An Autopsy Case of Spontaneous Splenic Rupture Due to Isolated Splenic Peliosis and Review of the Literature

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ABSTRACT

Peliosis is a rare entity that is characterized by cystic or blood-filled spaces in the parenchyma of solid organs. It is most commonly seen in the spleen in patients who also have peliosis hepatis. Isolated splenic peliosis is very rare and spontaneous rupture due to splenic peliosis is even more uncommon. This diagnosis has high importance in forensic pathology as it is a rare cause of atraumatic splenic rupture. We present a case in which the diagnosis of isolated splenic peliosis with massive hemoperitoneum was made at autopsy. *Acad Forensic Pathol.* 2016 6(2): 331-337

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INTRODUCTION

Peliosis is a rare disease characterized by cystic or blood-filled spaces, usually in the parenchyma of solid organs, that belong to the mononuclear phagocytic system such as the spleen, liver, lymph nodes, and bone marrow. Peliosis has also been described in other organs such as the lung, the parathyroid glands, the pancreas, and the pituitary gland (1-4). Peliosis is often associated with underlying immunodeficiency, malignancy, infections, steroid use, hemodialysis, or other conditions (5). Although peliosis in the spleen is usually seen in conjunction with peliosis in the liver, isolated splenic peliosis can rarely occur. Typically, splenic peliosis is an incidental autopsy finding. However, this entity can lead to atraumatic rupture of the spleen and death; in forensic pathology it can be easily confused with traumatic rupture. We present a case in which the diagnosis of isolated splenic peliosis with massive hemoperitoneum was made at autopsy.

CASE REPORT

A 45-year-old male with a past medical history of Hepatitis C virus infection and an unknown seizure disorder complained of severe left lower quadrant abdominal pain and dizziness. Three hours after the symptoms started, he was found to have mild anemia (Hgb 9.3 g/dL), normal platelet count, normal coagulation studies, and a negative fecal occult blood test. Approximately 24 hours later, he suffered a cardiopulmonary arrest, was taken to the hospital, and could not be resuscitated.

FINDINGS

External examination at autopsy revealed pale buccal membranes and conjunctivae, a flat abdomen with a periumbilical blue green discoloration, and no contusions or injuries in the skin or soft tissue of the abdominal wall. Upon internal examination, the abdominal cavity contained 4400 mL of liquid blood and blood clots. There were no peritoneal adhesions, hematomas in the retroperitoneum or mesentery, or no rib fractures on the left side. The spleen was enlarged, weighed 560 g, and showed a subcapsular hematoma

along the antihilar aspect (**Image 1**). There was also a tear along the superoposterior portion of the capsule, with focal involvement of the hilum. The parenchyma was soft, pale, and friable, with multiple well-circumscribed, unevenly distributed blood clots ranging from 0.3 cm to 5.5 cm with branching, tubular, angiod shapes (**Image 2**). Grossly, no cyst lining was noted. The liver surface appeared to be slightly granular with rounded edges, and the cut surface showed a light brown, homogenous lobular pattern with a firm consistency. There were no cystic lesions identified in the liver.

Image 1: Macroscopic appearance of the enlarged spleen with subcapsular hematoma and capsular tear.

Microscopic sections of the spleen showed extensive areas of hemorrhage involving the white and red pulp (**Image 3**). There were multiple lakes of blood of different sizes with an uneven distribution. The lakes of blood did not have a cell lining or associated desmoplasia (**Images 4 and 5**). The uninvolved white and red pulp showed sinusoidal dilatation and congestion, but were otherwise unremarkable. No atypical lymphocytes, vascular proliferation, or organisms were

identified on routine stains, confirming the diagnosis of peliosis and ruling out neoplasia or infection. Microscopic sections of the liver showed moderate to severe bridging fibrosis of the portal tracts with occasional nodule formation (ISHAK fibrosis stage: 4-5/6) and mildly active chronic hepatitis (MHAI: 5/18) (6), most consistent with Hepatitis C virus infection. The liver did not show any evidence of peliosis. There were no anatomic signs of portal hypertension or se-

Image 2: Serial sections of the spleen showing multiple cyst-like and angiod lesions with an uneven distribution pattern, subcapsular blood collection, and normal splenic parenchyma.

Image 3: Low power view of the spleen showing multiple lakes of blood and a small area of normal splenic parenchyma (H&E, x8).

Image 4: Medium power view of the spleen showing a well-demarcated pool of blood (H&E, x80).

vere liver disease such as ascites, peripheral edema or other signs of volume overload, gynecomastia, testicular atrophy, caput medusa, hemorrhoids, or esophageal or gastric varices.

DISCUSSION

Peliosis in the spleen is usually seen in conjunction with peliosis in the liver; however, isolated splenic peliosis can rarely occur. Isolated peliosis of the spleen is a rare entity associated with many underlying diseases and risk factors. These include Hepatitis C virus infection, liver cirrhosis, tuberculosis, AIDS, post-transplant immunodeficiency, intravenous drug abuse, chronic alcoholism, androgenic steroid therapy, corticosteroid therapy, oral contraceptives, advanced malignancies including hematologic malignancies, and hemodialysis. To date, spontaneous rupture of the spleen due to peliosis has been previously described few times in the literature (7-16). Peliosis of the spleen has characteristic gross appearance of cystic blood-filled spaces with well-defined margins, commonly involving the red pulp, distributed in a sporadic, clustered, or disseminated pattern. This unique

gross appearance provides an immediate clue to the pathologist as to the diagnosis of splenic peliosis. Histologically, the cystic spaces may lack a cell lining or be focally lined by endothelium. At autopsy, clinical history and initial findings might point to a traumatic death and having this differential diagnosis in mind is very important.

The pathogenesis of peliosis is unknown. It has been proposed that peliosis hepatitis results from loss of the integrity of the microvasculature of the liver, either due to disruptions in the microcirculation such as alteration in local intravascular pressure or congenital malformation in the vessels. It is thought that damage to the microvasculature of liver, along with additional unknown factors, is responsible for the development of peliosis hepatitis (17). An article published in 2005 proposes that peliotic lesions are venous malformations that have been created by alterations in local intravascular pressure conditions in the spleen, a similar pathogenesis to peliosis hepatitis (5). It is also possible that there are multiple pathogenic mechanisms giving rise to this disease since it has been associated with multiple conditions and risk factors.

Image 5: High power view of a cyst-like lesion filled with blood showing lack of endothelial cell lining (H&E, x400).

Microscopically, peliosis hepatis has been described as having two morphologic patterns: 1) blood-filled spaces that are not lined by endothelium (parenchymal) and 2) centrilobular blood-filled spaces that are lined by endothelium and have associated fibrosis (phlebactatic) (18). Senf proposed that these two patterns are observed in different stages of the disease (13). It is thought that the disease starts with a hemorrhage into a necrotic area in which the lesion will not have a lining. Later, as the disease progresses, there will be an endothelial lining around the initial area of hemorrhage (19).

Differential diagnoses to consider in these cases are traumatic splenic rupture, infectious mononucleosis, hemangiomas, and hairy cell leukemia (**Table 1**). Traumatic spleen rupture is always a possibility that needs to be considered and ruled out based on history, scene findings, and other autopsy findings. Infectious mononucleosis has a typical clinical picture of sore throat and fever in a child or young adult, and can rarely cause spontaneous splenic rupture. At autopsy, there would be splenomegaly with hematoma and

the microscopic sections will show atypical immunoblasts with red pulp expansion. Hemangiomas would present as vascular channels with plump endothelial cell lining. Hairy cell leukemia would show a proliferation of pale small lymphocytes in the red pulp, tumor cells invading subendothelial lining, and “lakes” of blood lined by abnormal lymphocytes.

Splenic peliosis should be considered by the pathologist in cases of splenic rupture with no history or signs of trauma, and the characteristic cyst-like blood-filled spaces in the splenic parenchyma. In this case of unexpected hemoperitoneum due to splenic rupture, the decedent was carefully examined for signs of trauma without any found. With the clinical history of Hepatitis C infection and fibrosis in the liver, splenic rupture due to portal hypertension was considered (20). However, available history and laboratory studies did not suggest decompensated liver disease, nor did autopsy reveal any anatomic signs of portal hypertension. Upon serially sectioning of the spleen, the well-circumscribed, unevenly distributed, round and angiod shaped blood clots were very striking. The gross find-

Table 1: Differential Diagnosis of Splenic Rupture

Pathologic Conditions	Clinical Presentation	Gross Findings	Microscopic Findings
Splenic Peliosis	Usually incidental finding in imaging; can present as acute abdomen due to splenic rupture Abdominal pain, anemia, altered mental status	Splenomegaly, cystic blood-filled spaces in the parenchyma with well-defined margins, commonly involving the red pulp, distributed in a sporadic, clustered, or disseminated pattern	Cyst-like spaces filled with blood; might be endothelial lining or no cell lining at all
Traumatic Splenic Rupture	History of blunt trauma to the abdomen Abdominal pain, anemia, altered mental status	Hematomas, fractures, capsular tear, often the superior pole or hilum	Neutrophils below capsular tear, often in superior pole or hilum
Infectious Mononucleosis	Child or young adult with sore throat, fever, bilateral posterior cervical lymphadenopathy; spontaneous rupture occurs often 10-21 days after disease onset	Splenomegaly with hematoma	Atypical immunoblasts with red pulp expansion
Hemangiomas	The most common primary tumor of the spleen. Average age is the sixth decade. May have abdominal pain, can rarely present as rupture and hemoperitoneum	Well-defined lesion, usually solid Larger lesion may be partially cystic	Vascular channels with plump endothelial cell lining
Hairy Cell Leukemia	Older males with weakness, weight loss, pancytopenia, splenomegaly, and associated atypical mycobacterial infections	Diffuse marked enlargement of the spleen without nodules	Proliferation of pale small lymphocytes in the red pulp, tumor cells invading sub-endothelial lining, and “lakes” of blood lined by abnormal lymphocytes

ings are characteristic and nearly diagnostic, and led us to consider splenic peliosis as a cause of spontaneous splenic rupture.

CONCLUSION

The spleen is a highly vascular organ that can rupture under various circumstances, leading to death. In forensic and hospital autopsies, spontaneous splenic rupture due to isolated splenic peliosis should be in the differential diagnosis. Splenic peliosis should be considered by the pathologist in cases of splenic rupture with no history or signs of trauma, and with the characteristic cyst-like blood-filled spaces in the splenic parenchyma. It is important for pathologists to identify the gross characteristics of splenic peliosis and to differentiate this entity from traumatic rupture of the spleen. The correct determination of the cause of splenic rupture is key to identify the appropriate cause and manner of death.

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