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Case report

Acute portal vein thrombosis in a 59-year-old male with JAK2 V617F mutation

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ABSTRACT

Portal vein thrombosis is an uncommon finding that typically arises in the context of cirrhosis. In the acute setting, it may present with abdominal pain, portal hypertension, ascites, gastrointestinal bleeding, or mesenteric ischemia. Local risk factors that predispose its formation include: cirrhosis, hepatocellular carcinoma, pancreatitis, and intraabdominal infection. Systemic factors, including hypercoagulable states and sepsis, also pose an increased risk. JAK2 V617F positive myeloproliferative disorders are associated with systemic prothrombotic states and are a less frequently identified cause of portal vein thrombosis. We present a case of acute unprovoked portal vein thrombosis diagnosed in a 59-year-old male without local disease factors. Computed tomography, magnetic resonance cholangiopancreatography, and ultrasound demonstrated the presence of portal vein thrombosis with neighboring periportal and pancreatic head edema. Peripheral blood testing detected the presence of JAK2 V617F mutation. The patient was discharged on 6-month anticoagulation therapy and outpatient follow-up.

Introduction

Portal vein thrombosis (PVT) is defined as a partial or complete obstruction of the portal vein by a clot resulting in impeded flow [1]. It is an infrequent occurrence that has been increasingly recognized with the broader utilization of radiological imaging in clinical practice [1]. Population prevalence is estimated to be 1% with 0.6%-16% occurring in asymptomatic liver disease, 15% of patients awaiting liver transplant, and 35% in the setting of cirrhosis with hepatocellular carcinoma [2].

The portal vein is formed by the confluence of the splenic and superior mesenteric veins. It divides superiorly at the porta hepatis into left and right branches supplying the left and right hepatic lobes, respectively. PVT forms in the trunk of the portal vein and can extend into the left and right intrahepatic branches or the splenic and superior mesenteric

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Table 1 – Classifications of PVT based on anatomic extent by Yerdel et al [5].

Classification	Occlusion of portal vein	Extension of thrombus
Grade 1	Minimal or partial thrombosis of the portal vein with <50% occlusion	With or without minimal SMV involvement
Grade 2	>50% and complete thrombosis of the portal vein	With or without minimal SMV involvement
Grade 3	Complete thrombosis of the portal vein	Proximal SMV is occluded with patent distal SMV
Grade 4	Complete thrombosis of the portal vein	Proximal and distal SMV occlusion

Abbreviation: SMV, superior mesenteric vein.

Table 2 – Differential diagnosis for PVT associated findings with risk and frequency of underlying causes.

Cause	Labs	Clinical findings	Imaging	PVT frequency	PVT risk
Liver cirrhosis	CBC, liver chemistry [3]	Ascites, jaundice, hepatic encephalopathy, gastrointestinal bleeding, portal hypertension, abdominal pain [2]	Irregular liver outline, portal vein dilation [2]	Incidence: 11.2%-16.6% [2,3]	Odds ratio: 17.1 in cirrhosis with primary hepatic cancer, 5.2 in cirrhosis without primary hepatic cancer[3]
Liver carcinoma	Serum AFP levels [9]	Advanced stage, major vessel involvement, low serum albumin, high serum AFP levels [3]	Filling defect with rim enhancement of vessel wall, disruption of vessel wall, expansive effect due to tumor mass [2]	Incidence: 20%-44% [3], 35% in combination with cirrhosis [2]	
Liver transplant	-	Decreased caliber of portal vein, donor/recipient portal vein diameter mismatch [2]	-	Incidence: 13.8% no portosystemic shunt, 38.9% prior portosystemic shunt [3]	-
Pancreatitis	Serum amylase/ lipase [15]	Premature activation of digestive enzymes and inflammation, acute severe epigastric pain radiating to the back [15]	Acute pancreatitis: Diffuse enlargement of pancreas, heterogeneous enhancement, peripancreatic stranding [15]	Incidence: 23% in acute pancreatitis, 57% in pancreatic necrosis [2]	-
Hypercoagulable states (factor V leiden, protein C deficiency, protein S deficiency, antithrombin III deficiency, prothrombin mutation, antiphospholipid syndrome, paroxysmal nocturnal hemoglobinuria)	Factor V mutation, protein C & S levels, ATIII, G20210A, cardiolipin, lupus anticoagulant, anti-beta2 glycoprotein antibodies, CD55/CD59 [2]	Acute PVT: Abdominal pain, fever, ascites, splenomegaly [2] Chronic PVT: Recurrent upper abdominal pain, portal hypertension, varices, splenomegaly, anemia, thrombocytopenia [2]	Acute PVT: hypoechogenic/ hypodense/ hypointense thrombus, absence of porto-systemic collaterals [2] Chronic PVT: absent (fibrotic) portal vein with cavernoma, portal hypertension, wall/thrombus calcifications hyperdensities on CT [2]	-	Relative risk: 10-20 in Protein C, Protein S, and Antithrombin III deficiency; 8 in Antiphospholipid syndrome [3]
Myeloproliferative disorders (PCV, ET)	JAK2 V617F [2,3,7]			30%-40% [2,3]	Odds ratio: 3.0 [10]

veins. Proposed grading of PVT by Yerdel et al is based on the percentage of portal vein occlusion and extension into the superior mesenteric vein (Table 1) [1–5]. Mesenteric vein involvement is associated with an increased risk of intestinal infarction and mortality [2]. Over time, if portal vein obstruction persists, surrounding vessels can dilate to permit collateral flow resulting in the formation of a cavernoma [1].

Predisposition to PVT is based on features of Virchow's triad: hypercoagulability, endothelial injury, and stasis [1]. Local factors, such as cirrhosis and intraabdominal infections carry an increased risk of developing PVT. Reduced portal flow and abdominal inflammation leads to endothelial activation and release of prothrombotic factors [2]. Systemic factors including hypercoagulable states and sepsis predispose