Abstract Pathological or spontaneous rupture of the spleen has been described in a variety of diseases affecting the spleen, with infections being cited as the cause in most cases. In hematological malignancies it is a rare event, despite the frequent involvement of the spleen in these diseases. It has, however, been described in patients with acute and chronic leukemia, Hodgkin’s disease, non-Hodgkin’s lymphoma of B-cell origin, mycosis fungoides, and so-called histiocytic lymphoma. Here, we present a fatal case of splenic rupture caused by infiltration of a peripheral T-cell lymphoma, unspecified according to the REAL classification. The importance of a correct diagnosis and fast surgery is emphasized.

Key words Splenic rupture · T-cell lymphoma · Immunohistochemistry

Introduction

Splenic rupture has been described in a variety of diseases known to affect the spleen, most often in connection with infections, malignant white cell proliferation, or connective tissue diseases [11]. However, being a rare event, it is still not clear what actually leads to the rupture. Furthermore, diagnosis can be very difficult, since symptoms at presentation vary considerably and often resemble other serious intra-abdominal or thoracic visceral damage.

Spontaneous rupture of the spleen in the presence of leukemia or lymphoma is a rare event, although the exact prevalence has yet to be determined. Knoblich [10] presents five examples occurring in patients with lymphatic and myelomonocytic leukemia and three occurring in patients with Hodgkin’s disease, and Oinonen et al. present two cases of splenic rupture in patients with mantle cell lymphoma [13]. A review by Bauer [2] presents five cases of spontaneous splenic rupture in patients with lymphoma or leukemia and reviews 48 previous cases from the literature, in an attempt to identify common risk factors or possible beneficial factors for survival.

The only known cure for splenic rupture and the only factor found to be of importance for the outcome is emergency splenectomy. A survival rate of 78% was reported in a group of hematological patients in which all those who did not have surgery died [2]. A recent review of 136 cases [6] confirms this finding, with a survival rate of 63% after surgery and 40 deaths among 43 patients not operated upon. Recently, splenic rupture has also been described after treatment with granulocyte-macrophage colony-stimulating factor (GM-CSF) in a patient with acute monocytic leukemia [16] and during treatment with G-CSF in the course of autologous stem cell transplantation in another patient with acute myeloid leukemia [9]. Two additional cases of spontaneous rupture of a spleen in primary splenic lymphoma and ‘diffuse histiocytic’ lymphoma have also been described [1, 15].

Regarding T-cell malignancies, only a single case of splenic rupture caused by mycosis fungoides has previously been reported [3]. It was therefore considered worthwhile to describe the present case with an atypical presentation of symptoms and sudden death 3 days after admission to hospital.
**Case report**

A 59-year-old man was admitted to the Department of Hematology with an enlarged spleen, peripheral lymphadenopathy, and pancytopenia. Additional blood tests revealed severe anemia with a slight reticulocytosis (40/1000 erythrocytes), a total white blood cell count of 1.5 x 10^9/l with a normal differential count, except for the presence of 10% abnormal leukocytes, and severe thrombocytopenia (45 x 10^9/l). LDH was elevated to eight times the normal level, while the other liver enzymes were only marginally raised. Similarly, coagulation factors II, VII, and X were marginally decreased. Renal function was normal, and there was no elevation of urate or calcium in the blood. An examination program for disease staging was planned, while a bone marrow aspiration and trephine biopsy were performed immediately.

During the first 2 days after admission the patient complained of mild abdominal pain, and on the evening of the third day he developed more severe, diffuse abdominal pain and shortness of breath. On his way to the bathroom he fainted and slid down to the floor. He was briefly unconscious and woke up with just a small superficial bruise on the right side of his cheek, but otherwise without evidence of injury. During the following hours the patient gradually developed increasingly severe abdominal pain and his blood pressure declined. There was no vomiting and no sign of peritoneal reaction. The abdominal tenderness was diffuse. The patient was by now clearly in distress, pale, and hyperventilating, with diffuse cold sweating, and a condition of shock was developing rapidly. The surgeons suspected a visceral perforation and suggested an abdominal plain film. However, the patient developed a hypovolemic shock with unconsciousness, apnea, and unmeasurable blood pressure. The condition was stabilized by intravenous infusion of human albumin, natrium chloride, adrenaline, and atropine. The patient was transferred to surgery and an explorative laparotomy was performed, revealing large amounts of blood in the retroperitoneum and the spleen enlarged with bleeding cracks. There was no sign of any other intra-abdominal damage. The spleen was removed and homeostasis achieved, but despite transfusion of large amounts of blood, the patient died on the operating table.

Autopsy showed infiltrates of T-cell lymphoma similar to that in the spleen (see below) in the liver, portal nodes, and bone marrow. The stomach and intestines were unremarkable, without evidence of lymphoma. In the liver, the infiltrates were mainly portal in distribution. Sinusoidal infiltrates were minimal, and features of angiocentricity were not seen. The coronary arteries showed moderate atheromatosis, but there was no evidence of myocardial infarction. The death was attributed to splenic rupture due to infiltrates of T-cell lymphoma.

**Pathology**

The spleen weighed 1.5 kg and measured 22 x 16 x 8 cm. In the capsule there were three small fissures in the facies diaphragmatica, and near the hilus of the spleen there was a small area where the capsule was missing. The cut surface was unremarkable. The hilar lymph nodes were moderately enlarged.

Examination of routinely processed, formalin-fixed specimens from spleen and hilar lymph nodes showed effacement of the normal architecture and diffuse infiltrates of large cells with pleomorphic, irregular nuclei with a vesicular structure and one or more nucleoli (Fig. 1). There were scattered mitotic figures. The pleomorphic cells were positive for CD 3, TIA-1, and granzyme B and partly for CD 43. Staining for CD4, CD5, CD8, CD15, CD30, epithelial membrane antigen (EMA), BNH9, and ALK-1 was negative. There was no expression of CD20, CD68, CD79a, myeloperoxidase, or lysozyme.

In the peripheral blood, a slight monocytosis and a few immature erythropoietic cells were seen. Furthermore, there were a few large, pleomorphic lymphoid cells, possibly a small population of circulating neoplastic cells. The bone marrow was hyperplastic with diffuse and interstitial infiltrates of T cells similar to those in the spleen and hilar lymph nodes (Fig. 2).

**Conclusion:** Spleen, hilar lymph nodes, and bone marrow with infiltrates of peripheral T-cell lymphoma with cytotoxic phenotype, unspecified according to the REAL classification [8], and pleomorphic large cells according to the Kiel classification [14].

**Discussion**

This is, to the best of our knowledge, the second reported case of spontaneous splenic rupture due to infiltrates of T-cell lymphoma.

![Fig. 1](image1) **The spleen with a diffuse infiltration of large, pleomorphic cells.** HE, x 400

![Fig. 2](image2) **The bone marrow with an interstitial infiltrate of large, pleomorphic, CD3-positive T cells.** CD3, x 100