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Spontaneous Splenic Rupture in Acute Myeloid Leukemia with Mixed-Lineage Leukemia Gene Rearrangement

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Abstract

Spontaneous splenic rupture is a rare and potentially devastating entity associated with a wide variety of causes including, but not limited to, infections, inflammation, and neoplastic and iatrogenic causes. The clinical presentation of spontaneous splenic rupture is typically acute onset abdominal pain which is non-specific and may be confused with other causes of abdominal pain; failure to recognize this rare entity can result in sever morbidity and mortality. Here we present a 26 year-old obese female with a history of childhood immune thrombocytopenia (ITP), presenting for medical care with flulike symptoms.

Keywords: Spontaneous splenic rupture; Acute myeloid leukemia; Mixed-lineage leukemia gene

Introduction

Spontaneous splenic rupture or, non-traumatic rupture of the spleen, is one of the many causes of acute onset abdominal pain and can result in severe morbidity and mortality of not recognized quickly with an overall mortality rate of 10-15% [1]. Splenic rupture typically presents nonspecifically with left upper quadrant abdominal tenderness, hypotension and syncope; shock and altered mental status may also be observed. Due to a non-specific presentation, and relative rarity, spontaneous splenic rupture can be confused with other abdominal pathology. Spontaneous splenic rupture must be considered in patients presenting with acute abdominal pain even without evident history of trauma, as early recognition and treatment may prevent serious morbidity and mortality.

Case Presentation

The majority of cases of splenic rupture are secondary to trauma; however, spontaneous ruptures can occur. A wide variety of conditions can lead to spontaneous splenic rupture across all ages and include infections, inflammatory conditions, neoplasia, and iatrogenic causes. Spontaneous splenic rupture is a rare and devastating presentation of hematologic neoplasms but has been reported in both plasma cell [2] and acute myeloid [3-8] leukemia. Here we show images of the spleen and bone marrow from a 26 year-old obese female with a history of childhood immune thrombocytopenia (ITP), presenting for medical care with flulike symptoms. A physical examination was unremarkable, and, in particular, her spleen was not palpable. The complete blood count (CBC) and peripheral blood morphological review were notable for a normal white blood cell (WBC) count and differential count and isolated thrombocytopenia with a platelet count of $10 \times 10^3/\mu L$. She was admitted to the hospital and treated with corticosteroids and intravenous immunoglobulin (IVIG) for a presumed relapse of ITP triggered by a viral infection. The following day she developed abdominal pain and collapsed in her room from severe hypotension. A hemoglobin level was 4.4 g/dL (decreased from 11.7 g/dL the day before), and bedside imaging showed extensive free fluid in the abdomen suspicious for intra-abdominal bleeding. She underwent an emergent laparotomy where findings included an engorged spleen, which was fractured at multiple sites and actively bleeding. Postoperatively she was admitted to the intensive care unit (ICU) in critical condition but expired the following day from multiorgan failure due to refractory hypotension, metabolic acidosis and coagulopathy. Pathology evaluation of the spleen showed complete obliteration of the normal architecture (Figure 1A and inset, hematoxylin and eosin (H and E stain) and diffuse infiltration of the red pulp by monocytic precursors that were shown by immunohistochemistry to contain lysozyme (Figure 1B) but not myeloperoxidase (Figure 1C). A post-mortem bone marrow exam showed a hypercellular marrow (Figures 1D, H and E stain) comprised predominantly of monoblasts with round to slightly irregular nuclear contours and abundant vacuolated cytoplasm (Figure 1E, Wright-Geimsa stain). Fluorescence *in situ* hybridization (FISH) analysis identified an atypical MLL/KMT2A/11q23 rearrangement with an extra copy of the 5' MLL/11q23 gene along with two intact MLL/11q23 loci (Figure 1F, arrows). The results of these studies led to revision of the original diagnosis of ITP to acute myeloid leukemia (AML) with a rearrangement in the mixed-lineage leukemia (MLL) gene.

This patient with AML presented with clinical and laboratory features suggestive of ITP. Even in retrospect, the benign history, physical exam and peripheral blood morphology provided no clues to the final diagnosis or to the impending splenic rupture. Acute leukemia with rearrangements in the MLL gene on chromosome 11q23 is generally associated with an unfavorable prognosis and high relapse rate after treatment with chemotherapy alone. A recent study, however, showed improved outcomes with allogeneic hematopoietic cell transplantation [9].

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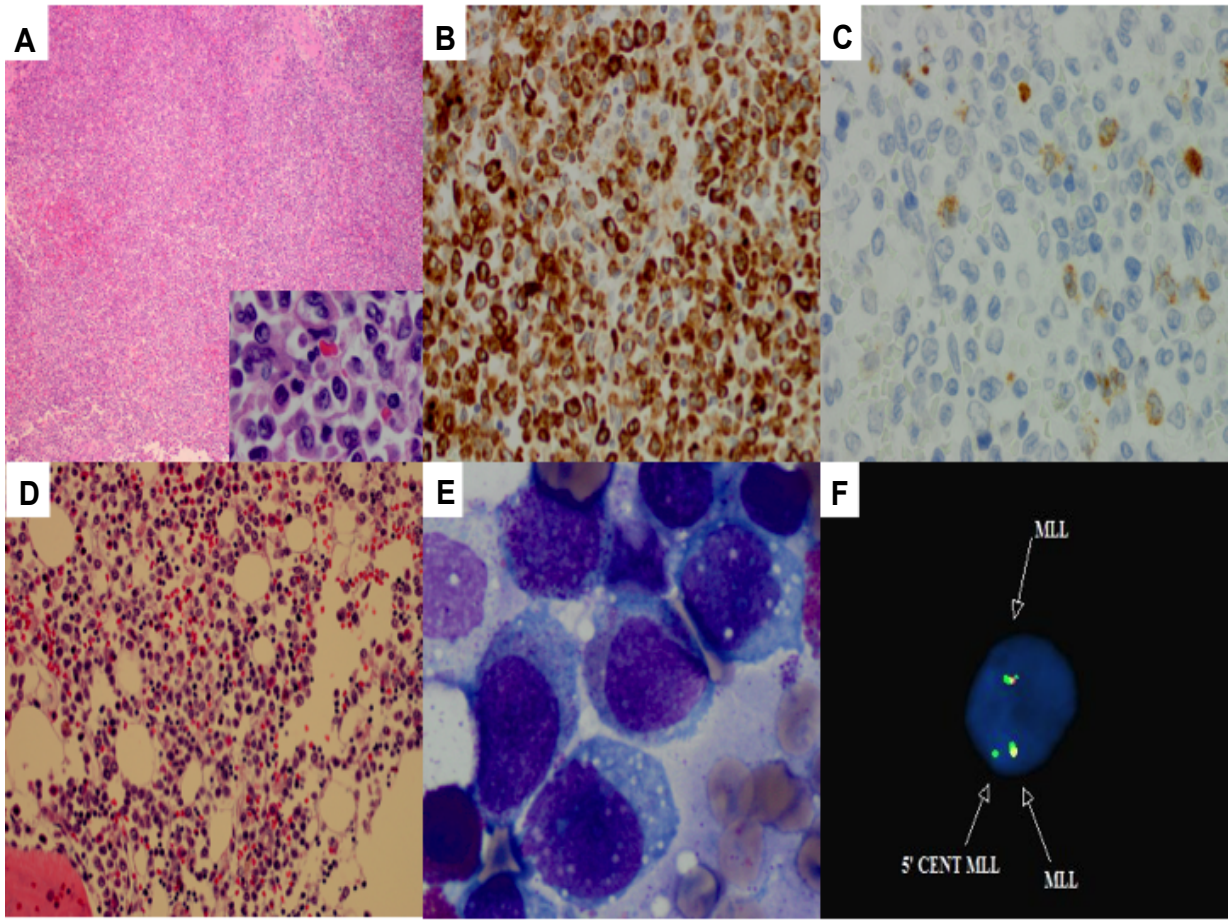

Figure 1: Pathology evaluation of the spleen.

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