



Case Report

Spontaneous splenic rupture: A rare complication of acute myeloid leukemia. Report of a case

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ABSTRACT

Introduction: Spontaneous rupture of the spleen (SPR) is a rare and severe affection, difficult to diagnose, with multiple causes such as: Infectious and hematologic affections which represent more than half of the cases. Among this subset of patients, acute myeloid leukemia is one of the causes.

Presentation of case: A 48-year-old man undergoing chemotherapy for acute myeloid leukemia presented with acute intense abdominal pain. Computed tomography showed Abdominal CT scan showed a splenic rupture with abundant hemoperitoneum and bilateral pleural effusion. The patient presented hemodynamic instability and was immediately operated, splenectomy were performed.

Discussion: Spontaneous rupture of the spleen usually presents as a severe abdominal syndrome, which may accompany non-specific symptoms. Two signs are suggestive of splenic rupture: Kehr's sign (left diaphragmatic irritation resulting in referred pain to the left shoulder) and Balance's sign (palpable tender mass in the left upper quadrant). Diagnostic methods of choice are computed tomography and ultrasound. The prognosis is depending on the quality of care and the nature of the etiology. Splenectomy remains the cornerstone of the treatment of splenic rupture. It is important to include splenic rupture as a differential diagnosis for acute abdominal pain, especially in patients with hematologic malignancy, since early recognition and treatment increase patient survival and improve the prognosis.

Conclusion: Even if spontaneous splenic rupture is rare, every clinician should have in mind the reflex to think of it, especially in patients with hematologic malignancies.

1. Introduction

The spleen is an immunological organ involved in hematological and non-hematological affections. Spontaneous splenic rupture (SSR), also called atraumatic splenic rupture, is a rare abdominal emergency that requires immediate diagnosis and prompt surgical treatment to ensure the patients survival [1]. Multiple pathologies have been associated with splenic rupture: infectious, hematological, congenital and autoimmune diseases. Acute Myeloid Leukemia (AML) is one of the main causes.

The objective of this study is to highlight the seriousness of this condition and the different diagnostic and therapeutic modalities.

This work has been reported in line with the SCARE criteria [2].

2. Case report

A 48-year-old man with no history of recent trauma undergoing chemotherapy for acute myeloid leukemia, **was admitted to our hospital's emergency department for acute abdominal pain.** At admission, the patient had the following vital signs: heart rate 120 beats/min, respiratory rate 30 breaths/min, arterial pressure 90/60 mmHg, body temperature 37.4 °C, with a flexible abdomen. Ultrasound revealed abundant hemoperitoneum with a heterogeneous upper segment of the spleen. Abdominal CT scan showed a splenic rupture with abundant hemoperitoneum and bilateral pleural effusion (Fig. 1).

Biological testing revealed hyperleukocytosis at 22,000 mm³, hemoglobin at 8.9 g/L and thrombocytopenia at 66,000 platelets/mm³. The hepatic and renal check-ups were normal: aminotransferase (AST): 29U/L, alanine aminotransferase (ALT): 16U/L, serum creatinine (4.3

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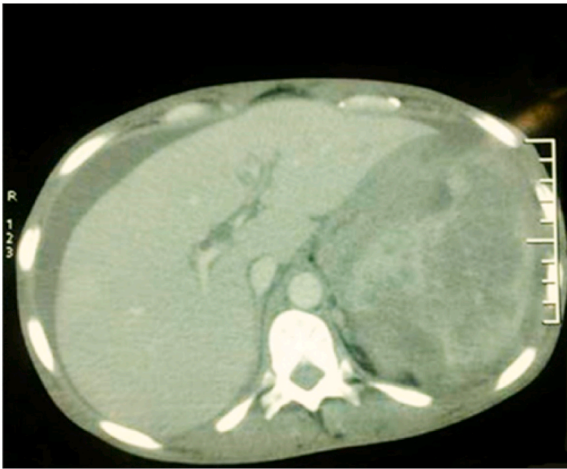


Fig. 1. Computed tomography scan showing a splenic rupture with hemoperitoneum.

mg/dL), urea (0.22g/L), potassium (5.9meq/L), and C-reactive protein (175mg/L).

The patient presented hemodynamic instability, **After optimization of his general condition with a Naso-Gastric tube suction and intravenous fluids resuscitation, a decision was taken to proceed with an emergency laparotomy, under general anesthesia with endotracheal intubation. Performed by senior general surgeon.** Preoperative prophylactic antibiotics were administered. Intraoperative exploration revealed a high abundance of hemoperitoneum, with capsular rupture on the posterior aspect of the spleen (Fig. 2). Splenectomy was performed successfully. During the procedure, **the patient required transfusion of 3 units of red blood cells and administration of abundant crystalloids.** Postoperatively the patient presented a cardiovascular decompensation and died on the seventh day. An anatomopathological study of the surgical specimen revealed a splenic localization of acute myeloid leukemia.

3. Discussion

Splenic rupture is a rare and life-threatening complication in which

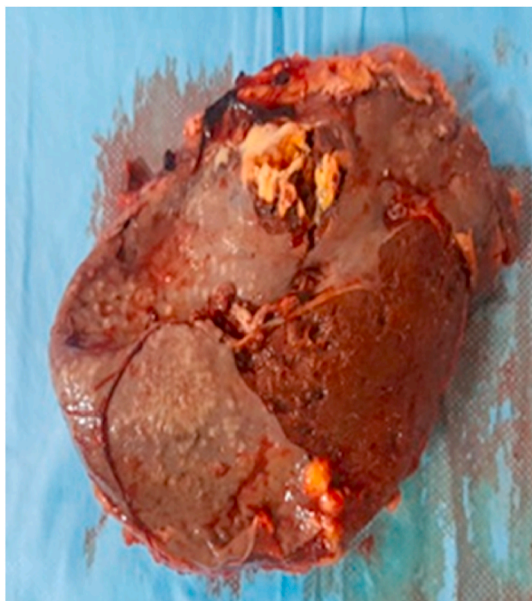


Fig. 2. Surgical piece showing the capsular rupture of the posterior face.

the spleen is damaged, producing internal hemorrhage in the abdominal cavity. According to the Orloff and Peskinin 1958 criteria, splenic rupture should be considered “spontaneous” only if it occurs without trauma history or other spleen pathologies [1]. Spontaneous rupture of the spleen is a very rare affection. Infectious and hematological diseases represent more than half of its causes, infections are due to: infectious mononucleosis, malaria and cytomegalovirus, while hematological causes are mainly due to malignant tumors such as leukemia, Hodgkin’s and non-Hodgkin’s lymphomas (NHL) [3]. Other cases of spontaneous rupture of the spleen include: congenital splenic lesions such as hamartoma and hemangioma, spleen infarction, clotting disorders, thrombocytopenia, autoimmune diseases, hemolytic anemia, pregnancy, amyloidosis, portal hypertension and focal splenic lesions [1,4].

Renzulli et al. summarized 845 patients with atraumatic splenic rupture from previous reports dating from 1980 to 2008, the etiological analysis found that the neoplastic cause accounted for 30.3% of the cases, followed by infection in 27.3% of the cases, inflammation in 20.0% of the cases and drug/treatment-related causes in 9.2% of the cases [5].

According to the study conducted by Kianman et al. [6], spontaneous rupture of the spleen was more frequent in men than women, the median age of presentation was 42 years, one third of the patients had hypovolemic shock and 8% died before surgery. The most frequent symptom was pain in the left upper quadrant [6]. The mechanism of onset for SSR is non specific, Hynes et al. suggested three mechanisms: the mechanical effect of leukemic infiltration in the spleen, especially if the capsule is invaded, splenic infarction with consequent subcapsular hemorrhage and subsequent rupture of the splenic capsule, and abnormalities of blood coagulation [7]. Regarding the SSR in acute myeloid leukemia, incidence is not known, the pathogenesis is still unclear and the mechanisms of leukemic infiltration of coagulopathy and infarct are incriminated.

Spontaneous rupture of the spleen usually presents as a severe abdominal syndrome, which may accompany non-specific symptoms simulating acute coronary syndrome, acute pancreatitis and ruptured aortic aneurysm, delaying diagnosis and treatment [8], no clinical manifestation can be used to definitively diagnose splenic rupture [9]. Two signs are suggestive of splenic rupture: Kehr’s sign (left diaphragmatic irritation resulting in referred pain to the left shoulder) and Balance’s sign (palpable tender mass in the left upper quadrant). The patients are often found in hypovolemic shock with signs of peritonitis on physical examination [10,11].

Abdominal ultrasound is the main examination to be performed to confirm the diagnosis of spontaneous rupture of the spleen. The Focused Abdominal Ultrasound Technique (FAST) can be used in the emergency room as a fast and safe method. CT scan has a sensitivity and a specificity of at least 95% in the detection of splenic lesions, it is particularly cost-effective when used to answer targeted questions that define the need for specific interventions, and define the nature and severity of the splenic lesion. MRI is an excellent tool for the diagnosis, the evaluation and the characterization of various focal splenic lesions and pathological aspects [10].

There is no doubt that a surgical intervention (splenectomy) is a necessity in patients with abundant hemoperitoneum and refractory hypovolemic shock. The use of conservative approach seems very limited in this disease. The selection of patients who would benefit conservative surgery remain controverted, integrity of the splenic pulp on ultrasound and CT scan, hemoglobin and hematocrit levels should be verified every 4–6 hours for the first 24 hours until the patient is stabilized [11–13].

Spleen rupture due to neoplastic disorders has the particularity of causing a high mortality rate, the risk factors for spontaneous rupture of the spleen that are associated with mortality include: splenomegaly, age over 40, neoplastic disorders [5]. The overall mortality rate reported in the literature is 14%. After a splenectomy, regardless of the indication, it is conventional to prevent infection by vaccinating against

Streptococcus pneumoniae, *Haemophilus influenzae* and *Neisseria meningitidis* [11].

4. Conclusion

Even if spontaneous splenic rupture is rare, every clinician should have in mind the reflex to think of it, especially in patients with hematologic malignancies. The diagnosis is based on the ultrasound or the abdominal CT scan, a detailed pathophysiology of the spontaneous splenic ruptures is still unclear. The treatment is splenectomy. The high mortality rate seems to be mainly related to delayed diagnosis and/or the severity of the underlying pathology.

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I declare on my honor that the ethical approval has been exempted by my establishment.

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Author contribution

AH: writing the paper.
 KY: Corresponding author writing the paper.
 SEL M: writing the paper.
 DE: study concept.
 RB: study concept.
 SR El J correction of the paper.
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Registration of research studies

Not applicable.

Guarantor

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Consent

Written informed consent for publication of their clinical details and/or clinical images was obtained from the patient.

Declaration of competing interest

The authors declare having no conflicts of interest for this article.

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