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Spontaneous rupture of a normal spleen: a case report and literature review

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Highlights:

- Spontaneous spleen rupture occurs less than 1% of the cases in absence of identifiable underlying medical conditions (infectious, neoplastic, haematological diseases, or blunted minor trauma).
- Clinical manifestations include pain in the epigastric region or the left upper quadrant, tachycardia, and shock.
- Point-of-care ultrasound and abdominal computed tomography play a crucial role in diagnosis.
- Delays in diagnosis and treatment increase the risk of fatal outcomes.
- Standard guidelines for the treatment have not been established yet. Splenectomy is recommended in unstable haemodynamic patients; splenic artery embolization may be considered for selected patients.

Abstract

Spontaneous spleen rupture is a rare condition that occurs in less than 1% of cases without any identifiable underlying medical issues. Emergency clinicians should always include spontaneous

spleen rupture in the differential diagnosis of abdominal pain, as timely diagnosis and intervention are crucial for improving patient prognosis and preventing fatal outcomes. Clinical manifestations may include pain in the epigastric region or the left upper quadrant, tachycardia, and shock. Some patients may show signs of an acute abdomen upon physical examination while maintaining stable vital signs. Point-of-care ultrasound and abdominal computed tomography play a pivotal role in the diagnosis. Standard guidelines for the treatment have not been established. While splenectomy remains the mainstay of treatment, some selected hemodynamically stable patients may be suitable candidates for splenic artery embolization. In this minireview, we will begin by discussing the case of a 53-year-old man who experienced left lower back pain for five days, initially misdiagnosed as renal colic, and we will then conclude by examining the current literature on this important topic.

Case Report

A 53-year-old man presented to our emergency department with left lower back pain that had lasted for 5 days and was accompanied by fever and nausea. He was treated with intramuscular ketorolac for suspected renal colic without benefit. Since the onset of his symptoms, he had experienced episodes of presyncope, which were preceded by prodromal symptoms. The patient had a history of thrombocytopenia during an infectious episode in 2018, when he was treated for acute cholecystitis with a cholecystectomy; follow-up haematological tests had been negative. He reported not taking any medication and denied any recent history of trauma.

On admission, the patient was awake and alert with a Glasgow Coma Scale (GCS) of 15. His vital signs were stable, with blood pressure of 140/90 mmHg, heart rate of 80 beats per minute, oxygen saturation at 98% on room air, and a body temperature of 36.5 °C. Abdominal examination showed

no pain or tenderness upon palpation, and no defensive reaction was observed. An electrocardiogram documented sinus rhythm with normal ventricular repolarization. Point-of-care ultrasound revealed a slight left pleural effusion (approximately two intercostal spaces) accompanied by consensual pulmonary atelectasis. The spleen appeared normal in size but exhibited an irregular echo texture. There was also a slight perisplenic effusion and free fluid in the abdomen. An emergency computed tomography (CT) of the abdomen with contrast medium confirmed splenic rupture and hemoperitoneum (Figure 1). Laboratory tests showed the following results: white blood count 11,800/mm³, red blood count 3,740/mm³, haemoglobin 11.2 g/dL, mean cell volume 86.4 fL, platelets 291,000/mm³, and C-reactive protein 20 mg/dL (normal value <0.5). Hepatic and renal function, as well as coagulation time and lactate dehydrogenase levels, were within normal limits.

The surgical team was consulted. Given the low haemoglobin levels and the presence of hemoperitoneum without an arterial blush observed on the CT scan, the patient underwent an emergency splenectomy, which had a smooth postoperative course. Histological examination showed capsulo-parenchymal lacerations and intraparenchymal haemorrhage in an otherwise "normal" spleen. Comprehensive virological testing (HCV, HBV, HIV, CMV, EBV, Parvovirus, COVID19) resulted negative.

Patient was discharged on the seventh day in good clinical condition after receiving his post-splenectomy vaccinations.

Discussion

Abdominal pain is one of the most frequent ED complaints, accounting for approximately 10% of all visits,[1] and it continues to represent a diagnostic challenge for emergency physicians. Over a 50-year span, ED evaluation of abdominal pain in adults has evolved into a testing- and imaging-rich

process. This has resulted in fewer cases of missed surgical illness, but considerable prolongation in evaluation time and cost.[2]

Spleen rupture is usually due to trauma, while Atraumatic Splenic Rupture (ASR) is an extremely rare condition[3,4] usually associated with many systemic disorders affecting the reticuloendothelial system, including infections,[5-10] neoplastic,[11] inflammatory[13,13] or haematological diseases.[14,15] Ten cases of ASR have been reported in COVID-19 patients,[16-24] including a 13-year-old boy.[25]

Sometimes (9%), ASR can occur as a complication of drug abuse, such as cocaine[26] and heroine.[27]

Often, a triggering factor or minor physical event can be ascribed as the aetiology for rupture, including coughing,[28] vomiting[29] or minor medical procedures not involving the spleen, such as electrical cardioversion,[30] endoscopic procedures,[31-33] or surgical interventions, including cholecystectomy,[34] sleeve gastrectomy,[35] appendicectomy,[36] and hysterectomy.[37]

Anticoagulation therapy should be always considered as a possible cause of ASR. Direct Oral Anticoagulants (DOACs), apixaban, rivaroxaban, and dabigatran have all been associated with ASR.[38-40]

In extremely rare cases (<1%) ASR can occur spontaneously in absence of underlying diseases, as in our case.[41-44] The first case of occult ASR in absence of underlying diseases has been reported in 1861 by Rokitansky[43] and later, in 1874 by the English surgeon Atkinson.

The pathophysiology

The pathophysiology of ASR has not yet been completely clarified. Many theories have been proposed, including a pre-existing small, localized area of disease, a chronic passive splenic congestion, a rapid dissection of the splenic artery, a sudden increase in intra-abdominal pressure

associated with exercise, coughing, vomiting, or a forgotten minor trauma.[42] Probably, ASR is due to a combination of these conditions.

Clinical manifestations

Rupture of the spleen typically presents with severe abdominal pain and hemoperitoneum that requires splenectomy in 85% of the cases.[42] Clinical presentation can mimic other more common abdominal conditions, including peptic ulcer, pancreatitis, biliary diseases, myocardial infarction or angina pectoris,[45] pulmonary embolism, basal pneumoniae, and renal colic. It can also mimic thoracic or left shoulder pain.[46] It is interesting to note that even in presence of an acute abdomen, vital parameters can remain stable since a fair amount of intra-abdominal bleeding may be tolerated without signs of hypotension and tachycardia.

Diagnosis

The diagnosis can be missed due to the infrequency of ASR. In addition, spontaneous ASR is rare and poorly documented. The importance of bearing this possibility in mind is pointed out since the early recognition and management of spontaneous spleen rupture is crucial to optimize the patient's outcome avoiding catastrophic consequences, including fatal outcome.[3] In an extensive review dated 1958, Orloff and Peskin identified four criteria for diagnosing ASR,[47] as follows: the absence of a history of trauma or of unusual effort that could injury the spleen; the absence of perisplenic adhesions or scarring suggestive of previous trauma; the absence of pre-existing splenic disease; and normal microscopic and macroscopic appearance of the spleen except for haemorrhage and rupture. A fifth criterion has been introduced in 1991 to exclude a recent viral infection through a comprehensive virological study showing no significant rise in antibody titres.[48] Mononucleosis associated with Epstein-Barr virus remains the most common cause of ASR worldwide.[5]

Emergency physicians must keep in mind that normal vital parameters do not exclude spleen rupture in the presence of an abnormal abdominal examination. Point-of-care ultrasound should be performed as the first-line imaging test in these patients[49] since it plays a crucial role in a prompt diagnosis. Similarly in trauma patients, it can detect free fluid with 90% sensitivity and 99% specificity[50] and can aid in early diagnosis and urgent consultations and guide treatment plans in cases with undifferentiated hypotension.[51]

Contrasted enhanced CT scan of the abdomen is the gold standard technique for diagnosis, staging, and decision on surgical management, providing insights into splenic injuries and rupture, the extent of haemorrhage and guiding the appropriate management.

Management

Treatment guidelines do not exist. In the study by Wang and coll, the authors suggest conservative treatment – fluid resuscitation and analgesia - in blunt spleen rupture patients of CT classification grade of 1-4 with stable hemodynamical, while patients of CT classification grade 4-5 with instable hemodynamical should be treated surgically with splenectomy (Figure 2).[52] If the patient is on anticoagulation, initial management includes discontinuation of anticoagulants, volume resuscitation, and, if available, reversal of anticoagulation. Current guidelines recommend RBC transfusion, platelet, and cryoprecipitate transfusion to maintain target haemoglobin >7 g/dL, platelet count >50,000/μL, and fibrinogen >100 mg/dL, respectively.[53]

According to Harady *et al*., preoperative use of imaging studies in hemodynamically stable patients can sometimes obviate surgery, or in cases of massive hemoperitoneum reduce intraoperative time.[54] Embolization of splenic artery can be considered in selected patients with active contrast extravasation noted in CT angiography,[55] even if more clinical studies and research are necessary to

establish criteria for patient selection, appropriate timing for intervention and the long-term consequences.

Prognosis

The prognosis is strictly associated with prompt diagnosis and treatment. Spleen rupture of all causes is a potentially life-threatening event. ASR has an approximately mortality rate of 12%.[56]

Conclusions

This case should remind the emergency physician that nontraumatic spontaneous splenic rupture, although rare, should be considered in the differential diagnosis of unexplained shock or acute left upper quadrant abdominal pain, even if the patient has a stable haemodynamic and nonspecific symptoms. Early identification and treatment can be lifesaving.

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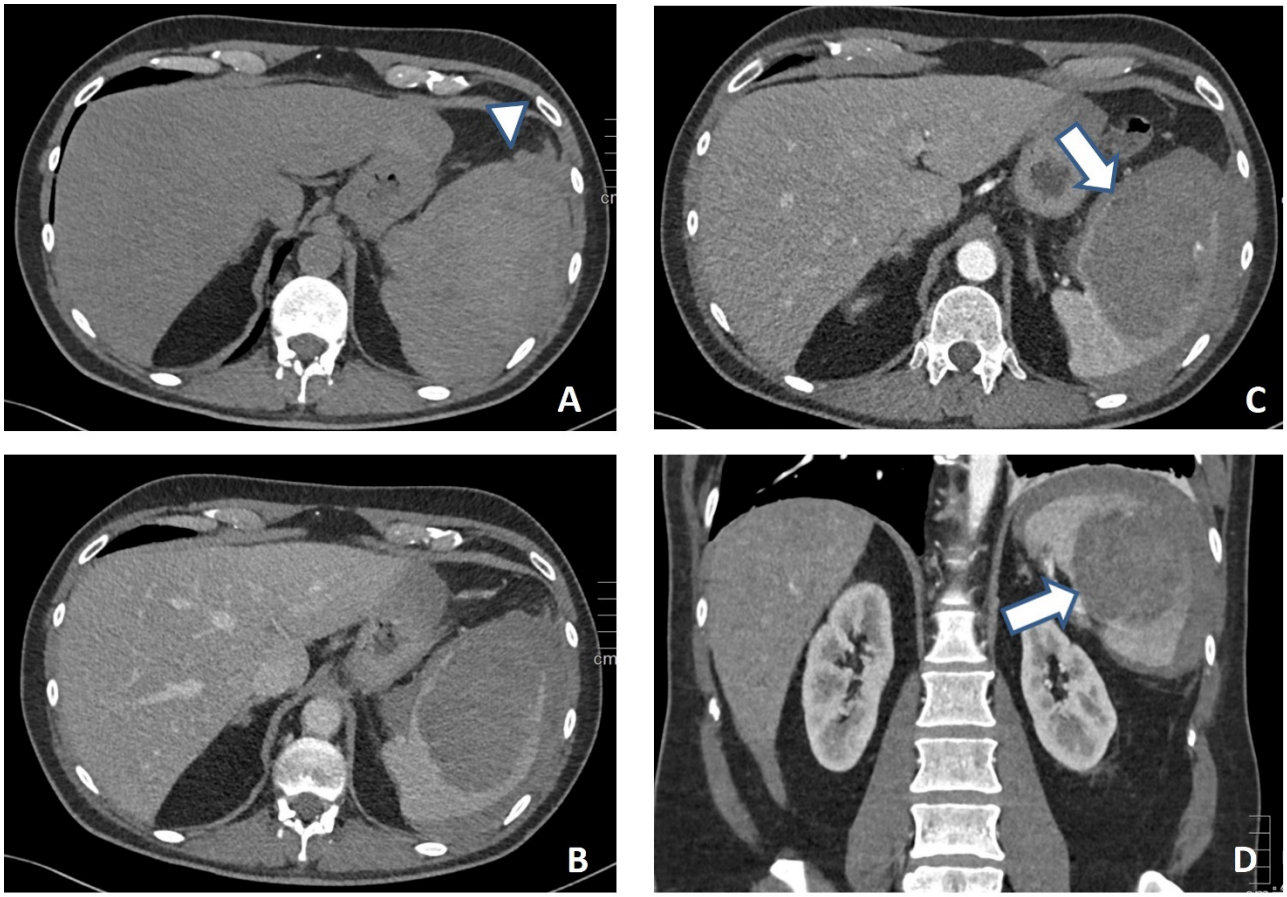

Figure 1. Contrast enhanced abdominal CT shows a perisplenic effusion measuring approximately 16 mm, hyperdense in the basal phase, consistent with recent blood (indicated by arrowhead, panel A). Following the administration of contrast medium, a large intraparenchymal hematoma measuring approximately 6.8 x 7 cm becomes visible (indicated by arrow, panel C and D) without arterial blush. Portal phase is showed in panel B.

AAST Grade and Type of Injury	Description
I	
Hematoma	Subcapsular, <10% of surface area
Laceration	Capsular tear, <1 cm of parenchymal depth
II	
Hematoma	Subcapsular, 10%–50% of surface area; intraparenchymal, <5 cm in diameter
Laceration	1–3 cm in parenchymal depth
III	
Hematoma	Subcapsular, >50% of surface area; ruptured subcapsular or parenchymal hematoma; intraparenchymal hematoma, >5 cm in diameter
Laceration	>3 cm parenchymal depth or involving trabecular vessels
IV	
Laceration	Laceration involves segmental or hilar vessels producing major devascularization (>25% of spleen)
V	
Laceration	Completely shattered spleen
Vascular	Hilar vascular injury that devascularized spleen

Figure 2. Spleen injury grading scale according to the American Association for the Surgery of Trauma (AAST).[57]