CASE REPORT

Chronic lymphocytic leukaemia and spontaneous rupture of spleen

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SUMMARY

Pathological or spontaneous splenic rupture is a rare but well-recognised complication of haematological malignancies. The authors present a clinical report of a 78-year-old woman with known clinical history of chronic lymphocytic leukaemia and atrial fibrillation under anticoagulation with apixaban which has spontaneous splenic rupture. Pathological examination revealed lymph node and splenic infiltration due to chronic lymphocytic leukaemia. The diagnosis of splenic rupture must be considered in all patients with haematological malignancies who experience acute abdomen. Given the severity, it requires a correct and timely diagnosis.

BACKGROUND

The spontaneous rupture of the spleen associated with haematological malignancies is a rare complication, with a few cases reported in the last 80 years. It requires a correct and timely diagnosis, because on that depends the patient's outcome and survival. So it is important that clinicians are aware of this pathology.

CASE PRESENTATION

We present a case of a 78-year-old woman with a previous clinical history of chronic lymphocytic leukaemia (CLL), treated with chemotherapy, with periods of remission and progression, but with the disease controlled in the past 3 years; atrial fibrillation anticoagulated with apixaban; type 2 diabetes mellitus and arterial hypertension.

She was admitted in our intensive care unit in shock with multiorgan dysfunction syndrome, with the diagnosis of leukaemic crisis, due to the sudden increase of lymphocyte count.

She had been previously admitted in the emergency room, after syncope at a wedding, with recovery of consciousness in <30 s, accompanied by nausea and abdominal pain. She denied incontinence of sphincters or convulsive movements, chest pain or palpitations. There was no history of trauma. She denied the use of any illicit drugs, tobacco and alcohol. Family history was unremarkable, including the absence of any malignancy.

She was lethargic, pale, appeared to be in moderate distress, hypotensive and tachycardic, in respiratory exhaustion and anuric.

She was intubated, ventilated and after an arterial and central line, vasopressor support was initiated.

The physical examination revealed an acute abdomen.

INVESTIGATIONS

Complete blood count revealed haemoglobin level of 5.1 g/dL, white cell count $197\,000 \times 10^9/L$, platelets $89\,000 \times 10^9/L$, international normalised ratio 1.5, activated partial thromboplastin time 28.8 s, creatinine 1.9 mg/dL, urea 81 mg/dL, negative myocardial necrosis markers; arterial gas analysis reveals metabolic acidemia with hyperlactacidaemia.

We immediately performed an abdominal CT scan: 'spleen sharply increased dimensions and heterogeneous density; ascites filling all the peritoneal recesses; no pneumoperitoneum' (figure 1).

DIFFERENTIAL DIAGNOSIS

The splenic rupture makes differential diagnosis with splenic infarction. The splenic infarction refers to occlusion of the splenic vascular supply, leading to parenchymal ischaemia and subsequent tissue necrosis and it is associated most commonly with haematological disorders. The most common presenting symptom is left upper-quadrant abdominal pain (up to 70%). Additional symptoms include fever, nausea and vomiting and left shoulder pain (Kehr's sign). These signs and symptoms also occur in the splenic rupture.

The splenic infarction can precede the splenic rupture. However, when the rupture occurs, the free intraperitoneal blood causes diffuse abdominal pain, peritoneal irritation and the signs of shock manifest (hypotension, tachycardia, tachypnoea, distress and decreased capillary refill). Our patient had signs of shock.

TREATMENT

Due to that, she was taken to the operating room and underwent urgent exploratory laparotomy, where, after it was found haemoperitoneum and rupture of the spleen, a splenectomy was performed.

She developed disseminated intravascular coagulation, requiring 12 units of erythrocyte concentrate, 10 fresh frozen plasma units, 8 g of fibrinogen, platelets and tranexamic acid.

The pathological examination revealed spleen weight of 1134.6 g and a size of $20 \times 12 \times 7.5$ cm, lymph node and splenic infiltration by chronic lymphatic leukaemia (B cells CD20+ with CD5 and BCL-2 expression) with splenomegaly and spontaneous rupture of the spleen.

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Figure 1 CT scan, coronal plan, spleen sharply increased dimensions; liquid filling all the peritoneal recesses.

OUTCOME AND FOLLOW-UP

The patient subsequently made a gradual recovery in the intensive care, being safely discharged to the surgical ward initially and afterwards home.

DISCUSSION

The presentation of splenic rupture in a patient varies, often posing a diagnostic challenge for clinicians. Spontaneous rupture of spleen is a rare but known complication of haematological malignancies.

It was first described by Rokitsky in 1861 and mentioned in multiple cases since.¹ The term ‘pathological splenic rupture’ was introduced in 1928 by Lange to define the non-traumatic splenic rupture that occurs in a diseased spleen. It occurs most commonly in patients with acute leukaemia, but is well documented in chronic leukaemia and lymphomas as well.²

Almost all patients experience abdominal pain at the time of rupture, although with variable intensity and irradiation, being often confused with biliary colic, aortic aneurysm, hollow viscera perforation, pancreatitis and acute coronary syndrome.³

In this case, it was initially assumed to be a leukemic crisis and the haemoperitoneum was confused with ascites on abdominal CT scan.

In a review of 48 cases, conducted by Bauer *et al* in 1981, the author describe that pain in the left hypochondrium with irradiation to the homolateral shoulder (Kehr’s sign), considered a classic haemoperitoneum sign, was only described in 17% of the cases. Hypotension was reported in 66%, fever in 74% and tachycardia in 75%. There is no consensus between spleen size and risk of rupture, since, according to Bauer *et al*, 40% of patients with splenic rupture did not have splenomegaly.⁴

Hynes and colleagues described three possible mechanisms of splenic rupture in patients with leukaemia: splenic infiltration due to haematological diseases, splenic infarction and coagulation disorders (which contributes to intracranial and subcapsular haemorrhages leading to an increased risk of rupture).⁵ In this patient the combination of the CLL with oral anticoagulation led to the outcome.

There is controversy about the influence of the haemodynamic profile on the prognosis of splenic rupture. Bauer *et al* showed

that haemodynamically unstable patients may have a better prognosis, which presumably correlates, according to those authors, with an earlier diagnosis and treatment in these situations.⁴

According to Bauer *et al*, postsplenic survival does not appear to be related to factors such as the type of haematological disease, of established therapy, platelet and white cell counts or spleen size. By far the most important predictor of survival was splenectomy.⁴

Several imaging methods may be useful in diagnosis, but CT scan is the examination of choice for detecting splenic rupture, subcapsular haematoma and haemoperitoneum, and allows the differential diagnosis with other causes of acute abdomen.⁶

The treatment of splenic rupture associated with malignant haematological diseases is splenectomy, with an immediate post-operative survival of 78%.⁶

With conservative treatment, the evolution of splenic rupture due to malignant haematological disease seems to be invariably fatal, electing surgical therapy as the most important factor to predict survival.⁶

In this case, the ascites, reported on CT scan, was later interpreted as haemoperitoneum and urgent laparotomy with splenectomy was performed, saving the patient’s life.

In short, pathological splenic rupture, though rare, should be considered in cases of acute abdomen in patients with haemato-oncological condition. Given the severity, it requires a correct and timely diagnosis. Splenectomy remains the therapy of choice.

Learning points

- ▶ Though rare, spontaneous rupture of the spleen is a known complication of haematological malignancies.
- ▶ It requires a correct and timely diagnosis, because on that depends the patient outcome and survival.
- ▶ Believing the clinic even when the complementary tests are negative, not delaying the treatment.

Contributors IMN contributed with conception and design, acquisition of data, analysis and interpretation of data. AIP contributed with acquisition of data, analysis and interpretation of data. RC contributed with revising the article critically for important intellectual content. AR contributed with final approval of the version published.

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