Non-Traumatic Splenic Rupture Disguised as Fall Injury
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Citation

Abstract
Most splenic injury and rupture is seen under the guise of blunt abdominal trauma but spontaneous splenic rupture is a phenomenon that also exists and may confound a patient’s initial presentation. Herein we describe a rare case of splenic rupture as the presenting symptom of undiagnosed hairy-cell leukemia. The patient arrived to the emergency department for evaluation of his light-headedness and abdominal pain. CT scan revealed a splenic laceration with large hemoperitoneum. The trauma service was consulted for possible fall injury. He had a history significant for weight loss and light-headedness. His laboratory data revealed pancytopenia and his vital signs suggested that he was in hypovolemic shock. The patient went to the operating room for a laparotomy and splenectomy. Pathologic evaluation of the patient’s spleen and peripheral smear confirmed the diagnosis of hairy-cell leukemia. Although spontaneous splenic rupture is a rare initial presentation of hematologic malignancy, it should still be kept in the differential diagnosis for patients who present with abdominal pain and splenic injury and a questionable history of trauma. Additionally this case illustrates the point that a thorough history and physical can raise the surgeon’s clinical suspicion and potentially diagnose a previously unknown medical condition.

INTRODUCTION
As surgeons we deal with splenic injury routinely. Most splenic injury and rupture is seen under the guise of blunt abdominal trauma. Non-traumatic splenic rupture (NSR) is defined as splenic rupture without antecedent injury. There are a number of case reports and series of patients who had NSR in the world literature and the usual causes are noted to be infection, malignancy, metabolic derangements, vascular disease, and hematologic disorders. The following presents a case of NSR disguised as a fall trauma and also exemplifies the importance of a complete patient history that, when obtainable, can help to point to underlying and undiagnosed disease processes in patients who are initially billed as trauma victims.

CASE
A 48-year-old male presents to the emergency department at a level 1 trauma center with abdominal pain worsening over two to three days and worsening lightheadedness and nausea. He also recounts falling three times in the last 24 hours secondary to coughing fits. The patient arrives diaphoretic and is tachycardic to 124 beats per minute initially with a blood pressure of 107/69 mm Hg. On further questioning it seems that the patient has slowly been losing weight, he has dropped two pant sizes in the last month and his ex-wife says she has noticed at least a 20 lbs weight loss in the last month. On physical exam, he has significant bitemporal wasting, blanched conjunctiva and pale fingernail beds. He also has significant tenderness in the epigastrium and left upper quadrant with involuntary guarding noted but no rebound tenderness. Focused Abdominal Sonography in Trauma exam revealed fluid in the pelvis and bilateral upper quadrants. The patient’s initial complete blood count revealed hemoglobin of 8.1 g/dL, and hematocrit of 23.2 %. Interestingly, he also was neutropenic with a white blood cell count of 2.6 x 10³/mL and had a platelet count of only 54 x 10³/mL (normal limits: 130-400 x 10³/mL). Since the patient was slightly more hemodynamically stable after some fluid resuscitation we proceeded to obtain an abdomen and pelvis CT scan that revealed massive splenomegaly (measuring 23cm), a large subcapsular hematoma and moderate amount of hemoperitoneum (Figure 1). At this point the decision was made to take him to the operating room for a laparotomy.
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Upon entering the abdomen there was a large rush of clot and old blood (approximately 850cc), and on examining the spleen it was noted to be markedly enlarged with a large subcapsular hematoma. All four quadrants were packed and splenectomy was performed in standard fashion. The rest of the abdomen was explored paying close attention to try to identify any enlarged lymph nodes in the mesentery or near the spleen that could have been biopsied, but there were none found. The spleen was found to have a mass of 1.03 kg and there were no other intra-abdominal injuries noted, the patient was closed and went to a monitored bed while his pancytopenia was being worked up. Peripheral smear revealed abnormal mononuclear cells with features of hairy cell leukemia. These findings included round oval nuclei, uniformly condensed chromatin, pale basophilic cytoplasm and well developed circumferentially distributed cytoplasmic projections (Figures 2, 3 and 4).

Figure 1
Figure 1: CT scan revealing splenomegaly, splenic hematoma and hemoperitoneum.

Figure 2
Figure 2: Peripheral smear demonstrating mononuclear cell with “hair-like” cytoplasmic projections (arrow), typical of hairy-cell leukemia

Figure 3
Figure 3: Histologic confirmation of hairy-cells within splenic parenchyma
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Figure 4
Figure 4: Immunostain of CD20 for B-cells. This is strongly positive in the splenic parenchyma, supporting diagnosis of B-cell lymphoproliferative disorder.

The hematology and oncology service was brought on board to help with this new diagnosis and its management, but overall the patient is doing well now his acute process has resolved.

DISCUSSION
NSR has been found to have an incidence of less than 0.5% in patients with no trauma history. There have been several proposed mechanisms to explain this phenomenon but as of yet there is no clear-cut consensus. Some of the pathophysiologic explanations include the possibility of increased intrasplenic tension due to cellular hyperplasia and proliferation. Another proposed mechanism is that the spleen is compressed by the abdominal musculature during physiologic activities that increase intra-abdominal compartment pressure (i.e.: sneezing, coughing, or Valsalva maneuver during defecation). Yet another explanation relies on the idea of vascular occlusion due to reticuloendothelial hyperplasia that leads to thrombosis and infarction of the spleen and subcapsular hemorrhage.

There is another entity that has been reported in the literature that is referred to as spontaneous splenic rupture (SSR). Knoblich distinguished between SSR with known primary disease or cause and idiopathic SSR. There have been several criteria that should be identified to make this diagnosis. The criteria are as follows: (1) no history of trauma or unusual activity that could conceivably injure the spleen, (2) no evidence of disease in other organs that are known to affect the spleen adversely and thereby cause it to rupture, (3) no evidence of perihilar splenic adhesions or scars that could indicate prior splenic trauma or rupture, (4) without findings of hemorrhage or rupture the spleen should be normal on both gross and histological examination, and (5) studies of acute phase and convalescent sera cannot show any rise in viral antibody titers that would suggest a recent infection by viruses that are associated with splenic involvement.

In the setting of hematologic malignancy such as leukemia and lymphoma the pathophysiologic mechanism of splenic rupture has been reported as involving both fragmenting and dissolving of the splenic capsule by invading atypical lymphocytes. The list of risk factors that could contribute to splenic rupture in the setting of hematologic malignancy includes; male sex, adults, splenomegaly and cytoreductive chemotherapy. Some of these criteria almost certainly require a prior diagnosis of malignancy be present. It is noted quite often that in this patient population, pathologic rupture of the spleen can occur unexpectedly, without antecedent event, and can present with new onset of acute abdominal pain, tachycardia and hypotension. However, there are only a handful of scattered case reports present that make note of splenic rupture being the first presenting symptom of a hematologic malignancy and they also draw the conclusion that this is possibly a sign of poorer outcome.

Review of the case series presented by Bauer et al. found that, for splenic rupture in patients with underlying hematologic malignancy, 40% of these patients had acute leukemia, 27% had Hodgkin’s or non-Hodgkin’s lymphoma and 18% had chronic leukemia. In patients presenting with severe abdominal pain, distention and signs of hemodynamic shock, it has been proposed that the entity of NSR be kept in the differential and that surgeons be alerted early to evaluate the possible need for laparotomy.

In the preceding case, this patient was billed as a fall trauma, as the patient recounted history of lightheadedness and fall with coughing fits in the prior 24 hours. The actual series of events was likely that the patient suffered an NSR secondary to his massive splenomegaly and underlying undiagnosed hematologic malignancy, and developed a slow bleeding after the splenic capsule was ruptured. Then, over the next several days, he became more hypovolemic and hypotensive leading to his lightheadedness and falls. That is why on presentation he was showing signs of hypovolemic shock, and on laparotomy he had a large volume of old blood identified. The patient had no other signs of traumatic injury noted either externally or internally. It is important for the trauma surgeon being consulted to perform a thorough history and physical that can help to explain or point to a
possibly undiagnosed medical problem, as was discovered in this patient. Although NSR and SSR are both rare entities they should be kept in mind when forming a differential diagnosis for a patient who has splenic rupture and a questionable history of trauma.

References

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