Chronic Idiopathic Non-Cirrhotic Portal Vein Thrombosis Treated with a Mesocaval Shunt Procedure and Anticoagulation

Hamid Shaaban · Nihar Shah · Ibrahim Sidhom

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Abstract Portal vein thrombosis (PVT) was first reported in 1868 by Balfour and Stewart and is a medical condition in which the lumen of the portal vein is completely or partially obstructed due to the presence of a thrombus [1]. Inherited (Factor V Leiden and Prothrombin gene mutation G20210A, Protein C, S and Anti thrombin III deficiency) and acquired thrombophilias (Lupus Anticoagulant, myeloproliferative diseases, malignancy, surgery and trauma) account for majority of the cases of PVT.

Portal vein thrombosis (PVT) is considered a rare even but its prevalence in the general population among cirrhotic patients ranges between 4.4 and 15 %, and is responsible for about 5–10 % of overall cases of portal hypertension [2]. The classic triad of PVT is splenomegaly, ascites, and variceal dilation [3]. The clinical presentation is mainly dependent of the acute or chronic nature of this diseases entity which determines the absence or presence of the development of the collateral circulation respectively. Intestinal congestion and ischemia, with abdominal pain, diarrhea, rectal bleeding, abdominal distention, nausea, vomiting, anorexia, fever, lactic acidosis, sepsis, and splenomegaly are common features of acute PVT. In contrast, chronic PVT can be completely asymptomatic, or characterized by splenomegaly, pancytopenia, varices, and, rarely, ascites [4].

63-Year-old Hispanic Female with history of hypertension, diabetes mellitus, coronary artery disease, hypothyroidism presented with complaints of epigastric pain and bloody vomiting. She initially had epigastric discomfort and two episodes of hematemesis. She denied any alcohol or oral contraceptive use. Screening mammogram/Pap smear and colonoscopy were negative for malignancy. Abdominal examination was unremarkable except for mild epigastric tenderness. She had no stigmata of liver cirrhosis. Laboratory analysis revealed a white blood cell count of $8,500/\text{mm}^3$ (neutrophils 74 %, lymphocytes 6 %), hemoglobin level of 10.5 g/dL, hematocrit of 36.8 %, MCV 85, platelets $157,000/\text{mm}^3$. Her electrolytes, liver function tests and aminotransferases were within normal limits. An upper gastrointestinal endoscopic examination was done which revealed grade 4 esophageal varices with fresh blood in the distal esophagus which were subsequently ligated. She was started on octreotide and propranolol. During the course of hospitalization, she had another episode of GI bleed requiring emergent endoscopic intervention. As part of her diagnostic workup she underwent a liver biopsy and pathology revealed only focal mild portal fibrosis with mild micro and macro vesicular steatosis but no evidence of cirrhosis (Fig. 1). A Doppler Ultrasound was also done that revealed PVT and concomitant portal hypertension. An extensive hypercoagulable work up was done which included thrombin, prothrombin and partial thromboplastin times, protein C and S levels, anti thrombin III, Prothrombin gene mutation G20210A, Factor V Leiden, Lupus anticoagulant, Anticardiolipin antibodies, Homocysteine level and they were all negative. We also tested her blood for flow-cytometry for CD 55 and CD 59 but the test was normal and in the process effectively ruled out paroxysmal nocturnal hemoglobinuria. She also was also tested for the JAK 2 mutation (sometimes positive in myeloproliferative

H. Shaaban (✉) · I. Sidhom
Department of Hematology/Oncology, St Michael's Medical Center, 111 Central Avenue, Newark, NJ 07102, USA
e-mail: hamidshaaban@gmail.com

N. Shah
Department of Gastroenterology, St Michael's Medical Center, Newark, NJ, USA

Fig. 1 Liver biopsy H&E with mild inflammation and no fibrosis

states) and it was negative. We gave her the clinical diagnosis of chronic idiopathic non-cirrhotic PVT. She subsequently had a meso-caval shunt done to relieve the portal hypertension. She was started on warfarin anticoagulation 3 days later with close monitoring for bleeding. She clinically got better and was then discharged home.

The prevalence of idiopathic PVT is not well known but experts agree it is rare and it is hypothesized that a combination of local and systemic factors contribute to its

development. In our case, it is not clear if the patient had an idiopathic portal hypertension that resulted in the development of the PVT. Acute cases need at least 3 months of anticoagulation and chronic cases need porto-systemic shunting. The use of anticoagulation in chronic cases should be decided upon on a case by case basis weighing the risk of bleeding versus thrombosis [5]. We opted to give long-term anticoagulation to our patient to prevent re-thrombosis of the meso-caval shunt/graft.

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