Case Report

Antiphospholipid Syndrome Presenting as Cavernous Transformation of Portal Vein Secondary to Portal Vein Thrombosis

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Introduction

Cavernous transformation of portal vein is a rare disorder caused by long standing portal vein thrombosis. As a compensatory mechanism there is dilatation of small blood vessels in and around the portal vein in about 3 to 4 weeks. However despite the opening of collaterals there is development of significant portal hypertension that may manifest after some time in the form of variceal bleed or splenomegaly leading to hypersplenism. The diagnosis is done on abdominal ultrasound, portal vein Doppler and CT angiography. Portal vein thrombosis can be idiopathic or secondary to cirrhosis, thrombophilia, myeloproliferative disorder or malignancy (hepatic or non-hepatic). Treatment of portal hypertension varies from beta blockers, anticoagulation to shunt surgeries. Hypersplenism leading to thrombocytopenia can be a limitation to anticoagulation therapy.

Case Report

A twenty five year old married lady visited Medical OPD as a referral case for treatment of hepatitis B. She was diagnosed as having hepatitis B during her antenatal visit. Patient was primigravida with no other health related issues. She had spontaneous vaginal delivery at 37 weeks. Her baby boy was given Hepatitis B Immunoglobulin and was vaccinated for hepatitis B. There was no history of melena, hematemesis or altered state of consciousness. On examination patient was mildly pale and had splenomegaly, 3cm below the costal margin. There were no other stigmata of chronic liver disease. Patient was advised to follow up with her blood complete picture, liver function tests (LFTs) and ultrasound abdomen to know the extent of liver damage secondary to hepatitis B. Her blood CP revealed anemia with Hb of 10.3 (microcytic hypochromic) and platelet count of 56,000. Her LFTs revealed ALT of 46. Her ultrasound abdomen showed normal liver echo texture with cavernous transformation of portal vein and splenomegaly with dilated splenic vein. Doppler ultrasound revealed amalgamation of numerous channels showing low velocity flow around porta hepatitis with possibility of portal vein recanalization after an episode of previous thrombosis.

Patient was worked up for hypercoagulable state to know the cause of portal vein thrombosis though there was no previous history of DVT, arthritis, oral ulcers or photosensitivity. She was found to have positive anti-cardiolipin antibodies and lupus anticoagulant. She was diagnosed as having thrombophilia secondary to Anti Phospholipid Syndrome. Patient was started propranolol and rivroxaban to decrease portal pressure and to prevent further episodes of spontaneous thrombosis. She was then referred to surgeon for consideration of shunt surgery. The surgeon advised to follow up with repeat endoscopy after 6 months and surgery was delayed till there is significant variceal bleed. Patient was advised to follow up after 6 months to see if she has spontaneous resolution of hepatitis B.

Discussion

Cavernous transformation is a rare phenomenon seen in patients who develop portal vein thrombosis.
This portal vein thrombosis can be idiopathic or secondary to malignancies, hematological disorders, cirrhosis or thrombophilic states which in our case was Anti Phospholipid Syndrome. After the thrombosis the liver loses two third of its blood supply. In order to compensate for this blood loss many small collateral blood vessels open both in and around the biliary channels to carry blood to the liver. However this compensatory mechanism is still insufficient to prevent the development of portal hypertension. If the thrombus extends into the mesenteric vein it may cause intestinal ischemia. The patients may be asymptomatic or present with a number of symptoms. There may be complaint of melena or haematemesis due to portal hypertension. There may be increasing pallor, gum bleed, easy bruising or fever as manifestation of hypersplenism. On examination patient may have caput medusa or splenomegaly. There may be pallor or bruises or jaundice due to biliary obstruction.

The diagnosis is made on ultrasound or color Doppler ultrasound examination that shows cavernous transformation with enlarged spleen. Patients in long standing cases may have small liver due to decrease in blood supply and slight impairment of liver function tests as well. Patient may also present as case of cholestasis or obstructive jaundice due to compression or (in long standing case) stricture of biliary channels. Further tests include CT angiography or MR angiography.

Our case presented in OPD for the management of Hepatitis B. Her ultrasound was done to see the liver status which revealed cavernous transformation. Cavernous transformation has very limited treatment options. It also depends on the cause of thrombosis. The literature lacks data on anticoagulation duration and outcomes. Patient may be considered for surgery (thrombectomy, venous reconstruction or vein graft), radiological endovascular intervention or leinorenal shunt at the cost of development of encephalopathy.

References