

Spontaneous Splenic Rupture in Infectious Mononucleosis

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Spontaneous splenic rupture is an uncommon complication associated with infectious mononucleosis. It was first described by Rokitansky in 1861, and several cases have been cited in recent years. Though it only occurs in a minority of patients with infectious mononucleosis, misdiagnosis can lead to death. The absence of a traumatic injury elicited from a patient's history does not rule out the diagnosis of a ruptured spleen. This article presents the case of a young woman who presented with spontaneous splenic rupture secondary to infectious mononucleosis. A review of this case as well as of the current literature underscores the importance of prompt identification and treatment of this entity. This article also emphasizes the importance of the unique management of the postsplenectomy patient.

CASE PRESENTATION

Initial Presentation and History

A 17-year-old Caucasian female presented to the emergency department (ED) complaining of malaise, sore throat, fever, and nonproductive cough for several days, with increasing abdominal pain. The patient reported vomiting and retching prior to coming to the ED, and this was followed by diffuse abdominal pain with radiation to her left shoulder. There was no history of trauma. The patient's past medical history was significant for a "mono-like" illness 3 years earlier. There was no history of allergies. The patient denied alcohol or tobacco use. The patient denied sexual activity. She rated her initial pain as 5 on a pain scale of 1 to 10, which increased to 10 over the previous 24 hours.

Physical Examination

On physical examination, the patient was an age-appropriate female weighing approximately 100 lb. Her initial temperature was 98.0°F. The patient was tachycardic with a heart rate of 120 bpm and an initial blood pressure of 100/60 mm Hg, which decreased to a nadir of 88/55 mm Hg while in the ED. Respiratory rate was 22 breaths/min. She was awake and alert but pale. Her pupils were equal and reactive to light with injected conjunctivae. The ear and nasal examinations were unremarkable. The oropharyngeal exam was significant for erythema. Cervical adenopathy was palpable. The patient had normal breath sounds without

any wheezing, rales, or rhonchi. Upon auscultation, the patient had a regular cardiac rate and rhythm with no murmurs appreciated. The patient was noted to have a slightly distended abdomen with diffuse tenderness and guarding with hypoactive bowel sounds. Inspection of the back revealed no costovertebral angle tenderness. The neurologic examination did not reveal any focal deficits. Examination of the patient's skin revealed diffuse erythema on the chest wall.

Laboratory Results

Laboratory testing revealed leukocytosis with a white blood cell (WBC) count of 20,250 cells/ μ L and the following differential: neutrophils, 43.5%; lymphocytes, 41.4%; monocytes, 2.3%; eosinophils, 0.4%; basophils, 1.6%; leukocytes, 10.8%; band neutrophils, 20%. Serum levels of aspartate aminotransferase and alkaline phosphatase were 156 U/L and 172.4 U/L, respectively. A quantitative human chorionic gonadotropin (hCG) test showed an hCG level less than 0.5 mIU/mL. Hemoglobin initially was 11.5 g/dL and several hours later decreased to 8.1 g/dL. Urine analysis showed 5 to 9 WBCs/hpf. Hepatitis B core antibody test was non-reactive, and hepatitis B surface antigen was negative.

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Figure 1. Abdominal computed tomography scan showing an enlarged spleen with heterogeneous enhancement.

The hepatitis B surface antibody level was greater than 1000 mIU/L. A hepatitis C antibody test was nonreactive at 0.16. The C reactive protein level was 0.66 mg/dL and the erythrocyte sedimentation rate was 20 mm/hr. Epstein-Barr virus (EBV) IgG antibody was positive at a level of 8.38 AU/mL. Cytomegalovirus IgG was negative at 0.24. Computed tomography (CT) scan of the abdomen (**Figure 1**) revealed a prominent spleen measuring 16 cm with heterogeneous enhancement consistent with splenic laceration.

Hospital Course

The patient underwent emergency laparotomy with splenectomy for a ruptured spleen. The gross pathology (**Figure 2**) revealed a ruptured and hemorrhagic spleen weighing 428 g. Microscopic examination revealed reactive lymphoid hyperplasia consistent with a history of infectious mononucleosis. A subsequent heterophile antibody test performed on a blood sample collected upon admission was positive for mononucleosis. The patient was stabilized in the intensive care unit and transferred to the general medical floor in stable condition.

DISCUSSION

Infectious Mononucleosis and Splenomegaly

Infectious mononucleosis, a herpes group viral infection, is caused by EBV. EBV infection is often a mild and nonspecific febrile illness in young children, while typical symptomatic “mono-like” illness is more common in young adults. The illness often lasts for 1 to 4 weeks. Symptoms commonly include fever, sore throat, and lymphadenopathy after a 4- to 7-week incubation. Splenomegaly occurs as a result of mononuclear lymphocyte infiltration of the splenic tissue. This splenomegaly is accompanied by a risk of rupture of the weakened

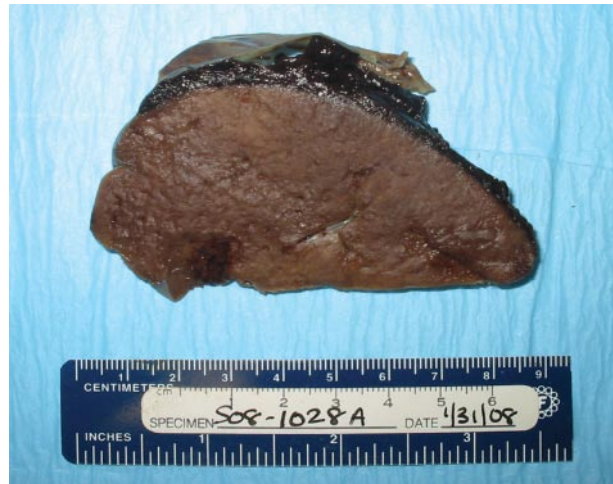


Figure 2. Gross pathology specimen revealing enlarged spleen with hemorrhage. (Photo courtesy of Gregory Palermo, MD, Department of Pathology, JFK Medical Center, Edison, NJ.)

splenic capsule; when rupture occurs, it is usually during the second or third week of the illness.¹ In fact, no documented reports of rupture have occurred after the first 3 weeks of symptom onset.² Nevertheless, patients are often advised to refrain from vigorous physical activity for 4 weeks after the initial diagnosis is made.³ With splenic rupture, a history of abdominal trauma is often revealed in the patient’s history.⁴ Spontaneous splenic rupture from infectious mononucleosis is the most common lethal complication of the illness and occurs in only 0.1% to 0.5% of patients.⁵ Estimates of mortality due to rupture vary from 30% to 100%.⁶

The high mortality associated with spontaneous splenic rupture seems to be secondary to a failure to diagnose the condition, as “atraumatic” rupture can be an elusive diagnosis.⁷ The actual incidence of spontaneous rupture is debated, and some have suggested that most cases of rupture are in fact spontaneous.² It has been hypothesized that spontaneous rupture occurs as a result of increased portal pressure associated with a valsalva maneuver. The contraction of the abdominal muscles during vomiting or coughing may induce rupture.⁶ Thus, there can be a hidden “traumatic” injury to the spleen that may not be obvious from a patient’s history and the label “trauma” may not be applied. The term *atraumatic rupture* may therefore be a misnomer.

Atraumatic splenic rupture was first documented in 1861 by Rokitansky. One analysis using the criteria set by Rutkow referenced over 8000 patients with mononucleosis and found only a handful of cases of atraumatic rupture of the spleen.⁸ These patients all had

common characteristics based on Rutkow's criteria, including a negative history of trauma, serologic evidence of mononucleosis, clinical symptoms of mononucleosis, as well as histological evidence consistent with the diagnosis. The degree of splenic enlargement in such cases varies. Normally, a spleen weighs about 200 g but in the setting of mononucleosis may enlarge to over 750 g.⁹

Presentation of Spontaneous Splenic Rupture

The presentation of spontaneous splenic rupture is variable. Commonly patients first have the typical prodrome of fever and pharyngitis seen with infectious mononucleosis as well as palpable splenomegaly. In rare instances, the rupture itself is the initial presentation of illness.¹⁰ One review of 55 cases revealed that only half of reported cases of rupture were associated with a palpable spleen on exam.³ In one study of patients with ultrasonographically identified splenomegaly, physical examination detected the finding in only 20% of patients. The characteristic abdominal pain radiating to the left upper extremity or shoulder, known as Kehr's sign, may also be an indication to suspect splenic rupture.¹ CT scan is often essential to making the diagnosis and may reveal splenomegaly with lacerations of the spleen and intraperitoneal or subcapsular bleeding.¹⁰ Characteristic laboratory data will show leukocytosis and anemia. In severe cases, the pain is often accompanied by signs of hypovolemia such as orthostasis and tachycardia secondary to hemorrhage.

Treatment

Splenectomy has historically been the treatment of choice in hemodynamically unstable patients. However, conservative treatment in those who are more stable may avoid complications such as postsplenectomy sepsis.¹¹ Selective splenic artery angiography with embolization, for example, may be used in lieu of surgery.¹² Patients who undergo conservative measures, however, should be selected carefully for certain criteria, including hemodynamic stability.¹³

Postsplenectomy Sepsis

Postsplenectomy sepsis (PSS) is a dreaded complication of splenectomy. The risk is highest in the first few years after the procedure. PSS may occur more commonly in childhood but is not limited to any particular age-group.¹⁴ The younger the patient, the shorter the interval to PSS is. Splenectomy secondary to trauma has the lowest risk for PSS, compared to splenectomy secondary to thalassemia, which has the highest risk. *Streptococcus pneumoniae* is the most important organism

implicated in PSS.¹⁵ Other organisms include *Haemophilus influenzae*, *Neisseria meningitidis*, and *Capnocytophaga canimorsus*.¹⁶ Early postoperative infections after splenectomy are caused by the usual organisms seen in other similar surgeries.

Therapeutic strategies for PSS include immediate self-treatment. Despite the absence of controlled studies, self-administration of antimicrobials at the first sign of suspicious illness is commonly recommended. Choices based on drug allergy include a quinolone, amoxicillin/clavulanic acid, trimethoprim/sulfamethoxazole, or a newer macrolide. Self-treatment should be promptly followed by clinical evaluation.

In terms of prophylactic antimicrobials, there is no statistically significant difference in the incidence of pneumococcal events among groups with and without prophylactic antimicrobials.¹⁷ However, recurrent episodes of PSS or perhaps even one episode of PSS can be an indication for lifelong prophylaxis. Immunization against pneumococci using the 23-valent unconjugated capsular pneumococcal polysaccharide vaccine (PPV23) is recommended following splenectomy. The Centers for Disease Control and Prevention recommend a one-time PPV23 revaccination after 5 years.¹⁸ Also recommended is either the conjugate meningococcal vaccine (MCV4) or the meningococcal polysaccharide vaccine (MPSV). Poor patient education about PSS increases the risk, and repeated instruction on follow-up visits is essential. Patients are advised to carry an alert card documenting asplenia, immunization, prophylaxis, and emergency plan.

SUMMARY

Spontaneous or "atraumatic" splenic rupture is rare in patients with mononucleosis but potentially fatal, often due to misdiagnosis. Consequently, atraumatic splenic rupture should be detected as early as possible using clinical, laboratory, and radiographic means. Hemodynamically unstable patients are often treated with splenectomy. When splenectomy is employed, the patient must be educated about the risk for PSS. **HP**

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