Portal Vein Thrombosis- Interesting Case Scenarios

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Abstract:
Portal vein thrombosis (PVT) refers to obstruction of blood flow in portal vein and its branches due to thrombus. Its prevalence in cirrhosis ranges from 5-15 and accounts for 5-10 of cases of portal hypertension. PVT can occur in two clinical entities, can be acute or chronic PVT. The clinical features and management differs between acute and chronic. Cause of PVT in cirrhosis is due to reduced portal vein blood flow, endotoxemia, acquired and inherited thrombophilic conditions. Risk factors for PVT can be local and systemic. Local risk factors include malignancies of abdominal organ most commonly pancreatic and hepatocellular carcinoma accounts for 21-242, Others include cirrhosis, appendicitis, pancreatitis, cholecystitis, diverticulitis, IBD or following surgeries such as colectomy, cholecystectomy, gastrectomy, liver transplantation, TIPS. Other thrombotic risk factors include Myeloproliferative disorders, Factor V Leiden mutation, Protein C and S deficiency, Antiphospholipid syndrome, OCP, pregnancy. We present interesting case scenarios of PVT detected in three different clinical situations such as Essential thrombocytosis, Appendicitis, Pancreatitis.

Keyword: Portal vein thrombosis (PVT), Inflammatory bowel diseases (IBD), Transjugular intrahepatic portosystemic shunt (TIPS), Oral contraceptive pills (OCP), LMWK (Low molecular weight heparin), PHT (Portal hypertension), CECT (contrast enhanced computed tomography)

Introduction:
Portal vein thrombosis is due to obstruction of portal vein and it branches by thrombus. It occurs in 1% of general population and 5-15% of cirrhotics. PVT can be recognized as two different clinical entities such as Acute PVT and chronic PVT. It accounts for 30-75% cases of PHT in adults and children [AASLD guidelines]. The clinical features, management differs between acute and chronic portal vein thrombosis. Acute portal vein thrombosis present as abdominal pain, abdominal distention, variceal bleeding, persistent non-spiking fever, bloody diarrhea, refractory ascites. Acute PVT is rarely reported in children. All manifestations of acute PVT are reversible. Extension of thrombus into superior
mesenteric vein causes mesenteric ischemia and intestinal infarction. It manifests as ascites, bloody diarrhea, and abdominal pain lasting more than 5-7 days. Chronic PVT may be asymptomatic, presents with splenomegaly, and pancytopenia. Gastrointestinal bleed is better tolerated than other cause of PHT. Occurrence of ascites or encephalopathy is uncommon. Hepatopulmonary syndrome occurs in 10% of cases. Risk factors for PVT can be local and systemic. Local risk factors include malignancies of abdominal organ most commonly pancreatic and hepatocellular carcinoma accounts for 21-24%. Others include cirrhosis, appendicitis, pancreatitis, cholecystitis, diverticulitis, IBD or following surgeries such as colectomy, cholecystectomy, gastrectomy, liver transplantation, TIPS. Other thrombotic risk factors Myeloproliferative disorders, Factor V Leiden mutation, Protein C and S deficiency, Antiphospholipid syndrome, OCP, pregnancy. Myeloproliferative disorders can be identified by screening for JAK2 mutation which is identified in 21-37% of patients with PVT. Combination of Splenomegaly and platelet count more than 200000/mm³ suggests presence of myeloproliferative disorder. Bone marrow biopsy shows dystrophic megakaryocytes.

We present 3 interesting case scenarios with portal vein thrombosis. We present clinical situations such as Appendicitis, Pancreatitis and Essential thrombocytosis apart from chronic liver disease in which portal vein thrombosis can be identified.

CASE.1 PVT DUE TO ESSENTIAL THROMBOCYTOSIS
59 years male presented with postprandial increase of abdominal pain, fever and loss of appetite. He had no episodes of UGI bleed. Clinically his physical examination was normal. On investigation we found he had elevated platelet count 7, 50,000 cell/mm³. Peripheral smear showed increased platelets and giant platelets. Liver function tests were within normal limits. Prothrombin time-117 sec and INR- 8.7. His Doppler USG showed thrombosis of portal vein (PV), superior mesenteric vein (SMV), and splenic vein (SV). CECT showed complete thrombosis PV and SMV with collaterals. (Fig.1)

We carried out prothrombotic work up such as Protein C, S, Antithrombin III, Anticardiolipin antibodies and found only protein S deficiency. Bone marrow aspiration showed hyper cellular marrow. We arrived at a diagnosis of PVT due to Essential thrombocytosis. The patient was not affordable for further workup such as JAK2 mutation. He was treated with Anticoagulation such LMWK Heparin, warfarin. The patient was regularly followed with INR and his dose of anticoagulants adjusted he had symptomatically improved.
CASE.2 PVT DUE TO PANCREATITIS
18 year female presented with complaints of continuous, boring epigastric pain with radiation to back for 6 months. It was associated with abdominal distention for 2 months. On examination her respiratory system revealed reduced breath sounds with basal crackles. She had ascites with diffuse tenderness. Complete blood count showed hemoglobin of 10.2 gm, platelet count of 1.20 L. Her serum amylase- 374IU, Lipase – 1551IU. Liver function tests normal. Ascitic fluid was hemorrhagic with lipase of 4039IU. CECT showed calcification in head of pancreas, multiple collaterals in Porta and pancreatic bed. MRCP showed Bilateral pleural effusion, thrombosis of main portal vein and confluence, thick walled collection 3.5x5 cm in lesser sac. Doppler Ultrasonogram showed thrombosis of portal vein, Splenomegaly, periportal collaterals and cavernous transformation of portal vein. ERCP showed a leak in pancreatic duct at genu. The leak was bridged with 5Fr7 cm pancreatic stent (Fig.2). Ascites improved and patient had weight gain.

CASE.3 PVT DUE TO APPENDICITIS
28 year old male presented with lower abdominal pain, fever and loose watery stools for 20 days. History of yellowish discoloration of urine and eyes for 7 days. Examination of abdomen showed mass in right iliac fossa with tenderness. USG showed aperistaltic blind ending non compressible dilated bowel loop in RIF with probe tenderness. Doppler showed PV to be thrombosed. CECT showed PV thrombosis (Fig.3), features of appendicitis. The patient was treated with higher antibiotics and anticoagulation.

FIG.3 PORTAL VEIN THROMBOSIS IN MAIN PORTAL VEIN

DISCUSSION:
Portal vein thrombosis may be acute or chronic. The clinical features and management are different for both conditions. Acute PVT to be considered in any patient with abdominal pain lasting more than 24 hours. Doppler Ultra sonogram forms the most important diagnostic modality. Thrombus within lumen has a sensitivity of 60% and specificity of 100%. Other findings are collaterals around porta hepatitis, loss of respiratory variation of PV with diameter > 13 mm. Presence of arterial flow within thrombus indicates malignant thrombus. Other investigating modalities are contrast enhanced CT, MRI, EUS, spin echo MRA. Spin echo has sensitivity of 85% and
specificity of 95%. EUS has sensitivity of 81% and specificity of 93%. EUS can detect small non-occluding thrombus. The goal of treatment in acute PVT is recanalisation of obstructed vein. It can be achieved with thrombolytic therapy with rtPA inhibitors. Anticoagulation is done with LMWK heparin followed by warfarin. In acute PVT anticoagulation indicated for at least 6 months. Patients of acute PVT with underlying prothrombotic conditions to be treated with lifelong anticoagulation. In chronic PVT anticoagulation is indicated when there is extension of thrombus into SMV. In setting of chronic liver disease associated with underlying prothrombotic state to be started on long term anticoagulation.

**BIBLIOGRAPHY**


