CASE REPORT

An Unusual Presentation of Acute Appendicitis Complicated by Portal and Mesenteric Thrombophlebitis

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ABSTRACT
Portal and mesenteric thrombophlebitis is a well-documented but rare complication of acute appendicitis. The condition is caused by ascending infection and is potentially life threatening. Prompt diagnosis and treatment with surgery, antibiotics, and anticoagulants are mandatory. This report is of a patient with acute appendicitis complicated by portal and mesenteric thrombophlebitis. The unusual presentation caused a diagnostic challenge, which delayed treatment.

Key Words: Appendicitis; Diagnosis; Mesenteric veins; Portal vein; Thrombophlebitis

INTRODUCTION
Pylephlebitis (suppurative thrombophlebitis of the portal vein) is a well-known but unusual complication of intra-abdominal and pelvic infections, such as acute appendicitis, diverticulitis, and inflammatory bowel disease. The exact incidence of this condition is unknown but it is considered rare due to the widespread use of antibiotics.1 Pylephlebitis can result in catastrophic complications of septic emboli or liver abscess. Prompt diagnosis and treatment are necessary to reduce the associated morbidity and mortality. However, atypical clinical manifestations can occur and result in diagnostic uncertainty.

This report is of a patient with acute appendicitis complicated by portal and superior mesenteric venous thrombophlebitis with an unusual presentation, causing a diagnostic challenge and delayed treatment.

CASE REPORT
An otherwise healthy 54-year-old man presented in 2004 with right upper quadrant abdominal discomfort and abdominal distension for 2 weeks, preceded by diarrhoea, decreased appetite, and passing of tea-coloured urine. At physical examination, he was febrile (38.5°C) and jaundiced. The abdomen was soft and non-tender on palpation. No other significant findings were detected.

Blood tests showed an elevated white blood cell count of 17.2 x 10^9/L (normal range, 4.5-11.0 x 10^9/L) and abnormal liver function tests, with isolated elevation of total bilirubin of 156 µmol/L (normal range, 5-21 µmol/L) and alkaline phosphatase of 216 U/L (normal range, 50-120 U/L). The clotting profile was also impaired with an international normalized ratio of 1.5, indicating liver dysfunction. At this point, the clinical picture was suggestive of obstructive jaundice or cholangitis.

Figure 1. Doppler ultrasound scan of the portal vein showing a distended portal vein with internal thrombus and absent Doppler colour signal (arrow).
Endoscopic retrograde cholangiopancreatography (ERCP) was performed the next day, but did not reveal any abnormality. Ultrasound of the liver performed subsequently showed evidence of portal vein thrombosis (Figure 1), but no liver mass or biliary tract abnormality was seen. The rest of the abdomen was not examined due to the clinical focus on the liver. Contrast computed tomography (CT) scan of the abdomen and pelvis was performed on the same day, confirming the diagnosis of portal vein thrombosis (Figure 2), which was noted to involve the superior mesenteric vein (Figure 3). The appendix was swollen (Figure 4), and a periappendiceal inflammatory mass (Figure 5) and fluid was noted. The findings were compatible with acute appendicitis complicated by a periappendiceal abscess and ascending phlebitis causing superior mesenteric and portal vein thrombosis.

Emergency laparotomy for appendicectomy and drainage of the periappendiceal abscess was performed 1 day later. The patient was given antibiotics (intravenous cefoparzone sodium and metronidazole), and anticoagulation (intramuscular nadroparin sodium for 4 days followed by warfarin) was also commenced for the venous thrombosis. He had an uneventful recovery and continued to improve clinically. Liver function also improved.
Follow-up CT scan done 5 months later showed recanalisation of the portal vein (Figure 6). Anticoagulation was subsequently stopped and the patient remained well.

DISCUSSION

Acute appendicitis is one of the most common causes of acute abdomen. Acute appendicitis occurs when the appendiceal lumen is obstructed, leading to inflammation and subsequent perforation. Accurate and prompt diagnosis is essential to minimise morbidity as major complications can occur when treatment is delayed. Septic seeding of the mesenteric vessels is a serious and potentially life-threatening complication. Ascending infection spreading along the draining mesenteric-portal venous system may result in pylephlebitis, pylethrombosis, and hepatic abscess formation. Pylephlebitis is a well-documented but rare complication of unknown incidence. The mortality rate has been reported to be as high as 32%. Therefore, it is important to recognise such potentially life-threatening complications, especially for patients with symptoms and signs of high-grade bacteremia. Imaging plays a significant role in the diagnosis of pylephlebitis, as the condition is often not detected at surgery.

Although the clinical diagnosis of acute appendicitis may be straightforward for patients who present with classic signs and symptoms, a wide variety of atypical presentations exist that may elude even the most experienced clinicians. Atypical presentation may lead to diagnostic confusion and treatment delay. This patient presented with classic symptoms and signs of obstructive jaundice (right upper quadrant discomfort, jaundice, fever, and abnormal liver function tests). Based on these clinical signs, the clinician decided to proceed to ERCP immediately. This highlights the difficulty of identifying the primary source of infection on the basis of clinical signs alone. In retrospect, ERCP was non-contributory and caused an unnecessary delay to making the correct diagnosis.

Cholestatic jaundice resulting from sepsis is not uncommon. The suggested pathogenesis includes direct bacterial action, circulating endotoxins, and inspissated bile syndrome secondary to changes in the water and electrolyte composition of bile.

More importantly, the delay in making the definitive diagnosis could have endangered the patient’s life. Although ultrasound examination showed portal vein thrombosis, the cause remained unknown. The CT scan confirmed the diagnosis of appendicitis and complication of venous thrombosis. This led to treatment by immediate laparotomy to remove the suppurative focus and subsequent antibiotic and anticoagulant therapies to control the sepsis and thrombosis. This patient underscores the importance of looking for causes of portal vein thrombosis apart from hepatocellular carcinoma invasion or portal hypertension, such as infective, inflammatory, or neoplastic conditions of the gastrointestinal tract, pancreas, or biliary tract.

Despite some controversy, anticoagulation has been recommended, not only to recanalise the thrombotic vein but also to reduce septic embolisation to the liver and subsequent liver abscess formation. A decreased recurrence rate of portal and mesenteric thrombophlebitis has also been reported. There is no consensus in the literature on the optimum duration for anticoagulation. Generally, a short duration (<3 months) is reasonable if there is no concurrent complication of bowel infarction or septic embolisation. Thrombectomy will only be required if the bowel is compromised.

In summary, this patient with acute appendicitis and pylephlebitis presented with symptoms and signs of obstructive jaundice. This led to unnecessary investigations and delayed treatment of a rare but potentially life-threatening complication of portomesenteric thrombophlebitis.

REFERENCES