

CLINICAL VIGNETTE

“Ouch, My Belly Hurts!” A Case of a Large Non-Infectious Splenic Cyst

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Case Presentation

A 67-year-old man with a type 2 diabetes mellitus and hepatitis C developed left upper quadrant abdominal and lower thoracic pain gradually over a few weeks. He presented to the emergency room and X-rays revealed old rib fractures and he was discharged with a presumed musculoskeletal etiology for his pain. He came to the outpatient clinic for follow up, at which time his exam revealed normal vital signs, a protuberant abdomen, normoactive bowel sounds, tenderness to palpation of the left upper quadrant with deep palpation, no overt splenomegaly, and no crepitus over the ribs or tenderness on the ribs.

Prior to his current presentation, testing for hepatitis C was already underway, which included an abdominal ultrasound to evaluate for cirrhosis. The imaging study was scheduled after the onset of his abdominal discomfort and the ultrasound incidentally noted a 7cm splenic cyst. He later underwent a CT scan that showed a mildly enlarged spleen with an approximately 7.5 x 6.6 cm cystic lesion with mildly thickened septations noted, not present on prior CT performed 10 years earlier. He denied a history of livestock or sheep exposure, although he did report pulling worms out of the flesh of a local inhabitant when he was stationed overseas during the Vietnam War. Serologies for Echinococcus, coccidiomycosis, HIV and TB were performed and were all negative. The remainder of his routine labs were within normal limits.

Discussion

Worldwide, the most common etiology for large splenic cysts is hydatid disease. However, the differential diagnosis for splenic cysts varies depending on the severity of the clinical presentation whether the patient appears ill or not and geographic location of the patient particularly if a history of significant exposure to sheep. Splenic cysts are divided into primary cysts, which by definition contain an epithelial lining, and secondary cysts, which have no epithelial lining, which tend to be posttraumatic.¹

Of the primary cysts, serologic testing and imaging studies are generally sufficient for diagnosis. *Echinococcus multilocularis* and *Echinococcus granulosus* are the two most common infective species to cause primary splenic cysts. *Echinococcus* preferentially involves the liver and lungs, but the spleen is the

3rd most common site.² On CT or MRI imaging of the spleen, hydatid cysts tend to have an irregular contour, no well-defined walls, central necrosis, and irregular calcification along the walls or within the cyst. Antibody testing has generally been superior to antigen testing and has a high sensitivity. Depending on the size of the cyst, treatment can range from solely medical therapy with albendazole to total splenectomy. Direct aspiration or biopsy can be attempted for definitive diagnosis. However, there remains a concern for potential secondary seeding of the peritoneal space and there are case reports of anaphylaxis as an acute hypersensitivity reaction from cyst rupture.

Other common causes of primary splenic cysts included lymphangioma, a rare malformation of splenic lymphatic channels mainly seen in children and primary non-infectious epithelial splenic cysts.³ The latter accounts for 10-20% of all benign cysts – most of which are diagnosed in childhood and rarely occur in later life.⁴ On imaging, primary epithelial cysts tend to be unilocular with thin, smooth walls. There is no clear evidence-based standard of care for managing primary non-infectious epithelial cysts. One option is serial monitoring, although complications can include massive enlargement of the cyst with splenomegaly, rupture, bleeding, or secondary infection. However, surgical options such as open partial splenectomy and laparoscopy cystic wall unroofing can be used for large and symptomatic lesions.⁵

In the United States, the vast majority of splenic cysts are secondary cysts – also called pseudocysts since they do not have true walls – and are usually the after-effect of infection, infarction, or trauma. Prior infection is usually obvious since bacterial infection with *Staph*, *Strep*, or *Enterococcus* species will have been obvious. Prior fungal infection usually causes multiple micro-abscesses, so it will have a different radiographic appearance than an isolated splenic cyst. Acute bacterial abscesses can manifest with fever, left upper quadrant pain, referred pain to the left shoulder or pleurisy, abdominal distension or splenomegaly. The patient will have had an antecedent history of illness – usually as a secondary complication of endocarditis, sepsis from pneumonia, or GI perforation since primary infection is unusual.⁶ They may have had prior abscess drainage with the splenic pseudocyst a residual finding. If the primary infection has already been treated and the

secondary cyst is an asymptomatic incidental finding, no further treatment is required.

Other causes of secondary splenic cysts, include splenic infarction with multiple different etiologies. The most common underlying cause is cardioembolic occlusion (from atrial fibrillation or atrial septal defect). Other causes include underlying malignancy, celiac or splenic artery atherosclerosis, hypercoagulable states such as heparin-induced thrombocytopenia, antiphospholipid syndrome or APS, sickle cell anemia, postoperative infarction, and secondary inflammation and subsequent infarction from acute pancreatitis.⁷ The underlying treatment for cysts resulting from splenic infarction depends on the underlying cause. Usually there is no specific intervention for small, asymptomatic cysts, but the underlying cause may require anticoagulation as in APS.

Other mimics of splenic cysts on ultrasound that are usually quickly ruled out with advanced imaging are liver hemangiomas (vascular, not truly cystic lesions) and splenic peliosis (irregular, ill-defined cystic blood-filled cavities; generally not a solitary lesion). There are rare cases of spontaneous involution of liver cysts, but most symptomatic cases of a benign etiology are treated with partial or total splenectomy.⁸

Clinical Course

Our patient was unusual as his cyst developed in later life with CT abdomen/pelvis ten years prior revealing no splenic abnormalities. It was assumed that his splenic cyst was likely post-traumatic given his history of being involved in multiple motor vehicle accidents with numerous broken bones including old bilateral rib fractures. He went through a trial with serial monitoring of the splenic lesion, but due to ongoing pain over the next 6 months, he elected for a laparoscopic total splenectomy. Pathology revealed no malignancy and he reported resolution of his left sided upper abdominal pain after surgery.

Figures

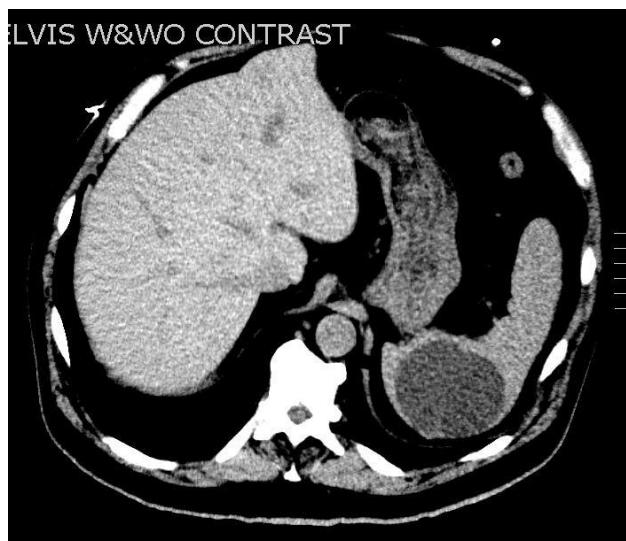


Figure A. Axial view of splenic cyst on CT imaging



Figure B. Sagittal view of splenic cyst



Figure C. Coronal view of splenic cyst

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