Road Blocked without Reason: An idiopathic case of acute portal vein thrombosis?

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LEARNING OBJECTIVES

1) To list the possible causes and clinical presentation of portal vein thrombosis in a non-cirrhotic patient.
2) To outline the algorithm for diagnosiss and management of acute portal vein thrombosis in the hospital setting.

CASE PRESENTATION

A 54-year-old female patient with a history of GERD, gastric bypass 1 year ago, presented with abdominal pain associated with fever, watery diarrhea and nonbloody-nonbilious vomiting for 3 days. Physical exam was significant for hyperactive bowel sounds, diffuse abdominal tenderness, with no signs of rebound, guarding, and/or rigidity. Laboratory analysis was unremarkable. On day 2, the patient presented with a significant increase in her liver function tests (Figure 1).

A computerized tomography of the abdomen and pelvis with intravenous contrast showed geographic hypodensity in the spleen, consistent with splenic infarction. Subsequently, MRCP and MRI of the abdomen with and without contrast showed a wedge-shaped signal abnormality within the right hepatic lobe due to thrombosis of a distal portal vein branch within hepatic segment V, with the remainder of the portal veins patent.

Hematology workup for secondary causes of thrombosis episodes were negative (protein C/S, factor V Leiden, ANA, anti-smooth muscle, anti-mitochondrial, B2-glycoprotein IgM/IgG, Cardiolipin IgM/IgG). JAK2 V617F mutation and JAK2 Exon 12 & 13 mutations, total RBC deficiency, and RBC Types II and III deficiencies were also negative. Age-appropriate cancer screening tests were unrevealing. Multidisciplinary decision making recommended medical management with full anticoagulation. The patient was started on therapeutic enoxaparin and discharged with hematology outpatient follow up.

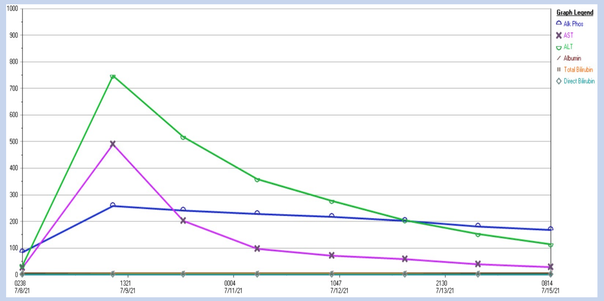

Figure 1. Liver profile trending during hospitalization course.

DIAGNOSTIC & MANAGEMENT ALGORITHM

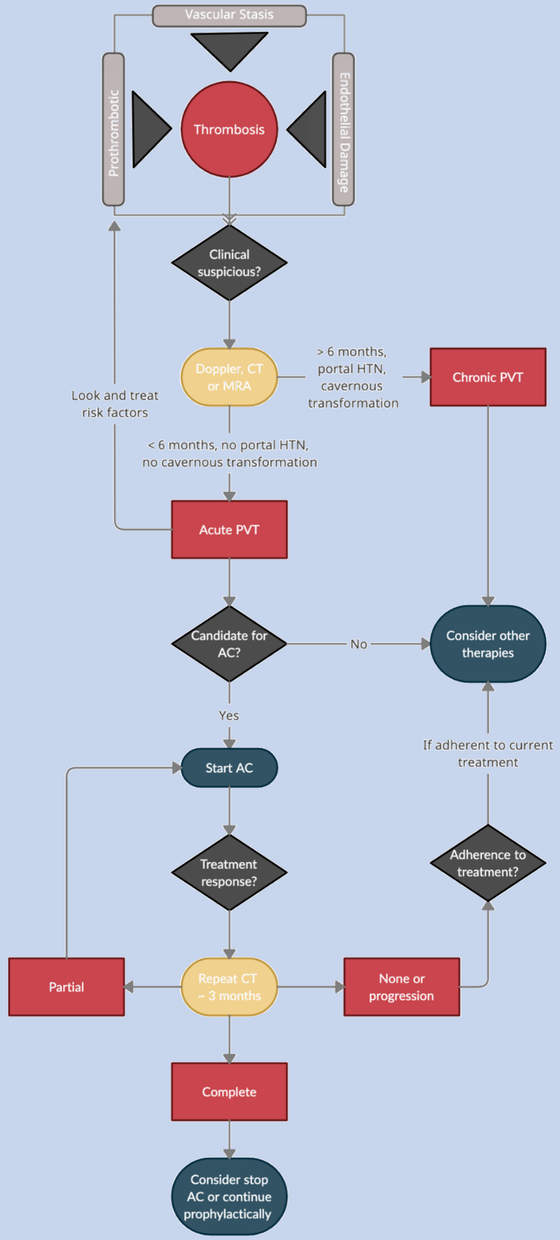

Figure 2. Algorithm of management of acute portal vein thrombosis.

DISCUSSION

Portal vein thrombosis (PVT) is a condition that results from thrombosis in the portal veins and subsequent obstruction of blood flow through the portal vascular system, ultimately precipitating with liver damage. PVT can be acute if it is present for less than 6 months prior to diagnosis. PVT is more common in cirrhotic patients (0.6% to 16%) [1]. However, it results challenging to discern the etiology in non-cirrhotic patients. It can be caused by a spectrum of conditions that affect the Virchow's triad [2].

Myeloproliferative disorders are the leading causes of PVT, reported in approximately 30% of patients. Other prothrombotic diseases have been identified as contributing factors, including paroxysmal nocturnal hemoglobinuria, antiphospholipid syndrome, protein C and S deficiencies, among others. Local intra-abdominal inflammatory conditions, such as malignancy (23%), acute pancreatitis (21%), and less frequently, intra-abdominal surgical procedures. However, in approximately 25-30% of cases no etiology is identified despite an exhaustive study [1, 2].

Acute PVT in non-cirrhotic patients can be accompanied with vague abdominal pain and fever. Often, the liver function tests, and another laboratory analysis can be not significant [1]. A doppler ultrasound (89-93% sensitivity and 92-93% specificity) has been considered the first-line diagnostic tool. However, CT digital subtraction angiography (DSA 91% sensitivity and 100% specificity) or magnetic resonance angiography (MRA 100% sensitivity and 98% specificity) are considered the gold standard [3].

Management with full anticoagulation for at least 3 to 6 months is recommended (conditional recommendation, very low level of evidence) for acute PVT, as it prevents thrombus extension, promotes recanalization, and reduces the rate of mortality [4, 5]. Low-molecular weight heparin continues to be the anticoagulant of choice. The advent use of direct oral anticoagulants (DOAC) have proven efficacy in acute PVT, but further studies are needed to be validated in clinical practice. Follow-up is imperative to evaluate the response to treatment. Most cases have a regressive disease course (47%), with a decrease in thrombus size or degree of occlusion. Nonetheless, if the occlusion has progressed, reconsideration of current therapy must be considered [6].

CONCLUSIONS

Our case of acute PVT in a noncirrhotic patient, with no history of prior liver disease and remote surgical history and no other apparent risk factors or identifiable underlying etiology could be found after extensive investigation was conducted. Moreover, some studies have stated that isolated thrombosis of an intrahepatic portal vein branch, either lobar or segmental, is considered a unique situation [7]. In such cases, early recognition of nonspecific abdominal symptoms followed by relevant workup and diagnostic tests can lead to early diagnosis and prompt initiation of anticoagulation therapy in the hospital setting, which is pivotal to optimize the clinical outcome of the patient (Figure 2).

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