

Case Report

Spontaneous splenic rupture in a patient with chronic granulocytic leukemia

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Received: 21 June 2021

Accepted: 06 August 2021

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ABSTRACT

Spontaneous splenic rupture is a rare phenomenon that is not associated with trauma. The most common causes of splenic rupture are hematological diseases (30.3%), inflammatory diseases (20%), infectious diseases (27.3%), drugs (9.2%), mechanical disorders (6.8%), and unknown causes (6.4%). Spontaneous splenic rupture secondary to hematological malignancy is rare; in this group of patients, chronic granulocytic leukemia is the main cause. The mechanism of spontaneous splenic rupture is uncertain. Three mechanisms have been suggested: the mechanical effect of leukemia and its infiltration in the spleen, especially if the capsule is invaded; splenic infarction with subsequent subcapsular hemorrhage and rupture of the splenic capsule, and coagulation abnormalities.

Keywords: Spontaneous splenic rupture, Hematological malignancy, Emergency splenectomy

INTRODUCTION

Spontaneous splenic rupture is a rare phenomenon that is not associated with trauma. The most common causes of splenic rupture are hematological diseases (30.3%), inflammatory diseases (20%), infectious diseases (27.3%), drugs (9.2%), mechanical disorders (6.8%), and unknown causes (6.4%).¹ Spontaneous splenic rupture secondary to hematological malignancy is rare; in this group of patients, chronic granulocytic leukemia is the main cause.^{2,3}

CASE REPORT

A 43 year old male patient with a history of type 2 diabetes mellitus came to the emergency department of our hospital due to abdominal pain. He complained of intense pain in the left hypochondrium and mesogastrium with 4 hours of evolution, accompanied by general

malaise, vomiting and drowsiness. On admission, his vital signs were heart rate 110 bpm, respiratory rate 22, blood pressure 100/50 mmHg, body temperature 37°C and oxygen saturation of 93%. On physical examination, he was pale with a distended abdomen and a predominance of pain in the epigastrium and left upper quadrant with rebound tenderness. He had no history of trauma. Acute diverticulitis, hollow viscus perforation, and acute pancreatitis were suspected as differential diagnoses. A complete blood count showed leukocytosis 266,000/mm³, hemoglobin of 9.1g/dl, hematocrit of 30%, and platelets 394,000. Hepatomegaly, a grade 5 splenic lesion, and perisplenic free fluid were seen in an abdominal CT (Figure 1).

After circulatory resuscitation and transfusion of packed red blood cells, platelets, and fresh frozen plasma, surgical treatment was decided. An exploratory laparotomy was performed where a hemoperitoneum of 2500cc was found together with a ruptured subcapsular

hematoma with active bleeding and multiple blood clots (Figure 2).



Figure 1: CT splenic lesion grade 5.

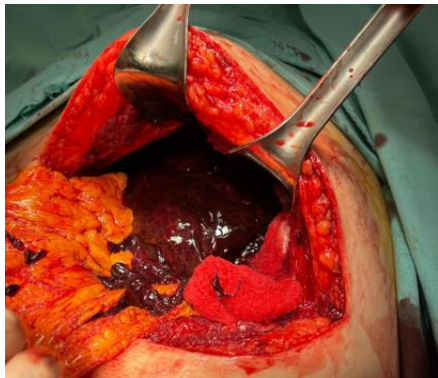


Figure 2: Hemoperitoneum.



Figure 3: Splenic rupture.

The stomach was mobilized through the greater curvature, and the splenic hilum and the tail of the pancreas was identified. Then, the splenic vein and artery were clamped, cut, and ligated. The area was irrigated, and the short gastric artery was clamped and ligated; the phrenicosplenic and splenocolic ligaments were released. A 20x13 cm specimen was obtained (Figure 3). On histopathology study, the spleen weighed 1.2 kg with chronic passive congestion associated with extramedullary hematopoiesis. The hematological evaluation was complemented with a peripheral blood

smear and a Coombs test. The diagnosis was chronic granulocytic leukemia. The patient had a favorable evolution, being discharged after six days without patient medical management with hydroxyl urea and imatinib by the hematology department and general surgery.

DISCUSSION

Pathological spleen rupture occurs without associated trauma. Any spontaneous rupture should be investigated for splenic pathology. Rokitsky first described this condition in 1861. The term "pathological splenic rupture" was introduced in 1928 by Lance to define the non-traumatic splenic rupture that occurs.⁴ In most cases, spontaneous splenic rupture is caused by three conditions, hematological malignancy; for example, leukemia or lymphoma (30.3%); infectious diseases; for example, malaria, mononucleosis, or other viral infections (27.3%); and inflammatory or neoplastic diseases; for example, acute or chronic pancreatitis (20.0%). Other causes include drugs, chemotherapy such as filgrastim therapy (9.2%), and mechanical disorders (6.8%).^{2,5,6} Splenic rupture has a mortality of approximately 12%; however, it could reach up to 21% in patients with neoplastic disease.^{2,7}

Spontaneous splenic rupture secondary to hematologic malignancies is rare. According to the literature, chronic granulocytic leukemia (15.8%) and Hodgkin's lymphoma (36.2%) are the main causes. Other causes include myeloproliferative entities (15.8), acute myeloid leukemia (13.8%), acute lymphoblastic leukemia (7.9%), and myelodysplastic entities (7.9%).^{5,6} The mechanism of spontaneous splenic rupture is uncertain. Three mechanisms have been suggested: the mechanical effect of leukemia and its infiltration in the spleen, especially if the capsule is invaded; splenic infarction with subsequent subcapsular hemorrhage and rupture of the splenic capsule; and coagulation abnormalities.⁸ The clinical symptoms of splenic rupture with hematological malignancy are the result of intra-abdominal hemorrhage and acute abdominal pain of variable location and intensity, sometimes radiating to the left shoulder (Kher's sign), which occurs in 50% of cases of spontaneous splenic rupture, together with splenomegaly on palpation, hypotension, and tachycardia.⁹

Abdominal ultrasound and abdominal CT are sensitive and specific for diagnosis. Treatment can be carried out by interventional radiology or surgery.^{10,11} In hemodynamically unstable patients, emergency splenectomy is indicated as a rescue measure.^{12,13}

CONCLUSION

A diagnostic suspicion of splenic rupture is of utmost importance in patients with hematological entities that are rare. Two treatment methods have been proposed, interventional angio-embolization and laparotomy splenectomy, in conditions of hemodynamic instability. Emergency splenectomy is a rescue measure in patients

with splenic rupture with an unfavorable immune condition and hematological abnormalities. It requires excellent multidisciplinary management in post-operative care.

ACKNOWLEDGEMENTS

Authors would like to thank the team of general surgery teachers for guiding us in performing emergency surgery, as well as the nursing staff who accompanied us, being of vital importance in the intervention process.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Miranda HV, Zarazúa ROM, Ramírez MAH, Briseño JAR, Maldonado GEM. Spontaneous splenic rupture in a patient with chronic granulocytic leukemia. *Int Surg J* 2021;8:2786-8.