	4	M	22	NR	None	950 g	Yes	Yes	Alive
Roshkow, 199011	3	M	31	NR	None	880 g	Yes	Yes	Alive
Solanki, 1986[2]	2	F	30	NR	None	NA	Yes	Yes	Alive
	1	M	44	NR	None	NA	Yes	No	Alive

Note: F = Female; M = Male; NR = Not Reported; NA = Not Applicable

In most cases it is not possible to identify a clear etiology of splenic sequestration, some literature has mentioned changes in altitude and systemic infections could trigger this complication [9,10]. In Solanki et. Al [2,] the first case of splenic sequestration related to pregnancy is described; however, the cause of the decompensation could not be identified. Similarly, in our case, etiology could not be determined despite conducting all the relevant studies. Of the 19 reported cases of splenic sequestration in patients with sickle cell disease and B thalassemia, 11 were men, their age ranged from 19 to 53 years, with no relevant comorbidities reported and 15% died because of this complication.

In splenic sequestration, certain imaging features can be observed. On computed tomography, multiple peripheral or large diffuse areas of low attenuation within the splenic tissue, along with mild to moderate splenomegaly, may be evident. Once the acute process resolves, these low-attenuation areas should no longer be present as this may be related to splenic infarction [11].

On abdominal MRI, the spleen typically exhibits an abnormally low signal intensity due to iron deposition. A shortening of both T1 and T2 relaxation times is also observed. Splenic scintigraphy with Tc-99m is useful for assessing splenic function, as it can demonstrate functional asplenia when there is no Tc-99m uptake, even in the presence of a palpable spleen. In cases of splenic sequestration, an increased uptake of the same tracer (Tc-99m) may be observed. Likewise, it has been shown that spleen size, as assessed by ultrasound, does not correlate with its functional status when compared to scintigraphy findings [12].

Acute treatment consists on transfusional support expecting to increase HbA levels; however, splenectomy should be considered in patients with recurrent acute splenic sequestration crises or in those who do not respond adequately to blood product administration [13]. Acute crises are recurrent in 50% of cases and are associated with mortality rates of 10-15%, therefore, elective splenectomy is recommended after the resolution of the crisis [14]. Of the 19 reported cases, 12 required elective splenectomy. All of them survived and presented a favorable clinical response. Our case required emergency splenectomy given the persistence of severe hemolysis despite multiple units of red blood cells transfused [3,8,15]. There were no early complications, and both the clinical response and hemoglobin levels observed at day 13 were consistent with expected outcomes reported in other documented cases.

This patient's heterozygous HbS/β-thalassemia pattern highlights the wide variety of phenotypes that can occur in hemoglobinopathies. Acute splenic sequestration crisis is not common as an initial manifestation of the disease, so it is not the first diagnostic consideration in patients with no pathological history; however, given the severity and clinical implications it should be considered to ensure an early diagnosis and treatment and prevent fatal outcomes.

Consent

Informed consent was obtained from the patient.

Funding sources

This work did not receive any grant from public, commercial or non-for-profit funding agencies.

Conflict of interests

There are no conflicts of interest to declare by any of the authors of this study.

Contributions

Contributors played a substantial role in conception, design, acquisition, analysis, interpretation, writing, and critical review of the manuscript.

Author's Bio

Leonardo José Enciso Olivera, médico especializado en medicina interna y hematología de la Universidad Nacional de Colombia, máster en ciencias biológicas de la Pontificia Universidad Javeriana.

Daniel Martin Arsanios, médico internista de la Universidad de La Sabana, actualmente cursando segundo año del programa de nefrología en la Pontificia Universidad Javeriana.

Elías Quintero Muñoz, médico especialista en medicina interna de la Universidad de La Sabana actualmente en el programa de reumatología de la Universidad de La Sabana.

Federico González Mahecha, médico especialista en medicina interna de la Universidad de la Sabana.

Viviana Poveda Hurtado, médico general de la Universidad Tecnológica de Pereira actualmente cursando tercer año del programa medicina interna en la Universidad de La Sabana.

María Camila Delgado Cañaveral, médico general de la Universidad de La Sabana

Laura Perdomo, médico general de la Universidad de La Sabana

References

1. Edo-Osagie E, Enofe I, Hakeem H, Rai M, Adomako E, Tismenetsky M, Janosky M. Splenic sequestration crisis as an index manifestation of heterozygous hemoglobinopathy in an adult. Oxf Med Case Reports. [Internet] 2019 12;2019(7):omz069. Available from: https://doi.org/10.1093/omcr/omz069.

2. Solanki DL, Kletter GG, Castro O. Acute splenic sequestration crises in adults with sickle cell disease. Am J Med. [Internet] 1986;80(5):985-90. Available from: https://doi.org/10.1016/0002-9343(86)90604-4.

3. Van Rhee F, Balsitis M, French EA. Fatal splenic sequestration crisis in adult sickle cell-beta thalassaemia. Postgrad Med J. [Internet] 1991;67(792):907-8. Available from: https://doi.org/10.1136/pgmj.67.792.907.

4. Naymagon L, Pendurti G, Billett HH. Acute Splenic Sequestration Crisis in Adult Sickle Cell Disease: A Report of 16 Cases. Hemoglobin. [Internet] 2015;39(6):375-9. Available from: https://doi.org/10.3109/03630269.2015.1070131.

5. Noreldeen SA, Oppenheimer C, Chapman C, Pavord S. Postpartum acute splenic sequestration in sickle cell disease. J Obstet Gynaecol. [Internet] 2008;28(4):440-1. Available from: https://doi.org/10.1080/01443610802064788.

6. Sheth S, Ruzal-Shapiro C, Piomelli S, Berdon WE. CT imaging of splenic sequestration in sickle cell disease. Pediatr Radiol. [Internet] 2000;30(12):830-3. Available from: https://doi.org/10.1007/s002470000341.

7. Koduri PR, Kovarik P. Acute splenic sequestration crisis in an adult with sickle beta-thalassemia. Ann Hematol. [Internet] 2006;85(9):633-5. Available from: https://doi.org/10.1007/s00277-006-0119-1.

8. Aslam AF, Aslam AK, Dipillo F. Fatal splenic sequestration crisis with multiorgan failure in an adult woman with sickle cell-beta+ thalassemia. Am J Med Sci. [Internet] 2005;329(3):141-3. Available from: https://doi.org/10.1097/00000441-200503000-00004.

9. Berry RA, Odumakinde EA, Lewis JP. Massive splenic infarction in doubly abnormal heterozygous sickling disorders. A new complication of acute splenic sequestration syndrome. West J Med. [Internet] 1991;155(5):531-2. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC1003162/.

10. Koduri PR, Nathan S. Acute splenic sequestration crisis in adults with hemoglobin S-C disease: a report of nine cases. Ann Hematol. [Internet] 2006;85(4):239-43. Available from: https://doi.org/10.1007/s00277-005-0056-2.

11. Roshkow JE, Sanders LM. Acute splenic sequestration crisis in two adults with sickle cell disease: US, CT, and MR imaging findings. Radiology. [Internet] 1990;177(3):723-5. Available from: https://doi.org/10.1148/radiology.177.3.2217790.

12. Dinan D, Epelman M, Guimaraes CV, Donnelly LF, Nagasubramanian R, Chauvin NA. The current state of imaging pediatric hemoglobinopathies. Semin Ultrasound CT MR. [Internet] 2013;34(6):493-515. Available from: https://doi.org/10.1053/j.sult.2013.07.002.

13. Tsikrikas S, Sialevris K, Tsochatzis E, Deutsch M, Vassilopoulos D, Vgontza N, Archimandritis AJ. Acute splenic sequestration crisis (ASSC) in an adult patient with beta-thalassemia sickle cell disease: a life-threatening complication. Ann Hematol. [Internet] 2008;87(6):499-500. Available from: https://doi.org/10.1007/s00277-008-0426-3.

14. Esterson YB, Sheth S, Kawamoto S. Splenic sequestration in the adult: cross sectional imaging appearance of an uncommon diagnosis. Clin Imaging. [Internet] 2021;69:369-373. Available from: https://doi.org/10.1016/j.clinimag.2020.09.012.

15. Hutchins KD, Ballas SK, Phatak D, Natarajan GA. Sudden unexpected death in a patient with splenic sequestration and sickle cell-beta+-thalassemia syndrome. J Forensic Sci. [Internet] 2001;46(2):412–4. Available from: https://doi.org/10.1520/JFS15038J.

16. Di Vincenzo A, Marson P, Puato M. Acute Splenic Sequestration Crisis After Red Blood Cell Exchange for Acute Chest Syndrome in an Adult With Sickle β-Thalassemia: What Went Wrong? Ther Apher Dial. [Internet] 2018;22(2):207-208. Available from: https://doi.org/10.1111/1744-9987.12643.