A Pediatric Patient with Pylephlebitis in the “Crazy” World of Antibiotics

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Abstract

Pylephlebitis is septic thrombophlebitis of the portal vein. It is an uncommon disease with high morbidity, mortality due to delayed diagnosis. It is associated with intra-abdominal infections, mostly acute appendicitis in childhood. We report a 16-year-old girl with a diagnosis of pylephlebitis and liver abscess secondary to perforated acute appendicitis.

Keywords: Portal vein thrombophlebitis; US; CT; Children

Introduction

Septic thrombophlebitis of the portal venous system is a rare ascending infection that arises from gastrointestinal origins such as diverticulitis, appendicitis, pancreatitis, cholangitis, inflammatory bowel disease, bowel perforation and pelvic infections [1]. In the pertinent literature, other terms have been variably used for the same scenario e.g. pylephlebitis, ascending septic thrombophlebitis, or suppurative pylephlebitis.

Patients with septic portal venous thrombophlebitis may present with nonspecific abdominal pain. Hence, the diagnosis may be delayed in pediatric patients. Mortality rates of 11% to 32% have been reported for untreated septic thrombophlebitis of the portal vein due to bowel ischemia, hepatic abscesses, and sepsis [2]. Contrast-enhanced Computed Tomography (CT) is an important radiological modality in the diagnosis of septic thrombophlebitis to identify portal venous thrombosis, portal venous gas, intrahepatic abscesses, moreover the gastrointestinal source of infection. In this report, we present a girl with septic thrombophlebitis of the portal vein due to perforated acute appendicitis.

Case Presentation

A 16-year-old girl presented to a local hospital with abdominal pain and vomiting one month ago. She was given oral antibiotics and rehydration therapy with the suspicion of urinary system infection. Her clinical status deteriorated despite treatment and ten days later she was admitted to another hospital due to persistent fever (38.9°C) and abdominal discomfort. On physical examination, jaundice and hepatosplenomegaly were detected. She was given intravenous immunoglobulin besides antibiotics with a probable diagnosis of Hemophagocytic Syndrome due to resistant fever and elevated levels of aminotransferases and ferritin. Heparin infusion was started due to portal vein and super mesenteric vein thrombosis. Pleural effusions were determined after examination, jaundice and hepatosplenomegaly were detected. She was given intravenous immunoglobulin besides antibiotics with a probable diagnosis of Hemophagocytic Syndrome due to resistant fever and elevated levels of aminotransferases and ferritin. Heparin infusion was started.

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tests for metabolic diseases and chronic hepatitis were in normal limits. Contrast-enhanced abdominal CT revealed thrombus in the main, right and left branches of the portal vein; causing non-homogeneous enhancement of the hepatic parenchyma. There was also a thrombus within the lumen of the superior mesenteric vein and multiple hypodense liver abscesses. Finally, CT images acquired at the level of the cecum showed evidence of fat stranding in the retrocecal area, wall-thickening in the cecum and ascending colon. The diagnosis of septic thrombophlebitis of the portal vein was considered after evaluation of the clinical, laboratory and radiological examinations of the patient. The liver abscess was drained in our interventional radiology unit. Culture analysis of the abscess yielded Streptococcus anginosus. No hereditary thrombophilic factors were determined and primary immunologic evaluation was normal. Her clinical status and laboratory tests improved under concomitant anticoagulation and antibiotic therapy. Three months later, perforated appendicitis was confirmed during a diagnostic laparoscopy and appendectomy was performed. Follow-up ultrasonography demonstrated cavernous transformation without any sign of portal hypertension. Written informed consent was received from the family.

**Discussion**

Pylephlebitis is septic thrombophlebitis of the portal venous system. It usually originates from an infection in the area drained by the portal system or in the structures contiguous to the portal vein. Pylephlebitis begins with thrombophlebitis of the small veins that drain the infected region [3]. Extension into larger veins leads to septic thrombophlebitis of the mesenteric vein, which can further involve the portal vein. The clinical scenario is associated with multiple possible causes that can present in any age group. Diverticulitis is the most common cause in the adulthood, but there are many other reasons including appendicitis, urinary/pelvic infections, biliary disease, inflammatory bowel disease and necrotizing pancreatitis. In our case, the pylephlebitis and pyogenic liver abscess were secondary to perforated appendicitis.

Pylephlebitis commonly presents with nonspecific symptoms including fatigue, fever, abdominal pain, nausea and vomiting. Furthermore, hepatomegaly and jaundice can be seen. Therefore, radiological studies demonstrating a thrombosis in the portal system are paramount for prompt diagnosis. Liver imaging findings include unopacified branches of the portal vein, transient parenchymal attenuation differences and intrahepatic abscesses. Up to 37% of cases of pylephlebitis are complicated by a pyogenic liver abscess [4]. A Gram-negative bacterium is most often the underlying cause and is often related to an inflammatory condition of the bowel [4]. Patients who have portal thrombosis secondary to abdominal sepsis may completely recover, with the recanalization of the vessel after successful treatment of the underlying sepsis. They may develop cavernous transformation or portal hypertension as well.

The morbidity and mortality of pylephlebitis have dramatically decreased with advanced diagnosis by the utility of multimodality imaging and treatment with broad-spectrum antibiotics. Nevertheless, the mortality rate still remains high due to delayed diagnosis and ineffective treatment. The worst scenario occurs due to sepsis or peritonitis while thrombus spread into the superior and inferior mesenteric veins may result in bowel ischemia and infarction [5].

Pylephlebitis with a pyogenic liver abscess due to intra-abdominal infection resulted from perforated appendicitis is a rare condition associated with a high mortality rate unless treated promptly. It should be kept in mind that the clinical presentation can be nonspecific especially in pediatric patients, and the imaging findings are often complex - resulting in delayed diagnosis and treatment. To this end, awareness of this disease and being familiar with its imaging
findings would indisputably be life-saving from the radiologist side of the story.

References


