

Case Report

Ischemic colitis due to antiphospholipid antibody syndrome

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ABSTRACT

Introduction: Portal system ischemia may present insidiously which may aggravates the prognosis.

Case presentation: A 26-year old man presented with watery diarrhea and generalized abdominal pain for 3 months. On physical examination, moderate splenomegaly was noticeable. Stool exam and culture was negative except for blood in stool. Colonoscopy was in favor of inflammatory bowel disease although the patient symptoms have worsened despite treatment. Abdominopelvic computed tomography (CT) showed thromboses in portal and superior mesenteric veins and as the ill patient evolved signs of peritonitis, he underwent laparotomy during which, total colectomy was performed due to significant bowel necrosis. The cause of venous thrombosis of the portal system revealed to be Factor V Leiden and the presence of antiphospholipid syndrome.

Conclusion: High mortality rates of portal and mesenteric thromboses despite therapy urge the need for early clinical suspicion, careful assessment of the differential diagnoses and timely treatment for fewer adverse events. Although the therapeutic plan is challenging, anticoagulation, angiography and surgical resection increase survival.

1. Introduction

Thrombophilia is not an uncommon process in accordance with rheumatologic and hematologic diseases. It results from hereditary or acquired disorder of blood clotting or in the presence of hypercoagulable states. Inflammatory bowel disease, the Arg506Gln mutation in Factor V coagulation named as factor V Leiden and antiphospholipid syndrome are all recognized as predisposing factors for hypercoagulability.¹ Portal vein thrombosis can be caused by various thrombophilic conditions; however, the acute or chronic presentations vary widely in symptoms and signs and the prognosis may be poor despite treatment. The ethics of this report have been approved by the University of Medical Sciences Institutional Review Board.

2. Case presentation

A 26-year old man complained of generalized abdominal pain and infrequent watery diarrhea for 3 months that occur shortly after oral intake. He had been treated for acute hepatitis A due to increased liver enzymes and high HAV IgG Ab but abdominal pain was not completely resolved, radiating to flanks and aggravating after food ingestion.

Besides, he had one episode of bloody diarrhea, night sweats and significant weight loss in a month. He exhibited epigastric abdominal pain with infrequent nausea/vomiting and the diarrhea continued to be watery. Past history was negative. No smoking or addiction was noted in his habits. Physical exam showed stable vital signs, mild epigastric tenderness and splenomegaly of about 14 cm span. Abdominal ultrasonography was unremarkable except for a moderate splenomegaly. The stool exam contained RBC and no WBC. Gastroenterologist and rheumatologist consultations were requested. Upper gastrointestinal endoscopy merely showed chronic gastritis. The histopathology of colon biopsy was suggestive of inflammatory bowel disease (IBD).

His symptoms were worsened with fever and several episodes of bloody diarrhea. He was treated by Mesalazine, Asacol enema and antibiotics for gastrointestinal coverage.

Peripheral blood smear (PBS) showed aggregated platelets. Doppler ultrasound of the portal system reported portal and splenic veins to have increased diameter, 16 and 11 mms respectively, with normal flow, spectral waves and normal patency of the superior mesenteric artery. Abdominopelvic computed tomography demonstrated mesenteric fat stranding, thick-walled bowels and portal and superior mesenteric vein thromboses (PVT) (see Fig. 1).

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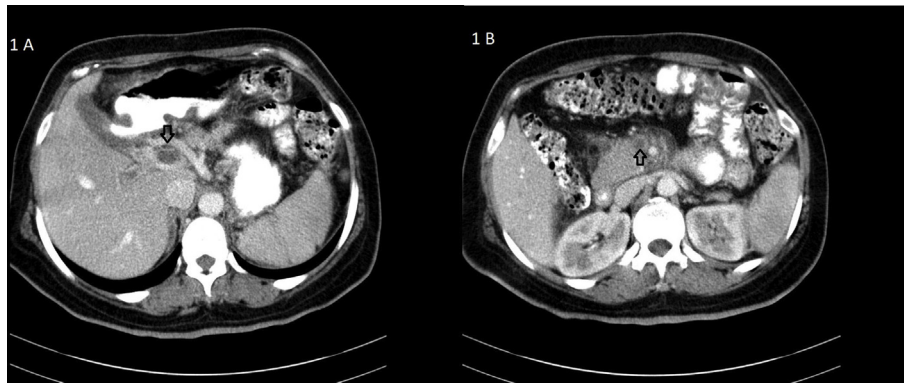


Fig. 1. Abdominopelvic computed tomography with intravenous and oral contrasts demonstrated mesenteric fat stranding, thick-walled bowels and portal vein thrombosis (1 A) and superior mesenteric vein thromboses (1 B) which are shown by black arrows.

The patient's abdominal pain turned to be intolerable, the diarrhea continued and generalized tenderness, rebound tenderness and guarding were evolved. Significant thrombocytopenia of 22000 was detected and check of peripheral blood smear, fibrinogen and fibrin degradation products (FDP) were requested. Thrombocytopenia was corrected and the patient underwent laparotomy which revealed significant bloody ascites and colon necrosis; thus, total colectomy and ileostomy were performed. The histopathologic results of resected colon showed mural necrosis and intraluminal thrombosis of vascular canals.

Two days after surgery, the patient was still critically ill in the intensive care unit and developed hypotension and dyspnea, abnormal coagulation studies, no schistocyte on PBS but thrombocytopenia, elevated fibrinogen and FDP levels that were treated with fresh frozen plasma according to hematologist consult. Unfortunately the moribund patient died and the post-mortem diagnosis was compatible with Factor V Leiden due to antiphospholipid syndrome (APS) which resulted in portal system thrombosis.

3. Discussion

We present a patient with subacute abdominal pain and diarrhea whom the catastrophic disease process rapidly progressed to peritonitis. Further investigations revealed portal and splenic vein thromboses which led to significant bowel ischemia and necrosis.

Although chronic portal vein thrombosis may present with subtle hepatic failure signs and symptoms, acute PVT usually result in abdominal distention, nausea, vomiting, rectal bleeding, splenomegaly, sepsis, intestinal infarction and perforation.²

A variety of disease processes are associated with splenomegaly that may reflect the exaggerated spleen function. These include infiltrative mechanism, immune response to infections, blood cells' destructions and sequestrations, neoplastic processes, congestion due to vein thrombosis or portal hypertension, medications, trauma, etc. Among local etiologies for portal and splanchnic vein thromboses, cirrhosis and abdominal sepsis are more commonly discussed.^{3,4} Besides, common systemic causes are myeloproliferative neoplasms and thrombophilia which results from inherited or acquired processes and may be exacerbated by some risk factors including sedentary states, iatrogenic catheter insertions, old age, obesity, surgery, trauma, hormone therapy, etc.⁵ The relationship between hypercoagulable states and certain rheumatologic diseases such as systemic lupus erythematosus and antiphospholipid syndrome has been established.⁶ In a systematic review and meta-analysis, Kaiser et al. suggested that factor V Leiden (FVL) has been a known risk factor for thromboembolic events; however, no significant interaction was detected between antiphospholipid disease and the FVL polymorphism. This indicates the additive risk of thrombosis in patients with the two risk factors. Inherited or acquired hypercoagulability in the presence of IBD is associated with PVT.

Routine screening for the Factor V Leiden is not recommended yet unless when a thromboembolic event happens in the presence of hypercoagulable states such as pregnancy, oral contraception, casting, operations, etc. It is prudent to evaluate the patient and family members carefully for probable previous events like recurrent miscarriage, deep vein thrombosis, etc.^{6,7} In acute settings, liver function tests remain usually near normal with elevated alkaline phosphatase and D-dimer.

Hepatic portal system imaging has been improved to assess the pathologic processes. Doppler sonogram may show flow abnormalities or intraluminal filling defect depending on the operator experience. Endosonography has been reported to be sensitive and specific to evaluate portal system thrombosis. Abdominal computed tomography with intravenous contrast and portal venous system magnetic resonance (MR) portography can also be helpful with the latter to follow partial thromboses, portosplenic collaterals and to make surgical plans for neoplasms involving portal system.

The standard of care is anticoagulation, systemic or catheter-directed thrombolysis through various access routes such as transjugular, percutaneous transhepatic or via the superior mesenteric artery, angioplasty & stenting and treatment of the underlying disease.⁸ Gastric varices due to splenic vein thrombosis can be managed endoscopically.

Surgical intervention is reserved for the resection of infarcted bowel and gas formation in hepatoportal system. The high recurrence rate hampers survival even with treatment, thus, it is crucial to focus on the underlying cause.⁸

To reach an early diagnosis, an emergency physician should be aware of the fact that the presence of diarrhea, abdominal pain and any history of lower gastrointestinal bleeding can be caused by an ischemic colitis, even without a risk factor⁹ and should consider the critical differentials in a timely manner.

4. Conclusion

Portal system thrombosis due to a hypercoagulable state is not uncommon and presents variably in acute and chronic settings. Ultrasonography, CT and MRI are useful diagnostic modalities. Anticoagulation and treatment of the underlying disease are the main therapies. Timely management of the portal vein thrombosis lessen adverse events although the secondary cause of the pathology principally determines the prognosis.

Conflict of interest

None.

Author contribution

M B, B CA, B H performed the research M B, B CA, B H, N S designed the research study, contributed essential reagents or tools and wrote the paper.

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