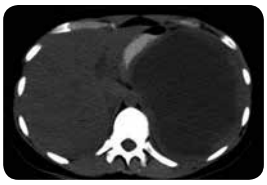

Pathological Spleen Rupture as Clinical Presentation of Diffuse Large B-Cell Lymphoma: Case Report*

Ruptura patológica del bazo como presentación clínica del linfoma difuso de células B grandes: Presentación de caso

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Summary

Non-traumatic splenic rupture is a rare entity; the frequency reported in the world literature is less than 1%. Most hematologic malignancies can affect the spleen, including various types of lymphomas, leukemia, and malignant plasma cell tumors. The most common haematological neoplasia affecting the spleen is Non-Hodgkin's lymphoma, among which the diffuse B-cell subtype is the most common. This article presents a case report of pathological splenic rupture in a 26-year-old patient as the first clinical manifestation of diffuse large B-cell lymphoma. Early diagnosis and clinical suspicion play a vital role in the outcome. A concise review of the available literature on the definitions and diagnostic findings of this event is made.

Resumen

La ruptura esplénica no traumática es una entidad rara; la frecuencia informada en la literatura mundial es menor del 1 %. La mayoría de los tumores malignos hematológicos pueden afectar el bazo, incluyendo varios tipos de linfomas, leucemia y tumores malignos de células plasmáticas. La neoplasia hematológica más común que afecta el bazo es el linfoma no Hodgkin, dentro de este, el subtipo difuso de células B es el más común. En este artículo se presenta el caso de ruptura esplénica patológica en una paciente de 26 años como primera manifestación clínica de un linfoma difuso de células B grandes. El diagnóstico temprano y la sospecha clínica juegan un papel vital en el desenlace, se hace una concisa revisión de la literatura disponible acerca de las definiciones y ayudas diagnósticas de este evento.

Introduction

Spleen rupture can be classified as traumatic, spontaneous or pathological (non-traumatic) (1). Trauma is considered the most common cause of splenic rupture (2) and its images are easily interpreted by radiologists; non-traumatic splenic rupture is rare, is usually diagnosed later and is a major challenge for radiologists (2).

Splenic rupture is a rare complication of pathological processes, but most haematological malignancies can affect the spleen, including various types of lymphomas, leukemia, and malignant plasma cell tumors. The most common haematological neoplasia affecting the spleen is non-Hodgkin's lymphoma and within this, the diffuse B-cell subtype is the most common (3).

The case of a patient diagnosed with secondary splenic outbreak with diffuse large B-cell non-Hodgkin lymphoma and its imaging findings are described below.

Case description

A 26-year-old female patient with no relevant history. Consultation with the emergency department for a clinical picture of 14 days of evolution consisting of abdominal pain located in the left hypochondrium and irradiated to the back, which is exacerbated by postural changes. Concomitantly, there is evidence of asthenia, adynamia and weight loss of 6 kg in the last 6 months associated with hyporexia.

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Physical examination is tachycardia, afebrile, with oral mucosa semiseca, skin paleness and without adenomegalias in the neck. The soft abdomen is palpated, with painful mass in the upper left quadrant, without other findings. Symptomatic management with parenteral and paraclinic hydration is initiated, in which: Leukocytosis at the expense of lymphocytes, mild anemia, thrombocytosis, mild elevation of liver function tests, and very high C-reactive protein (Table 1).

Table 1. Results of the paraclinical exams

Paraclinical	Results
Leukocytes	13.320
Neutrophils	67 %
Lymphocytes	16,5 %
Hemoglobin	11,4
Hematocrit	37,8 %
Platelets	673.000
MCV	67 Fl
Creatinine	0,76
BUN	8,4
ALT	52
AST	57
Alkaline phosphatase	211
Amylase	61
Sodium	137
Potassium	4,68
PCR	100,4
TP	11,9
TTP	28,5

A simple abdomen tomography with contrast medium is performed in 3 phases, in which splenomegaly is observed by mass of low density, with well defined contours, occupying the upper half of the spleen associated with low density collection, with irregular borders, which does not enhance with the contrast medium. Another nodular lesion of low density is seen in the lower pole of the spleen (Figure 1).

Faced with imaging findings and clinical suspicion of possible hematolymphoid neoplasm, bone marrow aspirate was performed with flow cytometry with the following results: mature T lymphoid population, 6.8%; lymphoid B mature population, 1.1%; myeloid mature population, 68%; erythroid population of 11%; no manifestation of myeloid or tumor lymphoid population. With these findings and immunohistochemistry, diffuse large B-cell lymphoma stage IIISX, of central-germinal origin, with giant focal splenic lesion was diagnosed.

The patient undergoes subphrenic cystic collection drainage and splenectomy. Subsequently, he received 6 cycles of R-CHOP chemotherapy (cyclophosphamide, doxorubicin, vincristine, prednisolone) for three weeks. In the PET-CT performed during the follow-up, complete remission was evidenced, with corresponding CHESON criteria with complete response.

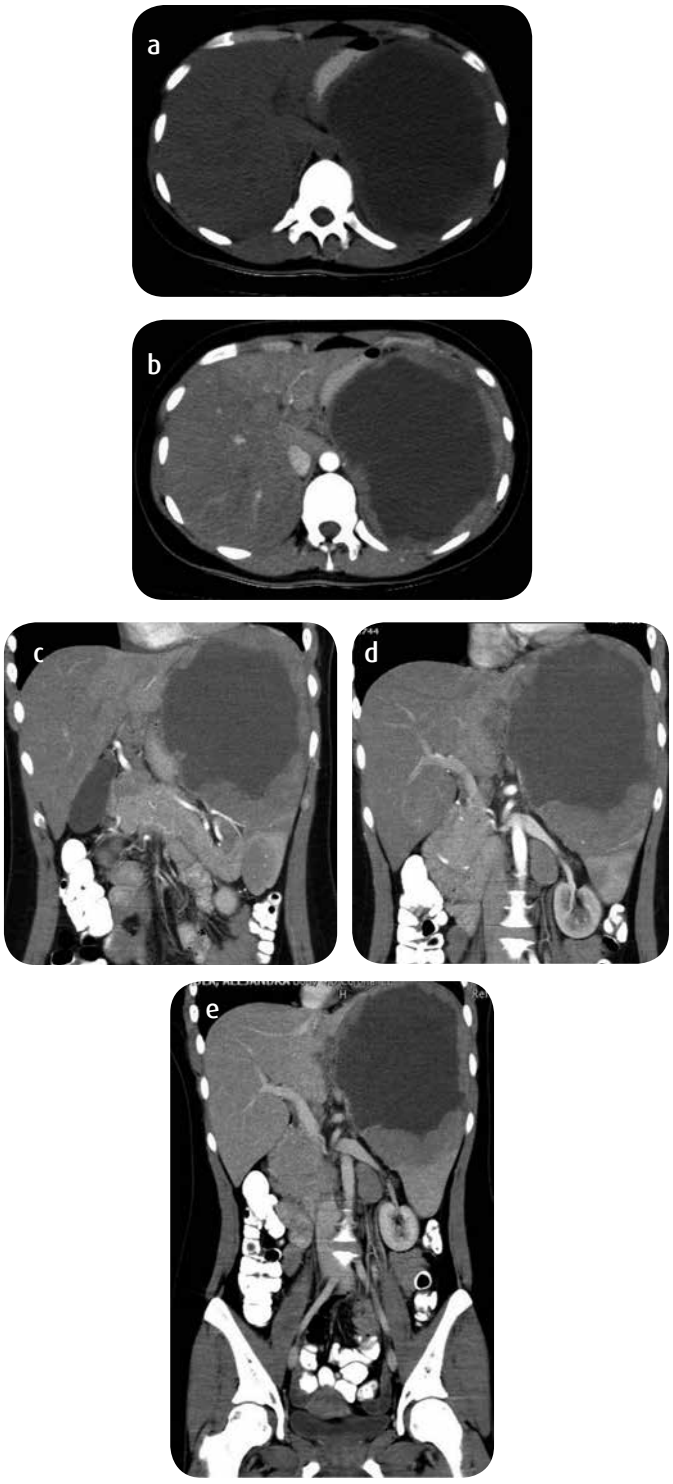

Figure 1. Chronic splenic rupture hematoma in a patient with NHL. a) Simple abdominal CT scan, axial cut, showing a splenic intraparenchymal collection, low signal, well defined borders, with thick and irregular wall, occupying practically the whole of the spleen, bulging its contour. In the images after the contrast medium, arterial phase, b) axial and c and d) coronal, there is absence of central enhancement with slight enhancement of the wall. e) In the portal phase there is still no enhancement of the contrast medium by the lesion. Note that the preserved splenic parenchyma in the lower pole has a normal behavior in the different phases after administration of the intravenous contrast medium.

Discusion

Etiology and epidemiology

Non-traumatic splenic rupture is a rare entity; the frequency reported in the literature is less than 1% (1). This is why there are no guidelines for handling and case series are therefore useful in establishing standards that will contribute to the realization of articles of greater scientific weight.

Debnath and Valerio used the terminology pathological and spontaneous rupture to distinguish the causes of non-traumatic rupture of the spleen and to give clarity to the classification, since the nomenclature of this condition is often confused. The difference is that pathological ruptures occur in a macroscopic or microscopically diseased spleen and spontaneous ruptures in a normal spleen (4).

Non-traumatic splenic rupture was first described by Atkinson in 1874 as a postmortem finding. The most common causes are malignant haematological disorders, viral infections (infectious mononucleosis and cytomegalovirus infection) and non-neoplastic inflammatory diseases (acute and chronic pancreatitis) (Table 2). Neoplastic disorders are the etiology with the highest mortality rate related to non-traumatic rupture of the spleen (5).

Half of all patients with Hodgkin's lymphoma and one-third of those with non-Hodgkin's lymphoma have spleen involvement; However, even in cases of non-Hodgkin's lymphoma, pathological splenic rupture as the first symptom of the disease is rare, as is the presence of primary splenic lymphoma (4).

Table 2. Etiology of non-traumatic rupture of the spleen

Causes	%
Neoplastic diseases Non-Hodgkin's Lymphoma, myeloproliferative disorders, acute myelogenous leukemia, angiosarcoma.	16,4
Infectious diseases Infectious mononucleosis, cytomegalovirus infection, endocarditis, tertiary malaria *(Plasmodium vivax)*.	14,8
Noninfectious inflammatory diseases Chronic pancreatitis, acute pancreatitis, primary amyloidosis.	10,9
Medicines Anticoagulation, hemodialysis.	7,2
Normal spleen	6,4
Mechanical disorders During pregnancy, hepatic cirrhosis (portal hypertension).	4,3

Source: Renzulli et al. (5).

Diagnostic imaging

If there is suspicion of diffuse or focal diseases of the spleen, studies that demonstrate the internal structure of this organ should be obtained, ultrasound (US), computed tomography (CT) and magnetic resonance imaging (MRI) (6).

With US, the size and morphology of the spleen can be estimated quite accurately using a 3.5-5 MHz transducer. Normal spleen parenchyma is homogeneous, with less echogenicity than hepatic parenchyma and greater than that of renal parenchyma. In the hands of an experienced radiologist, US is a highly reliable, fast, safe, cost-effective method and is the first option to identify spleen abnormalities (7).

In CT images, normal splenic tissue is homogeneous with an attenuation of 40-50 Hounsfield units (UH); the use of intravenous contrast media allows better characterization of splenic pathologies (8). Finally, MRI offers an advantage with multiphase images without the use of ionizing radiation. The signal intensity of the splenic tissue in MRI is smaller than that of the liver and greater than that of the muscle in the sequence with T1 information, has a signal similar to the renal cortex in sequence with information T1 and greater than the liver in sequence with T2 information (8).

In CT, US, and MRI, lymphomatous involvement of the spleen may manifest as a diffuse growth with homogeneous or multiple density, of varying sizes, replacing the splenic pulp (4). CT and MRI are not sensitive in the detection of splenic involvement due to lymphoma, since 45-70% of the lesions have diffuse infiltration or tumors of less than one centimeter (7).

In the images, 4 types of compromise can be found: diffuse splenomegaly, infiltrative miliary lesions (1-5 mm), multifocal lesions (2-10 cm) and solitary lesions (7-14 cm) (8). The characteristics in the different types of images are:

- In CT without contrast medium, the lesions are usually of low density, but they are not very striking.
- In MRI, lesions are often not very striking in sequences with T1 information or with T2 information.
- In CT and MRI with contrast medium lesions are more striking in late portal phase and at the first minute they are seen with density and signal similar to that of the spleen.

Primary spleen lymphoma shares the imaging characteristics of the systemic one, but it has two characteristics that are more common: central necrosis (70.6%) and local organ invasion (4).

Analysis of the clinical case

The non-traumatic rupture of the spleen is often fatal, the mortality rate approaches 12.2%, and of the six main causes, the neoplasias are considered the most risky (6). In this article we report the case of a diffuse large B-cell NHL that debuts with subacute splenic rupture and splenic hematoma, a rare form of presentation that requires a high index of suspicion. Splenic rupture frequently presents with severe abdominal pain, associated with hypovolemic shock; in this case, the patient had an insidious evolution, with atypical radiological findings, since not being acute bleeding can be easily confused with a splenic abscess or a cystic lesion.

It is important to emphasize the need for timely diagnosis and rapid intervention in patients with suspected non-traumatic rupture of the spleen to improve the prognosis of the disease, due to the high mortality rate generated by this entity; therefore, it is evident that early

diagnosis plays an important role in the natural history of this disease.

Because of its low frequency and low clinical suspicion, it is the radiologist who often comes to this diagnosis. In addition to describing the images, you must characterize them and approach a diagnostic impression. Although ultrasound is a low-cost and easily accessible tool, it has limitations because it is operator-dependent and because the echogenicity of acute bleeding with the normal splenic parenchyma is easily confounded. On the other hand, CAT is a technique with high sensitivity when the acute bleeding densities are demonstrated in the solid organs, so it is the image of choice for diagnosis until now (7).

The splenic lesion can be classified in degrees: (I) subcapsular hematoma, often in a semilunar form and closely related to the splenic margin; and (II) intraparenchymatous hematoma, which is broader and irregular, with a mass effect and enlargement of the spleen (3). The treatment of choice, regardless of the cause, will generally be splenectomy for several reasons, among which is that the histological diagnosis will be necessary to establish the etiology; on the other hand, as the most common causes are neoplastic the resection treatment will also be curative. Additionally, it is very probable that the function of the spleen is impaired despite conservative surgery of the organ (4)

Conclusion

Non-traumatic rupture of the spleen is a rare disease; this is why there are no guidelines for its management. Therefore, the publication of case series will contribute to the collection of more patients with this pathology to generate articles with greater scientific weight.

A high index of suspicion is required for its diagnosis so that a rapid intervention in the patients is carried out. This improves the prognosis of this entity that has a high mortality rate. The radiologist plays an important role in the early diagnosis of this pathology, taking it into account when evaluating the findings in images in patients with acute abdominal pain of atypical characteristics. CT is a highly sensitive technique and the preferred modality for diagnosis

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