Pregnancy Complicated by Chronic Portal Vein Thrombosis: A Case Report

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Introduction:

Women of childbearing age account for less than 25% of noncirrhotic portal vein thrombosis cases, with the presence of JAK 2 mutation in around 17-35 % of all patients with PVT. This case reports a pregnant female with extra hepatic portal venous obstruction thrombosis.

Case Presentation:

A 38-year-old G5P4004 Hispanic female with no past medical history presented at 8-weeks gestation, with two-week history of RUQ and epigastric pain complicated by nausea and emesis. She denied any fevers, chills, diarrhea, sick contacts, travels, and no prior pregnancy complications. Physical exam showed tenderness to palpation in the epigastrium and RUQ, with splenomegaly and normal bowel sounds. She remained hemodynamically stable, in no acute distress. Her hemoglobin was 12.6 and platelet count 194, PT 15.3, PTT 72.8, with normal liver enzymes. Ultrasound demonstrated complete thrombosis of the main, right and left portal veins (PV), as well as splenomegaly, with trace perihepatic and splenic ascites (Figure 1). MRI demonstrated thrombus extending into the distal superior mesenteric vein and portions of the splenic vein (Figure 2) as well as perisplenic, gastohepatic and retroperitoneal varices compatible with portal hypertension. CT demonstrated cavernous transformation of the PVs (Figure 3). She underwent emergent transjugular intrahepatic portosystemic shunt (TIPS) with percutaneous thrombectomy of the superior mesenteric vein, splenic vein and hepatic PVs. Her course was complicated by intraperitoneal hemorrhage and subcapsular hepatic hemorrhage, requiring percutaneous embolization. She later underwent emergent laparotomy with perihepatic packing and splenectomy for splenic ischemia and rupture, requiring mass transfusion. Unfortunately, intrauterine demise was noted, requiring dilation and curettage. JAK2 V617F mutation was discovered prior to discharge on anticoagulation.

Discussion:

Compared to acute PV thrombosis, chronic thrombosis tends to present with signs of portal hypertension such as hypersplenism, ascites, varices and formation of portal cavernoma. Risk factors include pregnancy and JAK2V617F mutation associated with myeloproliferative disorders.

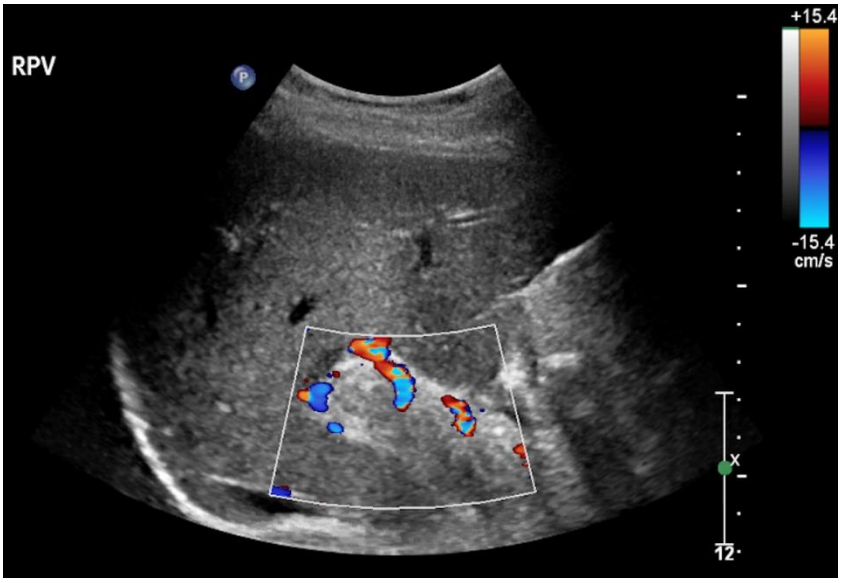

Figure 1: Ultrasound image showing complete thrombosis of the right portal vein.

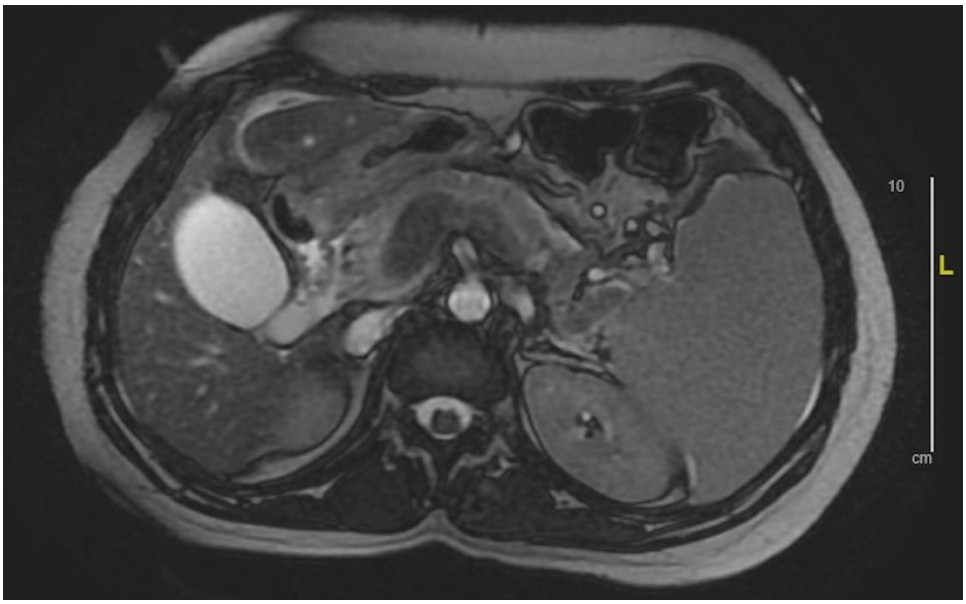

Figure 2: Axial MRI image showing complete thrombosis of the main and intrahepatic portions of the portal vein with thrombus extending retrograde into the distal superior mesenteric vein and into the distal and midportions of the splenic vein. There are perisplenic varices and splenomegaly compatible with portal hypertension.

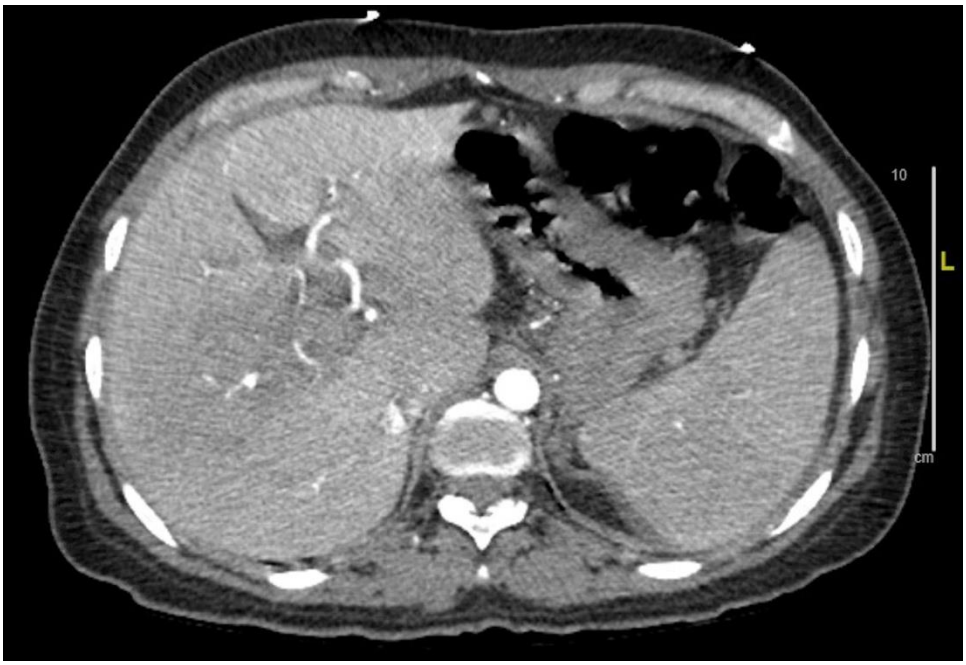

Figure 3: Axial CT scan image showing intrahepatic portal vein thrombosis with cavernous transformation. Splenomegaly is also noted.