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Guess the Case From the Ochsner Archives

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INTRODUCTION

A 57-year-old man was initially seen with several days of worsening crampy abdominal pain. The pain was not localized and did not radiate. No one else in the family was sick. His only bowel movement over the past few days was with the assistance of laxatives, which caused malodorous diarrhea. The patient had no significant medical history and had never had surgery. The patient denied any bright red blood from the rectum, melena, fever, chills, nausea, or vomiting.

On physical examination, the patient was afebrile with heart rate around 100 beats/min and in mild distress. The abdomen was diffusely tender. There was no rebound tenderness or guarding, and bowel sounds were decreased. Laboratory studies revealed a white blood cell count of 20,000/μL, normal pancreatic enzyme levels, and an elevated fibrin level of 780 (normal range, 182–366 mg/dL). Liver studies showed a bilirubin level of 1.6 mg/dL (to convert bilirubin level to micromoles per liter, multiply by 17.104) and an international normalized ratio of 1.1.

Kidneys, ureters, and bladder were nonspecific, but ultrasonography of the abdomen revealed thickened and edematous small bowel. Computed tomography was then performed (Figures 1, 2, and 3).

What is the diagnosis, and what treatment would you recommend? What other studies need to be performed?

WHAT IS THE DIAGNOSIS?

The patient has mesenteric venous thrombosis. This was confirmed by computed tomography, which showed thrombosis of the superior mesenteric vein and the portal vein, with resultant mesenteric edema.

The patient underwent an exploratory laparotomy because of his diffuse abdominal pain, elevated white blood cell count, and computed tomographic findings. The surgery consisted of small bowel resection and primary anastomosis, as well as a liver biopsy. Pathologic examination showed no steatosis in the liver and no fibrosis. The small bowel had ischemic changes with no evidence of malignancy and with thrombus identified in a large mesenteric vessel. If the patient had been asymptomatic or had only a mildly tender abdomen, the initial treatment would have been immediate anticoagulation therapy, with exploratory laparotomy only if the initial treatment failed. Alternative therapy with venous thrombectomy is unlikely to be successful and is of limited value.[1] Fibrinolytic therapy is not recommended because the congested bowel is at risk for hemorrhage. Endovascular therapy, angioplasty, and/or stent placement may be used in circumstances of mesenteric insufficiency due to mesenteric artery or celiac artery occlusion.[2]

Further workup revealed no finding of thrombus source on 2-dimensional echocardiography or on

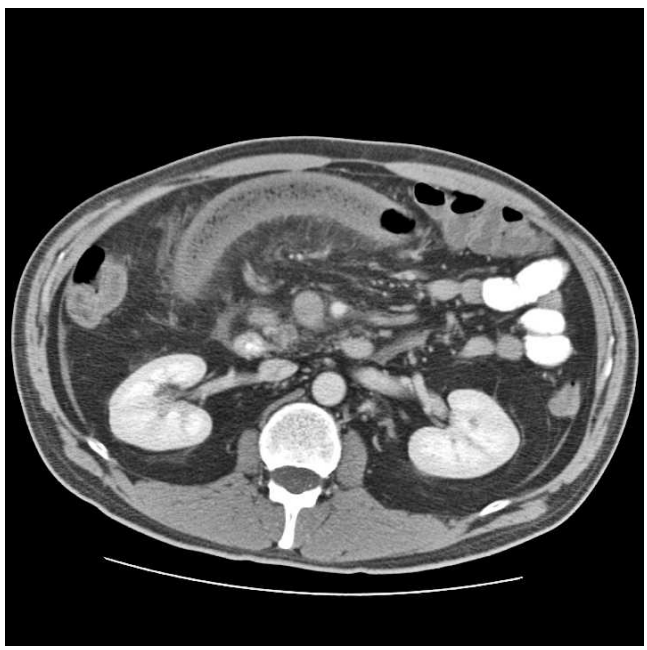

Figure 1. Computed tomographic scan showing thickened loops of small bowel.

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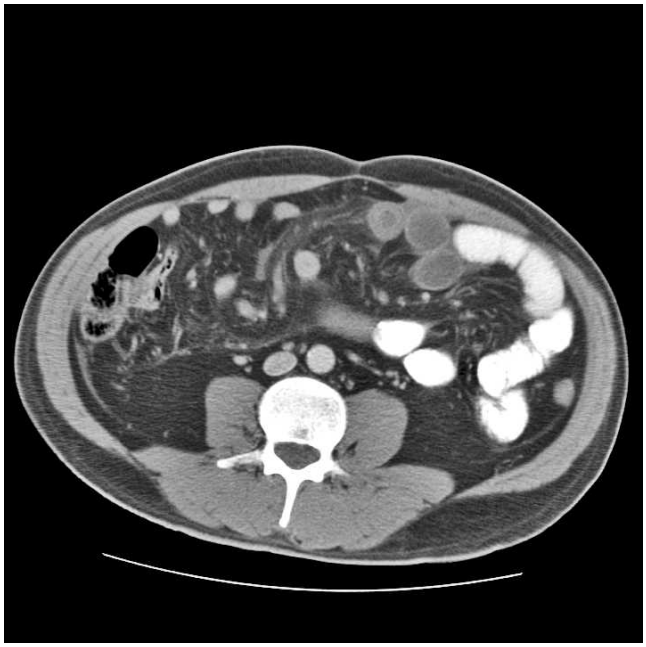

Figure 2. Computed tomographic scan showing target sign of thickened small bowel.

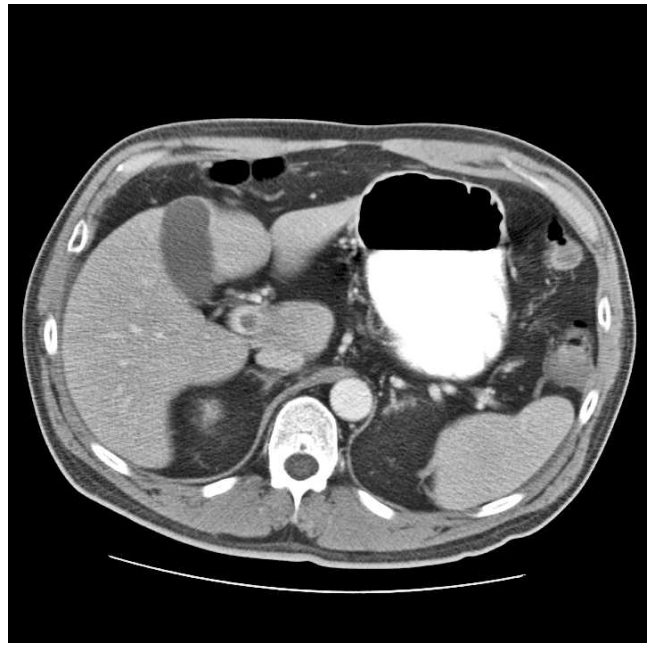

Figure 3. Computed tomographic scan showing a blood clot in the portal vein.

transesophageal echocardiography. Anticoagulation studies revealed no factor V mutation, no antithrombin III deficiency, no antiphospholipid antibody, and no lupus anticoagulant. However, the patient was found to be deficient in levels of protein C (7.0 g/dL [normal range, 8.0–14.0 g/dL]) and protein S (3.7 g/dL [normal range, 7.0–14.0 g/dL]). Heparin sulfate and warfarin sodium therapy was begun. Once the patient had a therapeutic international normalized ratio, he was discharged home.

DISCUSSION

Mesenteric venous thrombosis is a rare but deadly form of mesenteric ischemia. Mesenteric ischemia was first described by Antonio Hodgson in the latter part of the 15th century.[3] In 1935, Warren and Eberhard reported that intestinal infarction was a result of ischemia due to venous thrombosis, with a mortality rate of 34% in patients with venous thrombosis after resection. Unfortunately, despite improvements in therapy, this mortality rate still holds.[3]

One of the risk factors for mesenteric venous thrombosis is a hypercoagulable state. These risk factors include factor V Leiden mutation, protein C and S deficiencies, prothrombin 20210 mutation, lupus anticoagulant, and antithrombin III deficiency. Table 1 gives the prevalences and associated risks of these deficiencies based on work by Schick and Schick.[4]

Protein C and S deficiencies are rare but serious deficiencies that significantly increase the risk of venous thromboembolism. These account for about 5% of venous thromboembolism cases and are treated with heparin and warfarin. Proteins C and S are vitamin K–dependent clotting factors. Protein C works by degrading factors V and VIII along with fibrinogen. Protein S is a cofactor for protein C. Proteins C and S have the shortest half-lives of the

Table 1. Risk Factors for Venous Thromboembolism*

Condition	Prevalence in General Population, %	Prevalence in Persons With Venous Thrombosis, %	Increased Risk for Thrombosis
Factor V Leiden	5–15	20	3.8
Prothrombin 20210A	1–6	2	3.0
Protein C	0.2	3	25–50
Protein S	Unknown	1–2	10–15
Antithrombin III	0.02	1	10

*Based on work by Schick and Schick.[4]

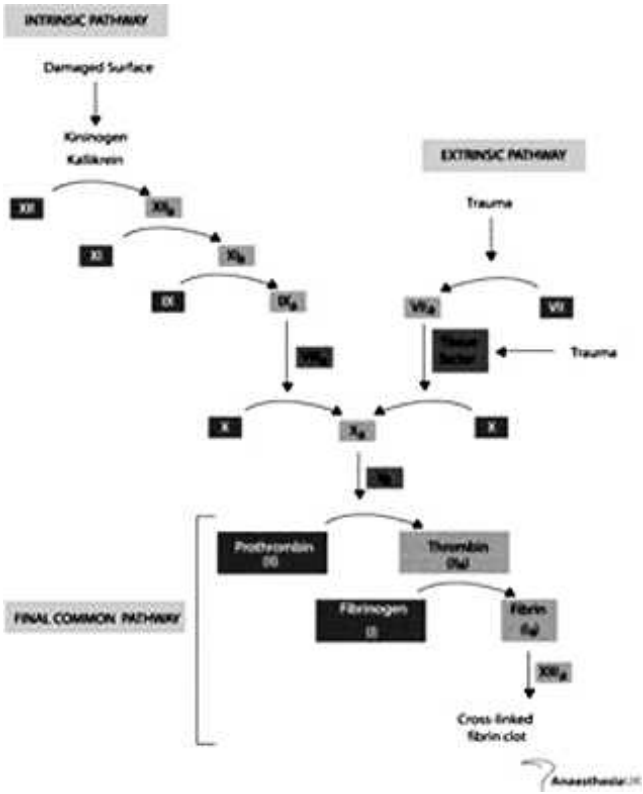

Figure 4. Clotting cascade.

vitamin K–dependent factors (factors II, VII, IX, and X and proteins C and S) and may result in a hypercoagulable state when they are depleted with warfarin therapy. Therefore, it is important to start patients on heparin whenever possible if warfarin is to be used, in an effort to prevent warfarin skin necrosis.[5]

Factor V Leiden is the most common congenital clotting disorder and accounts for about 20% to 30% of spontaneous venous thrombosis cases.[5] The defect is at the binding site of factor V for activated protein C and results in resistance. Treatment is with heparin and warfarin.

The key to anticoagulation is antithrombin III (Figure 4). This protein works by binding to and inhibiting thrombin along with factors IX, X, and XI. This is the site of action for heparin, which works by binding to and activating antithrombin III. Patients with this disorder will not respond to heparin and are treated with antithrombin III concentrate or with fresh-frozen plasma, followed by heparin and warfarin.[5]

Lupus anticoagulant is an antiphospholipid antibody that prevents binding of clotting factors. Patients with lupus anticoagulant have prolonged partial thromboplastin time that is not corrected with fresh-frozen plasma. Treatment is with heparin and warfarin.[5]

Venous thromboembolism is a serious and life-threatening event that should include a workup for hypercoagulable states. Surgical management often requires resection of affected areas, followed by lifelong anticoagulation with warfarin, even in patients with no known hypercoagulable state.[6]

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