

A Traumatic Splenic Rupture: Dreaded Complication of Splenomegaly

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Abstract

We discuss the causes of splenomegaly and the underlying abnormalities leading to the most feared complication of a traumatic splenic rupture. A case discussion is included.

Keywords: Splenomegaly; Traumatic splenic rupture

Case Presentation

A 21 year-old man presented to the Emergency Department (ED) with the chief complaint of abdominal pain that radiated to his left upper back and worsened with deep inspiration. The patient was nauseous and had vomited twice. He had a sore throat for approximately one month and had had a negative rapid streptococcal test at his student health center. He had been treated empirically for streptococcal pharyngitis at his university student health center within the previous two weeks with amoxicillin. He did not improve with the amoxicillin, but developed a diffuse maculopapular rash; his prescription was changed to clarithromycin for his persistent sore throat and he was given prednisone for his rash.

The patient denied any history of trauma as his symptoms began when he got out of bed earlier that morning. The patient appeared pale, felt clammy and was hypotensive at 95/60 mmHg and tachycardic with a heart rate of 123 bpm. He was a febrile, with an oral temperature of 97.9°F. On exam of the patient's abdomen he was found to have voluntary and involuntary guarding as well as rebound tenderness. A bedside ultrasound of the abdomen was found to have free fluid in the abdomen and an enlarged spleen with a large amount of complex fluid adjacent to the spleen; these findings were thought to represent hemorrhage from a ruptured spleen. Initial lab values were a white blood cell count of $22 \times 10^3/\text{mm}^3$, a hematocrit of 0.37, and platelets of $313 \times 10^3/\text{mm}^3$.

A surgical consult was obtained and the patient was taken emergently to the operating room. On opening the abdomen, a large amount of blood was appreciated. The spleen was found to be extremely enlarged with rupture of the capsule and active bleeding, so that a splenectomy was performed. No other abnormalities were found in the abdominal cavity. Pathologic examination of the spleen revealed that it was $14.0 \times 12.0 \times 3.5$ cm and weighed 630 grams; the capsule was intact except for the hilar area, which had an associated hemorrhage. Extensive immunoblastic hyperplasia was noted, and immunoperoxidase stain for latent membrane protein (LMP-1) was positive in scattered cells; in situ hybridization for Epstein-Barr virus (EBV) was positive. These findings were consistent with the diagnosis of a ruptured enlarged spleen secondary to acute infectious mononucleosis. The patient experienced a full recovery and was discharged home on hospital day four.

Discussion

Spontaneous splenic rupture has been reported as early as 1874 and was considered *Idiopathic* since there was no prior trauma except *violent retching...after eating some herring* [1]. It is a misleading term which would be better called atraumatic/non-traumatic or pathologic

rupture since it almost always occurs in a diseased spleen due to an underlying pathologic process. A true spontaneous rupture that would occur without trauma and in a normal spleen is considered to be exceptionally rare and is still debated if it indeed occurs [2-4]. Even with splenic injuries from blunt trauma there has been found abnormal lymphocyte populations that may have contributed to the risk of rupture requiring splenectomy [2]. The definition of a truly spontaneous rupture of the spleen in 1958 included: no antecedent trauma, no disease state with primary or secondary effects on the spleen, no adhesions or scarring of the spleen, and for it to be normal macroscopically and microscopically [5].

Anatomy and Physiology

The spleen is the largest of the lymphatic organs, and has among its functions filtration from the blood stream of all foreign matter, including damaged or parasitized blood cells, as well as it is a major site of erythropoiesis and hematopoiesis [6]. In a healthy adult it weighs between 75 and 250 g but decreases in size with age; it is usually 9-12 cm in length (cranio-caudal measurement) [4,7]. It lies beneath the 9th to the 12th thoracic ribs and is supplied by the splenic artery which is notable for its large size and tortuosity. The spleen consists of a capsule that is 1-2 mm thick and trabeculae that enclose the pulp. Blood passes from the splenic arteries into central arteries, which give rise to numerous arterioles that take off at right angles from the central artery. This perpendicular arrangement causes plasma to be "skimmed" from the blood flow; most red blood cells pass into the red pulp. Within the red pulp blood is collected in splenic sinuses, large, thin-walled spaces that drain into the pulp veins and then into the main splenic veins to enter the portal circulation. The tissue between the splenic sinuses is a reticular connective tissue network known as the splenic cords, or the cords of Billroth. At points of passage from cords to sinuses maximum deformability and flexibility are demanded of red blood cells so that they can squeeze through slits in the cord lining. The marginal zone separates the red pulp from the white pulp which contains follicles of B lymphocytes, named Malpighian corpuscles, and germinal centers of

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Flemming surrounding the T lymphocyte rich per arterial lymphatic sheaths [7-9].

In the presence of splenomegaly, blood flow through the spleen slows, becomes more circuitous, encounters more obstacles, and results in pooling of cells within the cords of Billroth. Erythrocytes have a lifespan of approximately 120 days; the spleen removes about 20 mL of aged red blood cells per day; with splenomegaly, greater accumulation of erythrocytes in the spleen may result in a hemolytic anemia. Neutrophils have a half-life of about 6 hours; in some hypersplenic states the removal of neutrophils is augmented and may result in neutropenia. One-third of the total platelet pool is normally contained in the spleen, but with splenomegaly up to 80% of the total platelet population may be sequestered in the spleen and results in thrombocytopenia [7,10]

Splenomegaly

Splenomegaly has traditionally been diagnosed by the physical exam finding of having a palpable spleen but in one study 16% of patients with such were found to have normal sized spleens by radiologic evaluation of either computerized tomography (CT) scan or ultrasound; physical exam alone is also not very sensitive as up to 30% of patients with acute mononucleosis due to EBV infection thought to have normal splenic size by exam were determined to have splenomegaly by imaging [7,10]. Splenomegaly is defined as cranio-caudal length greater than 13 cm by ultrasound [7]. Massive splenomegaly has previously been defined as a spleen that crossed the midline and reached the iliac crest but also can be diagnosed by CT scan measuring the splenic index (length×width×depth) greater than 120 cm³, or having a weight greater than 1500 g [7,11,12]. Splenomegaly was found in 0.3% of hospital admission in the U.S. between 1963-1965; an underlying diagnosis was found in 98% of cases but 12% required splenectomy in order to so [7].

Diseases Associated with Splenomegaly

Epstein-Barr virus is a gamma-group herpesvirus (human herpesvirus-4) that usually causes a self-limited disease characterized by the triad of fever, generalized lymphadenopathy, and sore throat [13]. Symptomatic infection peaks at age 15-24 years and up to 30% of college students are affected each year [10]. Incubation is approximately 4-6 weeks followed by a prodrome of fatigue, malaise, and myalgias for 1-2 weeks. Splenomegaly is usually most prominent in the second and third weeks of infection due to proliferation of mononuclear cells within the spleen and occurs in approximately 60% of patients. Although the overwhelming majority of these patients with splenomegaly are asymptomatic, splenic rupture occurs in 0.06-0.5% of cases and is the #1 cause of death from infectious mononucleosis [10,13]. The atypical lymphocytes that are characteristic of the disease are virus-specific cytotoxic T cells and suppressor T cells; these are enlarged cells with an increased amount of cytoplasm, vacuoles, and have indentations of the cell membrane. These atypical lymphocytes heavily infiltrate the spleen and, on histological examination, cause a prominence of splenic follicles and a blurring of the splenic architecture. Pleomorphic blast cells are present in the hyperplastic red pulp. Vascular congestion is coupled with focal and subcapsular hemorrhages. The spleen is very vulnerable to rupture secondary to infiltration of the trabeculae and capsule by atypical lymphocytes. Most ruptures occur by day 21 of illness and very few occur after day 28, so that return to sports should be at least four weeks from onset of illness [10]. The mechanism of rupture is thought to involve two mechanisms: 1) expanding subcapsular hematoma that eventually tears the capsule; 2) valsalva maneuver (cough, sneeze, vomit, or even turning over in

bed) increasing the portal venous pressure in an already engorged spleen accompanied by forceful diaphragmatic or abdominal wall contraction causing sudden compression of the spleen [10,13].

In general, there are four major groups of diseases that may cause splenomegaly: 1) those that cause increased demand for splenic function (reticuloendothelial system hyperplasia for removal of defective erythrocytes, extramedullary hematopoiesis, and immune hyperplasia), 2) abnormal splenic or portal blood flow (cirrhosis), 3) infiltration of the spleen (intracellular or extracellular depositions and benign or malignant cellular infiltrations), and 4) those of unknown etiology (idiopathic splenomegaly). The most common causes of splenomegaly in the U.S. are hematologic, hepatic disease and infectious [7] (Table 1). However, massive splenomegaly is less common with the vast majority of patients having non-Hodgkin's lymphoma, chronic lymphocytic leukemia (CLL), chronic myelogenous leukemia (CML), hairy cell leukemia, myelofibrosis with myeloid metaplasia, polycythemia rubra vera, essential thrombocythemia, and Gaucher disease [11]. In the tropics, the most common causes of massive splenomegaly include malaria (hyper-reactive malaria), schistosomiasis, and visceral leishmaniasis [14].

Splenomegaly is common in those patients with cirrhosis due to severe portal hypertension; these patients may exhibit a profound thrombocytopenia and leukopenia. In the absence of cirrhosis, splenomegaly and variceal bleeding indicate splenic or portal vein thrombosis [4].

Malaria commonly causes splenomegaly in endemic areas; splenic enlargement due to hyperplasia of the white pulp is likely due to repeated infections with the parasite, especially infections that have been treated multiple times in chloroquine-resistant areas and has a high mortality [6,15]. Sub acute bacterial endocarditis may cause splenomegaly as well as severe left upper quadrant pain and pleuritic chest pain secondary to thromboembolic (potentially septic) occlusion of splenic blood flow. Approximately 25-60% of all patients with endocarditis develop splenomegaly if the disease is of long duration [16].

Early sickle cell crisis prior to auto infarction may also cause splenomegaly and abdominal pain however, this is usually present in African-American children before the age of 10. Splenomegaly persists into adulthood in 10% of sickle-cell patients in the West Indies and is associated with high levels of fetal hemoglobin as well as sickle-hemoglobin C disease and sickle-Beta Thalassemia [12]. Massive splenic infarction can occur in these patients often precipitated by high altitude, acute chest syndrome or sepsis, and may require urgent splenectomy due to severe complications such as rupture, splenic abscess or pseudocyst formation [12]. Splenomegaly is most commonly asymptomatic in Sarcoidosis but when massive (3% of patients) it also can be a cause of massive splenic infarction [17].

Splenic Rupture

The recognized categories of etiologies of non-traumatic splenic rupture [1,3] (Table 2).

Infection	EBV, viral hepatitis, TB, HIV, malaria, endocarditis
Hematological	lymphoma, CML, CLL, hemoglobinopathies, myelofibrosis
Congestive	cirrhosis, heart failure
Inflammatory	Lupus, rheumatoid arthritis, Sarcoidosis
Neoplastic	hemangiomas, metastases
Infiltrative	Gaucher's, amyloidosis

Table 1: The recognized common etiologies of splenomegaly.

Infectious	Viral EBV (#1 cause in the US), CMV, Influenza, HIV, Dengue Bacterial Tuberculosis, Typhoid Fever, Syphilis Parasitic Malaria, Schistosomiasis, Leishmaniasis, Echinococcal cysts
Neoplastic	Non-Hogkins Lymphoma, AML, Immune Thrombocytopenic Purpura, Angiosarcoma (most common primary splenic malignancy), metastases
Inflammatory	chronic pancreatitis, Polyarteritis Nodosa, Lupus, Amyloidosis
Congenital	hemangiomas
Vascular	portal hypertension, pregnancy
Iatrogenic	anticoagulation, thrombolytics, G-CSF, colonoscopy

Table 2: The recognized etiologies of non-traumatic splenic rupture.

Evaluation

If splenomegaly is suspected by symptoms (left upper quadrant pain, fullness or early satiety) or physical examination, then radiographic evaluation should be performed in order to confirm splenic size, delineate splenic architecture, determine if it is focal lesions or diffuse enlargement, and for assessment of other potentially involved organs (liver, visceral lymphadenopathy) [7,16]. Laboratory tests should generally include complete blood counts, peripheral blood smear, coagulation studies, erythrocyte sedimentation rate (ESR), chemistry panel, liver function, anti-nuclear antibody (ANA), Rheumatoid Factor, urinalysis, and serology for Hepatitis B and C, human immunodeficiency virus (HIV), EBV, and cytomegalovirus (CMV) [7].

Treatment

Specific treatments for the underlying disorders causing splenomegaly are beyond the scope of this paper which will focus on the treatment of splenic rupture. Attention must first be paid to immediate stabilization and resuscitation of possible hemorrhagic shock due to intra-abdominal bleeding. At least two large-bore intravenous catheters should be placed for crystalloid and blood product administration. A bedside ultrasound evaluation is the preferred test for an unstable patient to detect hemoperitoneum but a diagnostic peritoneal lavage is an option for an experienced provider if ultrasound equipment is not available. A stable patient can have CT scanning of the abdomen performed. Urgent surgical consultation is warranted when the diagnosis is confirmed or highly suspected [2]. Although there has been an increase in the successful non-operative management of hemodynamically stable patients with blunt splenic rupture, total splenectomy is still recommended for patients with pathologic splenic rupture [2,13]. Observation, angioembolization, and splenorrhaphy have unacceptable risks since the underlying process that caused the initial rupture are still present so that the chance of re bleeding is quite high.

In summary, many disease processes may predispose patients to splenomegaly and subsequent splenic rupture, and although it is a rare occurrence, if not recognized and treated promptly the consequences are devastating. The definitive therapy is surgical splenectomy; immunizations against encapsulated organisms (streptococcus, meningococcus) should be administered prior to hospital discharge to minimize risk of subsequent overwhelming sepsis.

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