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| RESEARCH ARTICLE

Sepsis, Sickle Cell, and The Surgical Abdomen: A Triple Threat in a Young Adult

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| ABSTRACT

Anchoring bias poses a major obstacle in the timely diagnosis of surgical abdomen in patients with sickle cell disease (SCD). Clinicians must remain aware that not all pain experienced by patients with SCD is necessarily due to vaso-occlusive crisis (VOC). A high index of suspicion is needed, especially when pain becomes atypical, localized, or unresponsive to standard VOC management. This case illustrates the consequences of diagnostic delay in a 24-year-old Saudi male with known SCD who initially presented with what was presumed to be VOC. As his condition deteriorated into septic shock—with persistent fever, leukocytosis, and hypotension—broad-spectrum antibiotics were initiated, and an abdominal CT was finally performed. Imaging revealed acute appendicitis complicated by perforation and abscess formation. He underwent emergency exploratory laparotomy, which confirmed a perforated appendix with purulent peritonitis. Appendectomy, peritoneal lavage, and drain placement were performed, followed by targeted antibiotic therapy in the intensive care unit. The patient gradually improved and was later discharged in stable condition.

| KEYWORDS

Sickle Cell Disease, Acute Abdomen, Surgical Abdomen, Sepsis, Vaso-occlusive Crisis

| ARTICLE INFORMATION

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Introduction

Sickle cell disease (SCD) is a hereditary hemoglobinopathy in which a point mutation in the β-globin gene produces sickle hemoglobin (HbS) and causes chronic hemolysis and vaso-occlusion. SCD comprises a spectrum of related disorders, including homozygous sickle cell anemia, HbSC disease, and sickle–β thalassemia, that distort erythrocyte shape and impair blood flow [1][2]. Patients with SCD typically have chronic anemia and episodic "pain crises" due to microvascular infarction, along with

systemic features such as jaundice and immune dysfunction [1][2]. In SCD, the classic Glu6Val mutation in the β-globin chain causes HbS polymerization under deoxygenated conditions, leading to rigid, sickled red cells [1]. The most severe form, sickle cell anemia (HbSS), results from homozygosity for this mutation, whereas compound heterozygotes (e.g., HbSC or HbS–β^0 thalassemia) have variably milder phenotypes [1]. Overall, SCD afflicts millions worldwide, causing chronic organ damage and early mortality without intervention. Globally, the number of people living with SCD rose from about 5.5 million in 2000 to 7.74 million in 2021 [3], and an estimated half a million infants are born each year with SCD, primarily in sub-Saharan Africa [3]. The epidemiology of SCD is highly regional. It is most prevalent in malaria-endemic regions, notably sub-Saharan Africa, India, and the Arabian Gulf [3][4]. In the Middle East, SCD prevalence is especially high in the Arabian Peninsula. For instance, as per different literature and statistical papers, nearly 2.3% of the whole population of Saudi Arabia has SCD, with carrier status found in approximately 3.1% [2]. According to registry and screening reports from 2000 to 2021, the three countries; Bahrain, Oman, and Saudi Arabia have SCD prevalence rates exceeding 0.25% (>250/100,000), whereas Kuwait, Lebanon, Turkey, and the UAE report rates under 0.05% (<50/100,000). Additionally, there are a lot of newborn screening and premarital testing programs that are conducted in the Gulf region that confirmed these high rates; one cohort study in a Bahraini school (1999–2008) reported that 1.1% of students are affected by SCD [4]. Overall, these statistics highlight and confirm that SCD is extremely frequent and burdensome in the Middle East, varying substantially by region. Abdominal pain is a frequent and challenging complaint in SCD. Pain crises can occur in almost any vascular bed, including the mesenteric or splenic circulation, so diffuse or focal abdominal pain is common. In practice, abdominal pain is a recognized component of vaso-occlusive crises (VOC), but other causes are also common. Chronic hemolysis predisposes to pigment gallstones and cholelithiasis; a recent meta-analysis found that roughly 25% of patients with SCD have gallstones [5]. Cholecystitis, choledocholithiasis, and liver infarction are therefore important considerations. Splenic complications (acute sequestration, splenic infarcts, or abscess) were historically common, though autosplenectomy often limits this in adults. Other causes of abdominal pain in SCD include acute appendicitis, urinary tract infections, renal papillary necrosis, peptic ulcer disease, and intestinal obstruction (e.g., from adhesions or volvulus) [5][6]. In short, SCD patients may experience a broad differential for acute abdomen that overlaps both medical (VOC-related ischemia) and surgical causes. Distinguishing a simple VOC from a true surgical abdomen is notoriously difficult. Abdominal VOCs – thought to arise from mesenteric or serosal vessel infarction – often present with severe, colicky pain that may localize variably. Such pain can be accompanied by nausea, vomiting, and mild peritoneal signs. Critically, however, mesenteric VOCs can mimic any intra-abdominal surgical condition. As one case series notes, "pain emanating from abdominal organs will present with the acute abdomen" – tender, guarded, and rigid – yet it is "easy to misinterpret such a presentation as 'ordinary sickle cell crisis.'" [6]. Similarly, abdominal VOCs are "clinically indistinguishable from acute abdominal pain from other causes and can pose a diagnostic dilemma" [7]. In effect, symptoms of VOC (diffuse severe pain, low-grade fever, and leukocytosis) can obscure coexisting pathology like appendicitis or perforation. This diagnostic overshadowing can delay surgical recognition. For example, reports describe SCD patients with acute appendicitis initially treated as a pain crisis until repeat exams and imaging revealed the surgical disease [6][7]. Because abdominal crises in SCD are common, clinicians must watch for "red flag" findings that suggest a surgical process. Persistent focal peritonism (rigidity or rebound), progressive tachycardia or hypotension, or escalating leukocytosis out of proportion to a patient's usual crisis baseline should raise concern. High spiking fevers or lactic acidosis may likewise signal bowel ischemia or infection rather than uncomplicated VOC. Imaging (ultrasound or CT) can be invaluable: for instance, identifying gallstones or free fluid can prompt surgical referrals. In practice, one must maintain a high index of suspicion. As experts emphasize, "the need for an increased index of suspicion and repeated clinical examination" is critical in SCD patients with abdominal pain [6]. Serial exams and early surgical consultation are advised if the clinical picture is unclear. Another complicating factor is the sepsis risk inherent in SCD. Functional asplenia and immune dysregulation render SCD patients more susceptible to invasive bacterial infections [2]. In fact, one review notes that "sepsis is an ever-present problem in sickle cell disease and most of the time may be the precipitating factor in a crisis" [6]. Common pathogens include encapsulated organisms (e.g., Salmonella, S. pneumoniae) and gram-negative bacilli. In practice, systemic infection in SCD can itself cause severe abdominal pain (e.g., Salmonella osteomyelitis may present with abdominal pain from hip or psoas involvement) and further blur the clinical picture. Thus, an SCD patient with fever and abdominal pain might be suffering VOC, intra-abdominal sepsis, or both. The overlap of sepsis, VOC, and possible surgical disease creates a true "triple threat" scenario. This challenge exemplifies the problem of diagnostic overshadowing in SCD care. Diagnostic overshadowing refers to the tendency for common SCD-related symptoms (e.g., pain crises, anemia) to dominate the clinical picture and mask coexisting conditions [8]. Put simply, a clinician may ascribe new or severe symptoms to "just another sickle crisis," inadvertently overlooking a superimposed surgical or infectious pathology [8]. This cognitive bias can lead to dangerous delays. For example, recurrent case reports describe delayed appendectomies or unrecognized bowel perforations in SCD patients whose acute pain was initially attributed to VOC. Maintaining vigilance against this bias is crucial, especially when patients deteriorate or fail to improve with standard crisis management. The case presented here – a young adult with SCD who developed sepsis alongside an acute surgical abdomen – is noteworthy for precisely these reasons. It highlights the intersection of sepsis risk, VOC pain, and surgical emergency in one patient. Such cases are relatively rare in the literature but offer important lessons. They underscore that even in a well-known disease, clinicians must remain alert to atypical features and avoid anchoring on VOC as the sole diagnosis. The current report provides an instructive example of the diagnostic dilemma when VOC symptoms eclipse an emergent surgical

condition, and it illustrates strategies (e.g., serial exams, targeted labs/imaging, and broad differential diagnosis) to disentangle these overlapping crises.

Case Presentation

Patient's history and Physical Examination

Our case report delineates a 24-year-old Saudi male, known to have sickle cell disease (SCD), who typically requires hospitalization two to three times annually for episodes of vaso-occlusive crisis (VOC). He has no documented history of acute chest syndrome (ACS), avascular necrosis, or ischemic cerebrovascular events. A single transfusion had been administered previously in the context of a hemolytic crisis following an upper respiratory tract infection; however, there is no history of exchange transfusion. He has no prior surgical history. The patient commenced hydroxyurea therapy but demonstrated poor compliance, ultimately discontinuing the medication several years ago. He presented to our emergency department with a chief complaint of progressively worsening, severe, generalized pain—primarily affecting the lower back, shoulders, hands, and abdomen—of a sickling nature and unresponsive to home-administered analgesia. There was no vomiting at the outset, although the patient reported anorexia and mild nausea. Bowel habits remained unchanged. Over time, the abdominal pain gradually localized to the right lower quadrant, though this was initially overlooked due to the diffuse character of the overall pain. At presentation, the abdominal discomfort was attributed to his VOC. On physical examination, he was afebrile and normotensive, with a heart rate of 90 beats per minute and an oxygen saturation of 96% on ambient air. Cardiovascular and respiratory examinations were unremarkable. Abdominal examination revealed no tenderness, guarding, or rigidity Subsequently, however, the patient developed persistent fever and tachycardia. This clinical deterioration progressed to hypotension, at which point the abdominal examination revealed rigidity and rebound tenderness localized to the right lower quadrant.

Investigations

Relevant laboratory investigations were conducted as detailed in Table 1. Initially, no imaging modalities were employed. However, as the clinical picture evolved, an abdominal ultrasound was subsequently performed. The study was limited by overlying bowel gas, rendering the findings inconclusive. Nonetheless, no sonographic evidence of gallbladder wall thickening, cholelithiasis, or pericholecystic fluid was noted, and the biliary tree appeared unremarkable. No free intra-abdominal fluid was appreciated at that time. As the patient's condition progressed and signs of impending sepsis emerged, a contrast-enhanced CT scan of the abdomen was performed. The imaging revealed features highly concerning for a perforated appendix, associated with a localized abscess formation.

Test	Result	Normal Range
Hemoglobin	8.0	13-17 g\dL
WBC	$18x10^9$	$4.0\text{-}10x10^9$\L
Platelets	$490x10^9$	$150\text{-}450x10^9$\L
Hematocrit Value	26%	40-50%
Reticulocyte count	4.7%	0.5-1.5%
LDH	700	140-280 U\L
Total bilirubin	4.0	0.2-1.2 mg\dL
Direct bilirubin	0.9	0.1-0.3 mg\dL
Sodium	134	135-145 mmol\L
Potassium	4.6	3.5-5.0 mmol\L
Creatinine	1.4	0.7-1.2 mg\dL
ALT	73	<40 U\L
AST	65	<40 U\L
Procalcitonin	6 ng\mL	<0.5 ng\mL

Table 1: results of relevant laboratory investigations.

Management course

Upon admission, maintenance intravenous fluids were initiated in the form of 2.5 to 3 liters per day of normal saline. Intravenous morphine was administered at a dose of 4 mg every six hours for analgesia. On day 2 post-admission, the patient's clinical condition deteriorated, presenting with a temperature of 39°C, heart rate of 135 beats per minute, blood pressure of 90/55 mmHg, and oxygen saturation of 92% on room air. Laboratory findings revealed leukocytosis with a white blood cell count of 20 × 10^9/L and an elevated serum lactate of 3.5 mmol/L. A full septic workup was promptly initiated. Empiric intravenous antibiotic

therapy commenced with piperacillin-tazobactam (4.5 g every 6 hours) and aggressive fluid resuscitation was carried out. Intravenous morphine was carefully titrated in response to pain and clinical status. In light of the evolving abdominal findings and CT imaging suggestive of perforated appendicitis with abscess formation, the patient was taken urgently to the operating theatre for exploratory laparotomy. Intraoperatively, a perforated appendix with purulent peritonitis was confirmed. Appendectomy, extensive peritoneal lavage, and placement of intra-abdominal drains were performed. Postoperatively, the patient was admitted to the intensive care unit, where intravenous antibiotics were continued for 7 days. Once Escherichia coli was isolated from blood cultures and demonstrated sensitivity to ceftriaxone, antimicrobial therapy was de-escalated accordingly to ceftriaxone 2 g once daily. With gradual clinical improvement, the patient was transitioned to oral antibiotics and discharged home on amoxicillin-clavulanate 875/125 mg taken twice daily for completion of therapy.

Discussion

This case serves as a compelling illustration of anchoring bias as a diagnostic pitfall—particularly in patients with sickle cell disease (SCD), where physicians frequently attribute abdominal or musculoskeletal pain solely to vaso-occlusive crises (VOC). Such cognitive bias may delay the identification of life-threatening conditions, including surgical causes of acute abdomen. It is estimated that up to 30% of abdominal crises in SCD patients are attributable to surgical pathologies, with the remainder stemming from non-surgical causes [6]. This underscores the importance of revisiting the differential diagnosis when pain deviates from its typical VOC pattern—namely, when it becomes localized, progressive, or peritonitic in nature. While VOC-related pain is characteristically diffuse, migratory, and generally responsive to opioids, pain due to surgical pathology tends to be focal, persistent, and associated with peritoneal signs [7]. One study reports that appendicitis is initially missed in approximately 50% of SCD patients, often leading to severe complications such as perforation and generalized peritonitis [6]. Laboratory interpretation in such scenarios poses additional challenges. Leukocytosis and thrombocytosis are often chronic findings in SCD, secondary to baseline inflammation, and can obscure signs of acute infection [1]. This may contribute to diagnostic delays in conditions such as osteomyelitis, cholecystitis, or intra-abdominal sepsis [1]. Given these complexities, clinicians should maintain a low threshold for cross-sectional imaging, particularly abdominal CT, in any SCD patient presenting with concerning abdominal symptoms [7]. CT imaging offers a sensitivity exceeding 95% for detecting appendicitis, in contrast to abdominal ultrasound, which offers sensitivity ranging from 65% to 85%, and may be limited by patient habitus and bowel gas interference [7]. Moreover, due to functional asplenia resulting from repeated splenic infarctions, these patients are predisposed to Overwhelming Post-Splenectomy Infection (OPSI)-like syndromes, which can result in mortality rates exceeding 35% in cases where surgical abdomen is misdiagnosed or diagnosed late [8]. Perioperative complications also warrant particular attention. The incidence of acute chest syndrome (ACS) post-abdominal surgery in SCD patients is approximately 20%, often precipitated by perioperative hypoxia, hypothermia, and fluid shifts [8]. The American Society of Hematology (ASH) 2020 Guidelines for Sickle Cell Disease recommend maintaining a hemoglobin level above 9 g/dL perioperatively and the regular use of incentive spirometry to reduce the risk of ACS. While the guidelines acknowledge that exchange transfusion can lower postoperative complications by reducing HbS levels, this intervention should be reserved for selected high-risk patients rather than used routinely. In this case, the patient demonstrated biochemical evidence of organ dysfunction, including elevated liver enzymes and acute kidney injury. While such findings may reflect ischemic damage intrinsic to SCD, they may also signal underlying sepsis-related multiorgan involvement [9]. Emerging literature suggests that sepsis-associated organ dysfunction in SCD patients correlates with worsened long-term outcomes and remains a significantly underreported complication [9]. This case exemplifies the need for multidisciplinary collaboration, involving hematologists, surgeons, and critical care teams, to ensure timely recognition and appropriate intervention in complex SCD presentations. Furthermore, future research into novel biomarkers—such as presepsin and interleukin-6 (IL-6)—may enhance early detection of sepsis in this vulnerable population, though their clinical application remains under development.

Conclusion

Anchoring bias remains a formidable obstacle in the timely diagnosis of surgical abdomen in patients with sickle cell disease (SCD). Clinicians must recognize that not all pain in SCD is attributable to the disease itself. Overreliance on VOC as the default diagnosis may obscure critical findings—such as fever and leukocytosis—which, while sometimes present at baseline, may also serve as early indicators of surgical or infectious emergencies. In this context, a lower threshold for employing advanced imaging modalities, particularly abdominal computed tomography (CT), is essential. Given the high sensitivity of CT for detecting intra-abdominal pathology, it plays a vital role in distinguishing surgical causes of pain from VOC. This is particularly important considering that patients with functional asplenia are immunocompromised and therefore more susceptible to overwhelming infections and sepsis, with significantly elevated mortality risk. Furthermore, appendicitis in SCD is often only diagnosed after perforation, an outcome that greatly increases morbidity and should be proactively avoided. Early recognition and prompt intervention are therefore paramount to improving clinical outcomes in this high-risk population.

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