



## Case report

## A case of pylephlebitis complicating an acute appendicitis: Uncommon cholangitis-like situation

Mar Dalmau<sup>a,\*</sup>, Carlos Petrola<sup>a</sup>, Pablo Lopez<sup>a</sup>, Ramon Vilallonga<sup>a,b</sup>, Amador Garcia Ruiz de Gordejuela<sup>a,b</sup>, Manel Armengol<sup>a,b</sup>

<sup>a</sup> Department of General and Digestive Surgery, Hospital Universitari Vall d'Hebron, Barcelona, Spain

<sup>b</sup> Universidad Autónoma de Barcelona, Spain



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## ABSTRACT

**Introduction:** Pylephlebitis represents an uncommon but serious condition with significant mortality which can complicate intrabdominal sepsis of any etiology. One of the most common predisposing infections is appendicitis. **Presentation of case:** A 21-year-old male with 4 days of epigastric and right upper quadrant pain with associated fever and chills with hyperbilirubinemia and leukocytosis in blood test was orientated as cholangitis at first diagnostic. Poor response to antibiotic treatment with persistent fever and bacteriemia with *E. coli* and *S. constellatus* isolated in blood cultures led to complete the study with a CT scan which revealed an acute appendicitis complicated with thrombosis of the superior mesenteric vein (SMV) up to the splenoportal confluence. Appendectomy, treatment with broad-spectrum antibiotic and anticoagulation treatment led to full recovery. Follow-up after 6 months showed almost complete SMV patency.

**Discussion:** Pylephlebitis can present as a clinical cholangitis-like picture with hyperbilirubinemia with or without liver abscess formation. CT scan seems to be the most sensitive diagnostic test as it identifies the underlying focus of infection, the extension of the thrombosis and detects liver abscesses. Surgical removal of the source of infection as appendectomy and adequate antibiotic treatment adjusted by culture should be initiated promptly. Anticoagulant treatment should be considered in the case of poor clinical outcome or thrombosis progression.

**Conclusion:** Pylephlebitis should be suspected mainly in patients with appendicitis and diverticulitis with erratic behavior despite surgical removal and/or antibiotic treatment with abnormal liver tests and persistent bacteriemia. CT scan is the preferred image study.

## 1. Introduction

Pylephlebitis, or infective thrombosis of the portal vein system, results from intrabdominal sepsis of any etiology and represents a serious condition due to subacute presentation and delayed diagnosis with significant morbidity and mortality from 11 to 32 % [1–3]. Outcome in reports over the last 20 years have improved because of earlier diagnosis with sensitive image technics and prompt treatment [1].

A case of thrombophlebitis of the superior mesenteric vein as a complication of acute appendicitis with a clinical cholangitis-like debut is described. It was successfully treated with surgical removal of the first infection site, antibiotics, and anticoagulation therapy. The work has been reported in line with the SCARE criteria [2].

## 2. Case presentation

A 21-year-old male with no history of interest presented with a 4-day history of epigastric and upper quadrant abdominal pain, vomiting and fever with chills accompanied with dark tone urine. On admission, he had a high temperature (38.5 °C), his blood pressure was 117/80 mmHg, his pulse rate 110/min and his respiratory rate was 20/min. Physical examination noted right upper quadrant pain and tenderness without muscle guarding. Blood test results showed normal leukocytes (9.250/mm<sup>3</sup>; NV 4.000–11.000/mm<sup>3</sup>), elevate bilirubin (2.5 times normal value), increased cholestasis enzymes (alkaline phosphatase slightly elevated, and  $\gamma$ -GT 3 times elevated) and an elevated C-reactive protein (21.65 mg/dL; NV 0.03–0.5 mg/dL). Abdominal ultrasound described a minimal ectasia of the intrahepatic bile duct with no apparent obstructive cause and cholelithiasis. Under diagnostic

\* Corresponding author at: Hospital Universitari Vall Hebron, Universidad Autónoma de Barcelona, Paseo de la Vall d'Hebron, 119-129, 08035 Barcelona, Spain. E-mail addresses: [mardalmau6@gmail.com](mailto:mardalmau6@gmail.com) (M. Dalmau), [manel.armengol@vallhebron.cat](mailto:manel.armengol@vallhebron.cat) (M. Armengol).

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orientation of acute cholangitis in a patient with cholelithiasis, he was treated with amoxicillin clavulanic acid after extracting blood cultures and admitted to the Hospital. Blood cultures were positive for *Escherichia coli* and *Streptococcus constellatus*. During his stay, the patient remained feverish despite antibiotic therapy for 48 h. The treatment was switched to piperacillin-tazobactam, and new blood cultures were obtained. An extended study with magnetic resonance cholangiopancreatography (MRCP) showed hepatosplenomegaly, bile duct within the normal limits and hepatic periportal and subcapsular edema suggesting hepatitis. The microbiological study was extended with serology of hepatotropic viruses, which resulted negative. In the context of acute infection with persistent fever and bicytopenia with lymphopenia and thrombocytopenia, a CT abdominal scan was performed, with findings of acute appendicitis complicated with an abscess at the root of the mesentery ( $37 \times 26$  mm) and superior mesenteric vein (SMV) thrombosis (Fig. 1). A gangrenous appendix was removed, and the abscess was drained. Anticoagulation treatment with low molecular weight heparin (LMWH) was started, with prophylactic doses reaching complete treatment when platelet recount was normalized. He had erratic progress with fever on the 7th postoperative day with a CT scan that showed a surgical site fluid collection of  $87 \times 32$  mm with almost complete thrombus of the SMV. Initially, a percutaneous drainage was placed, resulting insufficient and requiring subsequent laparoscopic surgical drainage. Later surgical cultures resulted positive for *E. coli BLEE*, so the treatment spectrum was extended with ertapenem. Eventually, his postoperative progress was adequate, and he was discharged on the 24th postoperative day. He

received oral anticoagulation with acenocoumarin and completed 6 weeks of intravenous antibiotic therapy. The follow-up 6th month CT scan demonstrated patency of the SMV, finishing anticoagulant treatment by then.

### 3. Discussion

Pylephlebitis begins with the thrombophlebitis of small veins, draining an area of infection and leading to septic thrombophlebitis of the portal and mesenteric vein by extension. Diverticulitis and appendicitis are the most common predisposing infections [1,3] although it has also been associated with pancreatitis [3], cholangitis and infected choledocholithiasis [4,5], inflammatory bowel disease [6] and pelvic sepsis [7,8].

Bacteremia associated with pylephlebitis occurs in 44–88 % of cases and is usually polymicrobial, *E. coli* and *B. fragilis* being the most common isolations [1,4]. In the case presented, blood cultures in admission resulted positive for *E. coli* and *Streptococcus constellatus*. However, two days after initial antibiotic treatment, in the surgical liquid only *E. coli BLEE* was present.

The most common presenting symptoms are abdominal pain and fever. Clinical signs may include right upper quadrant or generalized abdominal tenderness and hepatomegaly. Although pylephlebitis usually presents as an acute illness, the course of the primary focus of infection is subacute and silent [9,10].

Leukocytosis and abnormal liver function tests, especially an



**Fig. 1.** Coronal section of abdominal CT-scan with thrombosis of the superior mesenteric vein (arrow) secondary to an appendicitis complicated with an abscess (asterix).

increase of cholestasis values, are common findings in blood tests. Clinical jaundice appears to be an unusual condition in pylephlebitis unless it is complicated with hepatic abscesses [1,11,12]. In the above case, an increase of bilirubin and mild abnormalities in liver function suggested an incipient cholangitis, which was later seen to have overlapped and masked the clinical picture of an acute appendicitis with thrombophlebitis, in the absence of liver abscess formation. Moreover, the inciting intrabdominal sepsis may also contribute to hepatic dysfunction.

In terms of complications, liver abscesses are reported in a third of cases, resulting from septic emboli release within the liver [3,11]. Mesenteric vein involvement, present in 42 % of cases could lead to bowel ischemia and infarction. However, bowel ischemia requiring resection is a rare condition described in just one case [13]. In the case presented, the thrombus involved the SMV up to the splenoportal confluence without intrahepatic implication or signs of bowel ischemia in surgical revision. Portal hypertension is a rare and long-term complication [14].

The diagnosis is reached by demonstration of portal vein thrombosis in a patient with bacteremia. As it is an uncommon condition, the diagnosis is usually delayed because symptoms are unspecific and clinical signs from the first infection site are typically subacute. In the past, the diagnosis was often done at laparotomy or in the autopsy in patients with severe sepsis.

Demonstration of thrombosis in the portal and mesenteric vein can be done by CT and ultrasonography. The advantage of the CT study is identification of the underlying focus of infection, extent of pylephlebitis, and intrahepatic abnormalities, such as liver abscesses [15].

It is also remarkable that in this case both abdominal US and MRCP were not able to find the real cause of the infection. It's likely that the biliary signs and symptoms led the radiologist to focus primarily on the biliary tract. It's possible that in such a severe case of sepsis an abdominal CT-scan should have been done earlier.

The principal treatment for the infection is by using empiric broad gram-negative and anaerobic spectrum antibiotics until culture results are available [1]. The treatment, which takes at least 4 to 6 weeks, can be completed by oral agents after a significant clinical response and availability, depending on the microorganisms isolated.

Surgery is needed when the main infective cause is an appendicitis, although Stizenberg et al. [16], reported that interval laparoscopic appendectomy could be delayed 3 months after treatment. Surgical or radiological percutaneous drainage of intrabdominal abscess associated may be needed.

The role of anticoagulant treatment is controversial, as there are no randomized controlled studies and no consensus on its use in thrombophlebitis. Extensive thrombosis to the mesenteric vein, progression of thrombosis in CT control, persistent bacteremia despite antibiotic treatment or presence of hypercoagulable state, are all reasonable indications for anticoagulation [14,17–19].

The most common regimen is initially anticoagulation with LMWH [14]. On discharge, it can be switched to an oral anticoagulant. Treatment duration is uncertain. It is reasonable to stop when image CT control suggests that the thrombus disappears or remains stable.

#### 4. Conclusion

Pylephlebitis is a serious condition with significant morbidity and mortality which should be suspected mainly in patients with appendicitis and diverticulitis with erratic progression, despite surgical removal and/or antibiotic treatment with abnormal liver tests and persistent bacteremia. The CT scan is the preferred image study because of the additional ability to identify the underlying focus of infection, thrombus extension and liver complication. Treatment is based on surgical removal if needed and antibiotic therapy which takes from 4 to 6 weeks. Anticoagulant treatment is not routinely recommended unless there is a poor clinical response or extended thrombosis progression.

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This study was exempt from ethical approval.

#### Consent

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#### Author contribution

Mar Dalmau: Managed the case, Wrote the original manuscript, Got informed consent.

Carlos Petrola: Managed the case, Reviewed and edited the manuscript, Got informed consent.

Pablo López: Managed the case, Reviewed and edited the manuscript, Got informed consent.

Ramón Vilallonga: Managed the case, Reviewed and edited the manuscript.

Amador Garcia Ruiz de Gordejuela: Managed the case, Reviewed and edited the manuscript.

Manel Armengol Carrasco: Reviewed and edited the manuscript, Supervised the process.

#### Guarantor

Amador Garcia Ruiz de Gordejuela.

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#### Declaration of competing interest

None of the authors have conflicts of interest.

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