

## Acute Splenic Sequestration: A Rare and Severe Complication of Sickle Cell Anemia in Adults: About a Case Report

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### Abstract

### Case Report

Acute splenic sequestration is a well-known complication of sickle cell disorders, marked by a sudden drop in hemoglobin levels and significant, painless splenomegaly. This condition is typically observed in children with homozygous sickle cell disease. However, it is rarely reported in adults with heterozygous sickle cell conditions. We describe the case of a 28-year-old patient with a history of hemoglobin SC disease who suffered an acute splenic sequestration crisis. We review the CT characteristics of splenic sequestration, including splenic enlargement and an irregular peripheral rim of hypoenhancing or hypoechoic tissue, and discuss differential diagnoses. While acute splenic sequestration is predominantly a severe complication in children, timely diagnosis and treatment particularly red blood cell transfusions can lead to full recovery.

**Keywords:** Splenic Sequestration, Sickle Cell Anemia, Splenectomy, Red Blood Cells Transfusion.

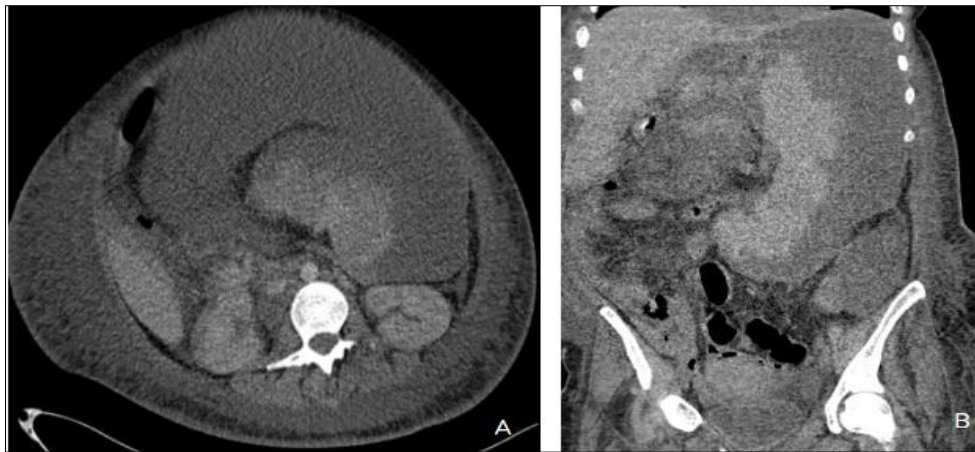
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## INTRODUCTION

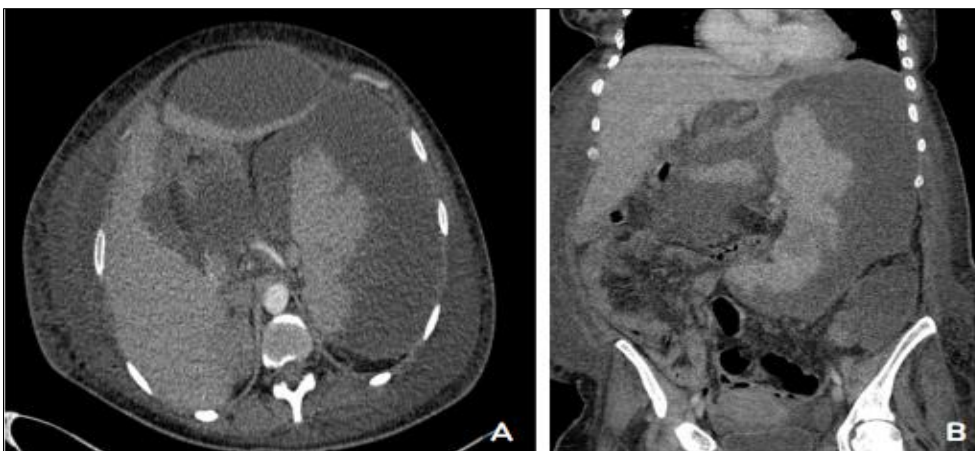
Acute splenic sequestration crisis (ASSC) is a severe complication of sickle cell disease which can be fatal. It presents with a new onset of splenomegaly (length > 2 cm and gauges as positive) and at least 2.0 g/dL decrease in Hb levels due to spleen encapsulation of blood [1]. This happens through a blockage of red blood cells in the spleen, leading to circulatory collapse and without treatment to death. ASSC presents most frequently in children, but it is an extremely rare disease with few cases being reported in adults. The diagnosis may be largely clinical with few imaging studies needed. Non-specific presenting symptoms in patients include pallor, lethargy, abdominal fullness and pain, tachycardia and tachypnoea, often mimicking other acute abdominal conditions [2]. Without an understanding of the patient's baseline physical examination and hemoglobin levels, diagnosing ASSC can be challenging. Accurate diagnosis is crucial to avoid unnecessary surgical interventions. This review aims to inform general physicians and radiologists about the diverse presentations of splenic sequestration on CT imaging [3].

## CASE PRESENTATION

A 28-year-old woman with a history of hemoglobin SC disease presented to our institution with lower back pain. In the emergency department, the patient was noted to be tachycardic at 113 beats per minute and febrile with a temperature of 38°C. Physical examination revealed splenomegaly, though other findings were unremarkable. Laboratory results were significant for low hemoglobin level of 8 g/dL, a low mean corpuscular volume of 77.2 fL, and an elevated reticulocyte count of 3.5%, attributed to anemia and hemolysis related to his hemoglobinopathy. Abdominal CT imaging showed marked splenomegaly with the spleen measuring 26 cm in craniocaudal dimension with increasing irregular hypoenhancement along the periphery of the splenic parenchyma, associated to a peritoneal effusion evident throughout all compartments (Fig. 1 et 2). The patient's condition deteriorated over the following days. Repeat blood work revealed a significant drop in hemoglobin (from 8 g/dL to 4 g/dL), elevated LDH, low haptoglobin, and increased indirect bilirubin. Despite multiple transfusions of packed red blood cells, platelets, and fresh frozen plasma, and aggressive supportive care, the patient did not survive.



**Figure 1: Non-enhanced axial (A) and coronal (B) CT scan showing mild splenomegaly and multiple low attenuation areas in the periphery of the spleen.**



**Figure 2: Abdominal CT: axial (A) and coronal (B) section at arterial and venous phase demonstrating a splenic enlargement with increasing irregular hypoenhancement along the periphery of the splenic parenchyma.**

## DISCUSSION

Splenic sequestration is a rare complication in adults with sickle cell disease, with only about forty cases documented [4]. This rarity is attributed to the spleen's anatomy, which includes areas of low oxygen pressure that promote red blood cell sickling and lead to repeated infarcts from early childhood. Diagnosing splenic sequestration can be challenging, especially in adults where it is less common, and the diagnosis may not be immediately apparent in acute settings [5].

Clinical manifestations typically include sudden weakness, severe pallor, tachycardia, and abdominal fullness. Aplastic anemia was ruled out because the full blood count did not show pancytopenia, and there were no hemorrhagic symptoms such as petechiae, ecchymoses, or gastrointestinal bleeding related to thrombocytopenia [6].

The symptomatology of splenic sequestration can be misleading. For instance, a study by Naymagon *et al.*, of 16 splenic sequestration cases found that only a minority of patients exhibited classic left-sided abdominal pain; more commonly, patients presented with symptoms similar to a typical vaso-occlusive pain

crisis [7]. Thus, patients with splenic sequestration might be referred for diagnostic imaging, and radiologists must be prepared to recognize the characteristic features of this condition, which may not be immediately considered by referring clinicians in adults.

This variability in presentation requires a high level of suspicion and appropriate imaging. Splenic sequestration can be identified using various diagnostic imaging modalities, including CT scans, ultrasound, and technetium-99m sulfur colloid liver-spleen scans [8-12]. CT imaging typically reveals an enlarged spleen with a thick, irregular peripheral rim of low attenuation, indicative of infarcts and hemorrhage. Alternatively, CT may show more diffuse areas of hypoattenuation within an enlarged spleen. Ultrasound similarly demonstrates an irregular peripheral hypoechoic rim in an enlarged spleen, with both modalities confirming the patency of the splenic artery and vein [9, 10].

On cross-sectional imaging, differential diagnosis for splenic sequestration include subcapsular splenic hematoma and splenic infarction [11]. A subcapsular splenic hematoma usually appears as a lenticular or crescentic perisplenic collection that

smoothly flattens the splenic contour, contrasting with the irregular peripheral appearance seen in sequestration. Splenic infarction typically presents as one or more peripheral, wedge-shaped hypoattenuated areas, whereas sequestration shows more diffuse peripheral hypoattenuation. Additionally, normal homogeneous attenuation of the spleen often returns following sequestration recovery, while wedge-shaped hypoattenuation from splenic infarction may persist, scar, or calcify on follow-up imaging [12].

The prognosis can be severe, with the risk of hypovolemic shock potentially leading to death. Nevertheless, the condition often resolves spontaneously. In our case, it was complicated by acute thoracic syndrome (characterized by a new radiological infiltrate and symptoms such as cough, fever, dyspnea, expectoration, chest pain, and abnormal auscultatory findings), which ultimately had a favorable outcome [13, 14].

The management of splenic sequestration is still debated. Treatment generally involves supportive care, including hydration and pain management. Blood transfusions may be required to address severe anemia and stabilize the patient (Power-Hays, 2024) [15]. Hydroxyurea, a disease-modifying therapy, has been shown to reduce the frequency of vaso-occlusive crises and may assist in managing splenic sequestration by decreasing the overall sickling tendency of red blood cells. Surgical options, such as splenectomy or partial splenic embolization (PSE), may also be considered. The choice of treatment should be individualized based on the patient's clinical status and history of splenic crises, highlighting the importance of early recognition and intervention to improve outcomes [16].

## CONCLUSION

In conclusion, splenic sequestration is a critical condition that requires prompt recognition and management, particularly in pediatric patients with sickle cell disease. The role of imaging, especially CT, is indispensable in diagnosing and differentiating this condition from other abdominal emergencies. Ongoing education for healthcare providers and parents is essential to improve outcomes and reduce the risks associated with this potentially life-threatening complication.

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