CLINICAL VIGNETTE

Acute Portal Vein Thrombosis

Sittiporn Bencharit, M.D. and Michael G. Quon, M.D.

Patient A

A 75-year-old female with history of NSAID-induced gastric ulcer was admitted for severe abdominal pain of several days duration. She also had thrombocytosis treated with hydroxyurea. CT of the abdomen revealed complete thrombosis of the splenic vein and intrahepatic and extrahepatic portal vein extending into the superior mesenteric vein. There was wall thickening of the ileum, moderate mesenteric edema, and vascular engorgement.

She was treated with IV heparin for acute portal vein thrombosis. However, her condition deteriorated, and she was taken to the operating room, where a long segment of necrotic small bowel was resected. She had a prolonged hospitalization stay, requiring TPN for short gut syndrome and continued with anticoagulation.

Patient B

A 54-year-old female with a history of alcoholic cirrhosis with prior variceal bleed was admitted to the hospital after several days of abdominal pain, nausea, and vomiting. CT of the abdomen revealed cirrhotic liver and a thrombus in the portal vein extending into the superior mesenteric vein. Both colon and small intestine had thickened walls.

In spite of her INR of 1.7 and platelet count of 23 K, she was started on heparin infusion and her abdominal pain resolved within 24 hours. She was discharged on warfarin.

Discussion

The portal vein is formed from the splenic and superior mesenteric veins. Thrombus formed in the portal vein may extend into splenic and superior mesenteric veins. Portal vein thrombosis (PVT) can be seen in 5-10% of patients with portal hypertension with increased incidence with more severe liver disease. Ultrasonography in unselected cirrhotic patients found prevalence of PVT ranging from 10-28%. In Sweden, the prevalence of PVT is 1% in general population based on autopsies between 1970-1982.[1]

Predisposing conditions for PVT include reduced portal blood flow, vascular endothelial injury, and hypercoagulable state. In non-cirrhotic patients, myeloproliferative disorders can be found in 35% of the cases. Patients should be screened for hypercoagulable states. The 2009 American Association for the Study of Liver Diseases guideline recommends screening for antiphospholipid syndrome, Factor V Leiden, Factor II gene mutation, protein C, and protein S deficiency.[2]

Endothelial activation of prothrombotic factors can occur as a result of various intra-abdominal inflammatory diseases including pancreatitis, cholecystitis, appendicitis, and abdominal trauma. Acute pancreatitis is the most common association, followed by cholecystitis and cholangitis.[2]

In cirrhotics, in addition to reduced blood flow, there appears to be prothrombotic state, in spite of elevated prothrombin time. A decrease in liver derived procoagulant factors is usually overcompensated by a decrease in anticoagulant factors and an increase in procoagulant factors VIII and Von Willebrand.[3]

Symptoms of PVT depend on the extent and the speed of the obstruction of the portal blood flow. Some patients can be asymptomatic; however, abdominal pain associated with nausea and vomiting, hematemesis, fever, and marked systemic inflammatory response can be seen. When mesenteric vein thrombosis occurs, intestinal ischemia, and infarction can develop. In cirrhotics, elevated portal pressure may lead to variceal bleeding. Acute septic PVT often present with high spiking fevers and liver tenderness. It is often associated with an intra-abdominal focus of infection.

PVT can be diagnosed with ultrasound, demonstrating hyperechoic material in the vessel lumen. CT without and with IV contrast is the preferred imaging study as it can detect underlying causes and assesses the extent of the thrombus and extent of intestinal ischemia. In patients allergic to IV contrast, MRI can be an alternative with a sensitivity of 100% and a specificity of 99%. Both are more sensitive than ultrasonography as portal veins may be difficult to visualize on ultrasound. The differential diagnosis of PVT includes malignant invasion of portal vein, most commonly hepatocellular carcinoma or constriction of the portal vein by the tumor, as in pancreatic cancer or cholangiocarcinoma.[4]

Treatment of acute PVT is acute anticoagulation. The success of recanalization of the thrombus, depends on the speed of anticoagulation.[5]

Low molecular weight heparin is recommended with transition to oral anticoagulation when stable. Thrombolytic therapy, administered through superior mesenteric artery or transjugular intrahepatic puncture into the portal vein, had resulted in complete or partial canalization of the thrombus in in 75%. However, 60% developed major procedure related complication.[6] Surgical thrombectomy is reserved for patients undergoing surgery for intestinal infarct.

Untreated acute PVT may lead to intestinal infarction, although the relative risk is unknown as there are those who remain asymptomatic. Limited data suggests that spontaneous recanalization of the thrombus without therapy is rare.

Anticoagulation can lead to complete recanalization in 41.5% and partial recanalization in 66.6%. Duration of therapy is 6 months, if there is no other prothrombotic risk factor.[2] However, rethrombosis of the portal vein occurs in 38% of patients after discontinuation of anticoagulation therapy.[5]

Anticoagulation can lead to bleeding complication in 0-6%. Treatment in cirrhotics results in bleeding complication in 3-4%, usually in the form of variceal bleeding. To mitigate the risk of bleeding, cirrhotics should undergo endoscopic treatment of varices, before starting on long-term anticoagulation.[4]

Thrombocytopenia less than 50 thousand is a known risk factor for increase bleeding in cirrhotics treated with warfarin. In addition, it is often difficult to maintain narrow therapeutic window, INR 2-3. Direct acting anticoagulants have been effective in the treatment of PVT and have comparable bleeding risk to warfarin.[7,8] More data are required to see if these agents should be the anticoagulant of choice.

Summary

Both patients presented with acute portal vein thrombosis with signs of intestinal ischemia on CT scan. Both were immediately started on heparin infusions. Patient A developed small bowel infarction requiring intestinal resection, while Patient B's symptoms resolved within 24 hours. Perhaps Patient B, who is cirrhotic, developed more extensive collateral venous drainage, sparing her from intestinal infarct.

Patient A has done well. She is now off TPN and is able to maintain her weight. She is maintained on warfarin with INR around 2.

Patient B was treated with warfarin for 8 months. Keeping her INR between 2-3 had been a challenge. Repeat CT of the abdomen showed complete recanalization of her PVT, and her anticoagulation was discontinued. However, 9 months after discontinuation, she developed abdominal pain, nausea, and vomiting. A CT scan demonstrated a new portal vein thrombosis with edema of the large and small intestine. She was started on heparin drip and transition to apixaban. The duration of her anticoagulation has yet to be determined.

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