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**Case Report**

# Pylephlebitis secondary to a gangrenous appendicitis a forgotten complication: a case report and review of the literature

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## ABSTRACT

Pylephlebitis is an uncommon condition which can complicate intrabdominal sepsis of any etiology. A 22-year-old male with 2 weeks of epigastric and right upper quadrant pain with associated fever, chills, nausea, vomiting, hyperbilirubinemia and leukocytosis in blood test documented. An acute cholangitis was the first diagnosis. Abdominal ultrasound described multiple abscesses and gallbladder wall thickening. CT scan revealed a case of acute appendicitis complicated with a thrombosis of the portal vein. Diagnostic laparoscopy was performed, observing an inflammatory process in the right lower quadrant, cholecystectomy with cholangiography was done with no complications, and then appendicectomy was performed. Broad-spectrum antibiotic and anticoagulation treatment led to full recovery. Follow-up after 12 months showed a full recovery and no signs of thrombosis in the serial CT scans for follow-up. Pylephlebitis can present as a clinical cholangitis-like scenario with hyperbilirubinemia with liver abscess formation. CT scan seems to be the most sensitive diagnostic test because it can identify the underlying infection foci, it also can show the extension of the thrombosis and other complications like liver abscesses. Surgical removal of the source of infection 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. Pylephlebitis should suspected in the case of poor clinical outcome of intrabdominal infections, a properly diagnosis with imaging tests and multimodal management can reduce the morbidity and mortality, and also short- and long-term complications of this pathology.

**Keywords:** Pylephlebitis, Portal vein, Portal vein thrombosis, Appendicitis, Cholangitis presentation, Hepatic abscess, Case report

## INTRODUCTION

Pylephlebitis or infective suppurative thrombosis of the portal vein system is a rare and severe condition, which is characterized as a result of intrabdominal sepsis of any etiology and represents a significant critical condition of morbidity and mortality due to. Its sub-acute presentation and delayed diagnosis.[1-7]

Pylephlebitis was first described by William Osler in 1882 who reported the case of a 28-year-old patient who died of complications of septic pylethrombosis.[8]

In the pre-antibiotic era this condition was universally fatal, nowadays has become treatable with the improve of the earlier diagnosis with sensitive image technics and the use of new therapeutic strategies and also the increase of awareness of this condition.[9]

Pylephlebitis is most commonly caused by diverticulitis followed by appendicitis, and then less commonly by pancreatitis and other focal infections.[10]

Due to the low incidence that ranges between 0.3 and 3 cases per 100 000 patients per year, no current

randomized controlled trials or prospective studies describe the risk factors, diagnosis and most important therapeutic strategies.[11]

Here we describe a clinical case of 22-year-old male with Pylephlebitis as a complication of an acute appendicitis with clinical cholangitis like debut

## CASE REPORT

A 22-year-old male with no medical history of interest to his current condition, presents with a 2 weeks history of epigastric and upper quadrant abdominal pain, vomiting and fever with chills, patient referred change in bowel habits, like diarrhea and pale stool and also dark tone urine. He was initially treated in a local hospital near his hometown. As his pain worsened and his symptoms did not improve after the treatment, his relatives decided to visit our hospital for further medical management. On admission to the emergency department, he had fever (38.5ºC), he had low blood pressure and the patient was tachycardic. On the physical examination noted right upper quadrant pain also in the epigastrium, he had abdominal tenderness.

His laboratory values on admission showed leukocytosis, low platelet count, elevated bilirubin, increased cholestasis enzyme (Table 1). Serology was made and was negative. Blood cultures were obtained and were positive for Escherichia coli.

**Table 1: Laboratory values upon admission.**

| Laboratory test | Normal range | On admission | Post op |
|---|---|---|---|
| **WBC count ($10^9$cells/l)** | 3.5-9.5 | 39.6 | 14.1 |
| **Neutrophil (%)** | 50-70 | 85 | 81.4 |
| **Hemoglobin (g/dl)** | 11.4-16 | 12.1 | 11.8 |
| **Platelet count ($10^9$cells/l)** | 125-350 | 56 | 186 |
| **PT (Prothrombin time)** | 11-16 secs | 18 | 16 |
| **Total bilirubin (mg/dl)** | 0.4-1.2 | 15.4 | 4.17 |
| **Direct bilirubin (mg/dl)** | 0-0.8 | 13.4 | 3.5 |
| **AST (U/l)** | 5-45 | 78 | 70 |
| **ALT (U/l)** | 5-40 | 27 | 30 |
| **Albumin (g/l)** | 3.5-5-5 | 3.4 | 3.0 |
| **Amylase (U/l)** | 0-140 | 30 | 32 |
| **Lipase (U/l)** | 0-60 | 31 | 33 |
| **Blood urea nitrogen (mg/dl)** | 8-20 | 266 | 46.9 |
| **Creatinine (mg/dl)** | 0.5-1.2 | 2.5 | 1.3 |
| **C-reactive protein (mg/dl)** | 0-6.45 | 38 | 15.8 |

Abdominal ultrasound was performed and described a general increase in its size of the liver with multiple rounded images, irregular edges with a wall, heterogeneous echogenicity, suggesting a probable abscesses. The gallbladder measures 71×38×42 mm, with a volume of 61.7 cc, its wall thickening up to 5.9 mm, inside there is no evidence of stones or bile sludge. No perivesicular collections are observed Figure 1.

A CT scan was performed and showed multiple abscesses and thrombosis of the left branch of the portal vein, also it was observed findings of complicated appendicitis Figure 2.

## RESULTS

In the above case, an increase of bilirubin and mild abnormalities in liver function suggested an incipient cholangitis, which was later seen to have over- lapped and masked the clinical picture of an acute appendicitis with thrombophlebitis.

Despite antibiotic and anticoagulation therapy patient remained feverish and had erratic progress.

Intraoperatively showed a gall bladder enlargement so cholecystectomy with intraoperative cholangiography was made without evidence of choledocholithiasis, an appendiceal remanent in retrocecal position, gangrenous appendix was observed so it was removed Figure 3.

Postoperatively, the patient showed an adequate progress, his leukocyte counts significantly decreased and no abdominal symptoms was shown.

Histopathology confirmed a chronic cholecystitis process and gangrenous appendicitis.

During his stay in the hospital, he was prescribed with anticoagulant therapy for portal vein thrombosis and continue with oral anticoagulation for three months, and 6 weeks with antibiotic therapy.

Histopathology showed a chronic cholecystitis process and gangrenous appendicitis.

He was discharged on the 17th postoperative day. The follow-up 12-month CT scan showed patency of the

portal vein in the two branches, finishing anticoagulant treatment by then.

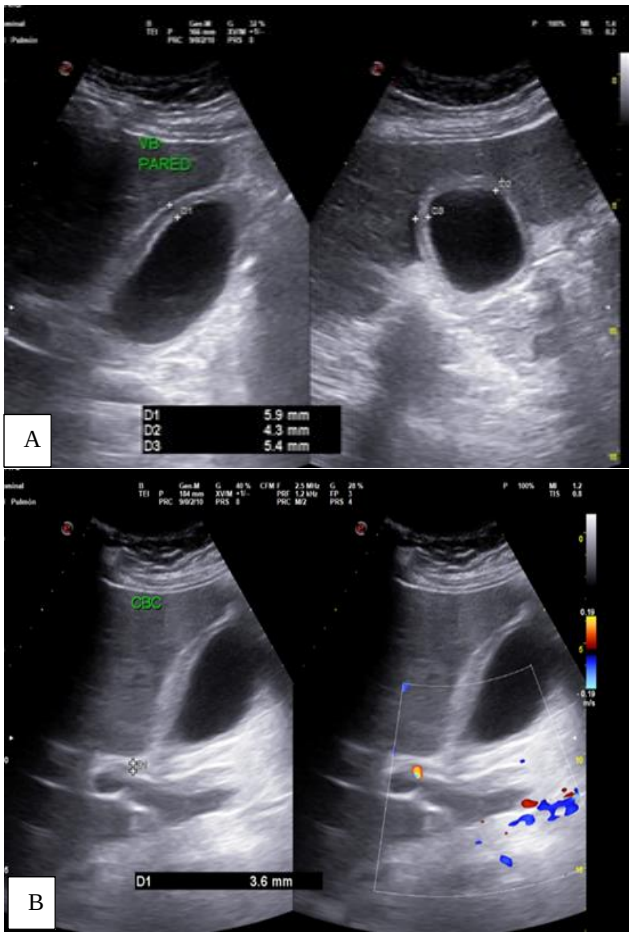


**Figure 1 (A and B): A great thickening of the wall. No stones inside the gallbladder.**

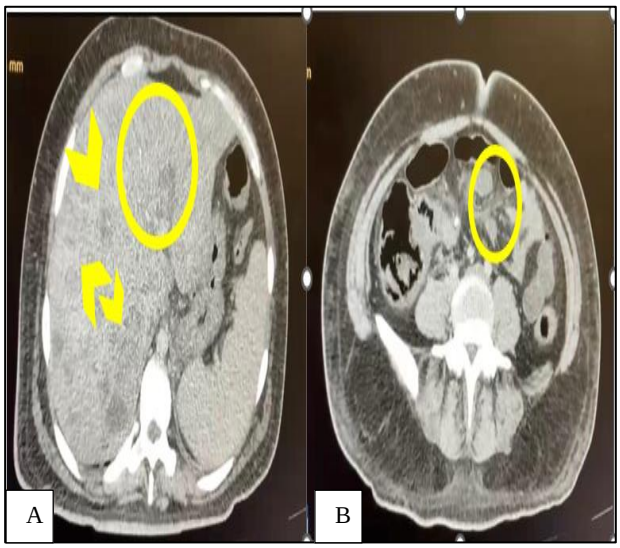


**Figure 2 (A and B): Multiple liver abscesses (arrows) and a thrombus in the left portal vein is shown in the circle. Fecalith and appendiceal process with local inflammation can be observed.**

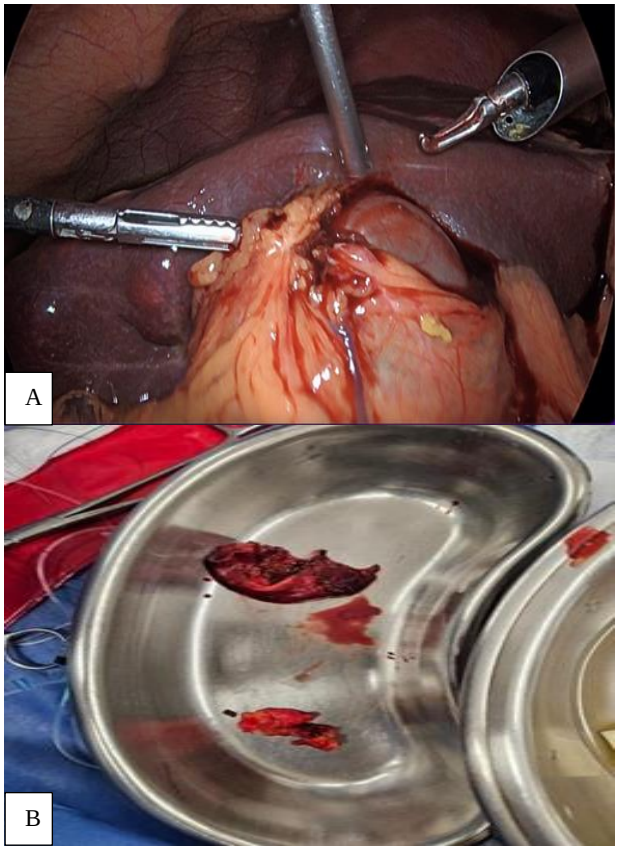


**Figure 3 (A and B): Intraoperative gallbladder and a gangrenous appendix.**

## DISCUSSION

Pylephlebitis can be a result of a complicated intra-abdominal or pelvic infection, that develop within areas drained by the portal venous circulation.[2,12] This process behind pylephlebitis starts with localized thrombophlebitis of small veins draining an area of infection and leading septic emboli migrate to the main branches of the portal vein.[13]

The anatomy of the portal system explains why diverticulitis (26.5%) and acute appendicitis (22%) are the two most common causes of pylephlebitis, it is worth to mention that contiguous infection can also lead to this entity such as cholangitis/cholecystitis (3.5%) or pancreatitis (5.5%), inflammatory bowel disease and pelvic sepsis.[14]

Pylephlebitis was caused by polymicrobial pathogens in most cases (44-88%), the most frequently isolated were *E. coli* (25%), *Bacteroides* spp. (17%) and *Streptococcus* spp (15%), it is important to mention that can be caused by fungal infections (3.5) all caused by *Candida* spp or parasitic infections (2%) due to intestinal worms and amoebas.[15]

The most frequently symptoms are fever, abdominal pain, other common symptoms. Include nausea or vomiting

and diarrhea.[16] Clinical signs may include right upper quadrant or generalized abdominal tenderness, hepatomegaly and splenomegaly.[17] Jaundice has been reported but is considered unusual, unless there is associated cholangitis or liver abscesses.[18]

All these symptoms are common in intra-abdominal infections, that is why the diagnosis makes it challenging to differentiate between symptoms caused by pylephlebitis or by the original infective process that caused pylephlebitis in the first place.[19]

Although pylephlebitis usually presents as an acute illness, the course of the primary focus of infection is subacute and silent.

Leukocytosis, C-reactive protein and abnormal liver function test, especially alkaline phosphatase with three-to-four-fold increase, gamma-glutamyl transferase with five- to tenfold increases, hyperbilirubinemia, with values of total bilirubin that could reach a six-fold increase, other common laboratory findings include anemia, hypoalbuminemia, and thrombocytopenia.[20]

In the above case, an increase of bilirubin and abnormalities in liver function suggested an incipient cholangitis, which was later seen to have over- lapped and masked the clinical picture of an acute appendicitis with thrombophlebitis.

The demonstration of thrombus in the portal vein is the principal finding leading to the diagnosis. Computed tomography (CT) scanning and abdominal ultrasonography can detect the presence of the thrombi in the portal vein.[21]

CT scan is preferred because of its higher definition and ability to identify possible abdominal or pelvic foci, also because it is not operator dependent.[22]

Magnetic resonance imaging (MRI), angiography, endoscopic ultrasound, or positron emission tomography (PET) can also be used to demonstrate portal vein thrombosis; however, their application remains limited to selected cases.[23]

The treatment of pylephlebitis consists of broad-spectrum antibiotics, with the choice of empiric antibiotics depending on the most probable source of infection and the most likely involved organisms, regardless of the presence of bacteriemia, this entity is mostly a polymicrobial infection, with gram-negative and gram-positive, aerobes and anaerobes bacteria, especially *E. coli*, *Streptococcus* spp., and *Bacteroides* spp. However, there is no randomized control studies that demonstrate a preferred empiric antibiotic regimen to this pathology.[24]

The suggested broad-spectrum antibiotic regimens are shown in Table 2.[25]

**Table 2: Broad-spectrum antibiotic regimen.**

| S. no. | Combination therapy |
|---|---|
| **1** | Metronidazole (500 mg every 6-8 h) plus one of the following |
| **2** | Ceftriaxone (2 g daily) |
| **3** | Cefotaxime (2 g every 6 h) |
| **4** | Ciprofloxacin (400 mg every 8-12 h) |
| **5** | Levofloxacin (750 mg daily) |
| **6** | Monotherapy |
| **7** | Based on beta-lactam/beta-lactamase inhibitor, with one of the following: |
| **8** | Piperacillin-tazobactam (4.5 g every 6-8 h or 13.5-18 g in continuous infusion) |
| **9** | Ampicillin-sulbactam (3 g every 6 h) |
| **10** | Or based on carbapenem, with one of the following: |
| **11** | Imipenem (500 mg every 6 h) |
| **12** | Meropenem (1 g every 8 h) |
| **13** | Ertapenem (1 g daily) |

The antibiotic treatment takes at least 4 to 6 weeks, and can be completed by oral agents after a significant clinical response and availability, depending on the microorganisms isolated.[26]

It is important to mention that there is no prospective randomized control trial demonstrates the utility of anticoagulants in pylephlebitis, the main purpose is to prevent thrombus extension and favor thrombus resolution.[27]

Various reports in the literature have found that anticoagulant administration can increase the rates of portal vein recanalization (58% vs. 21%) and reduce complications associated with chronic portal hypertension, in addition the anticoagulant therapy had lower mortality rates (6%) if compared who did not (22%). Nevertheless, there is no recommendation for prescribing anticoagulant therapy in all patients, they should be administered in patients with progression of the thrombosis on the serial imaging studies, or clinical worsening like persistent fever despite proper antibiotic therapy.[27]

The most common regimen is initially anticoagulation with LMWH that can be switched to oral anticoagulant, on discharge of the patient.[27]

Surgery is indicated for the treatment of the generalized peritonitis that originated the thrombophlebitis or for the drainage of suppurative complications, such as abdominal collections, liver abscesses, among others.[28,29]

The principal complications are caused by hematogenous dissemination of the pyogenic portal infection causing metastatic abscess, this thrombosis can persist and evolve in to a cavernous portal vein, characterized by veins forming within around the thrombotic segment.[30]

Pyogenic liver abscesses can complicate up to 37% of cases of pylephlebitis Other rarer sites include metastatic abscesses in the lung and the brain.it is important to note that Intestinal ischemia has also been described, in the literature.[31]

Long term complications include portal hypertension with dilated splenic veins and development of venous collaterals in the hepatoduodenal ligament.[32]

Mortality rates could range from 8.7 to 19% with overall mortality <10% for patients diagnosed after 2010. Sepsis has been found as a cause of death in 88.9% of the patients increasing the risk of death 17-fold. Positive blood cultures were also found to be an independent risk factor for death (approximately 2.2-fold).[33]

## CONCLUSION

Pylephlebitis is a rare condition which can virtually complicate any intraabdominal or pelvic infections that develop within areas drained by the portal venous circulation.

We would like to stress the importance of early diagnosis and treatment in cases of pylephlebitis. In the case of our patient, the diagnosis was elusive and delayed until we ruled out all diagnostic possibilities that met our patient's clinical criteria.

A high suspicion for pylephlebitis should be maintained in the context of intra-abdominal infections.

Likewise, early diagnosis of this disease must be made using tomography with intravenous contrast and timely treatment established by controlling the infectious focus, appropriate antibiotics and anticoagulants when it is indicated.

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