Antiphospholipid syndrome as a cause of nodular regenerative hyperplasia presenting as refractory ascites

FNU Bhawna, MD[1]; Justin J. Wagner, DO[1]; Vikash Kumar, MD[1]; Meron Debesai,MD[1]; Michael Bernstein MD[1]

[1]New York City Health + Hospitals, Coney Island

ACG 2022

INTRODUCTION

Nodular regenerative hyperplasia (NRH) and obliterative portal venopathy (OPV) are two causes of non-cirrhotic portal hypertension (NCPH), which is a vascular liver disease in which clinical signs of portal hypertension (PHT), such as esophageal varices, ascites, and splenomegaly develop in the absence of cirrhosis and portal vein thrombosis. The etiology often remains unidentified, but herein we present a case of a male with antiphospholipid syndrome who developed NRH and OPV.

CASE DESCRIPTION

We present a case of 56-year-old-male with NCPH and refractory ascites who underwent liver biopsy confirming NRH and OPV. Etiological work-up revealed beta-2 glycoprotein-1 and anticardiolipin antibodies, concerning for APS despite no prior history of thrombosis. The patient underwent a transjugular intrahepatic portosystemic shunt (TIPS) procedure for his refractory ascites and was started on prophylactic anticoagulation due to concern for APS with clinical improvement in his ascites and shortness of breath.

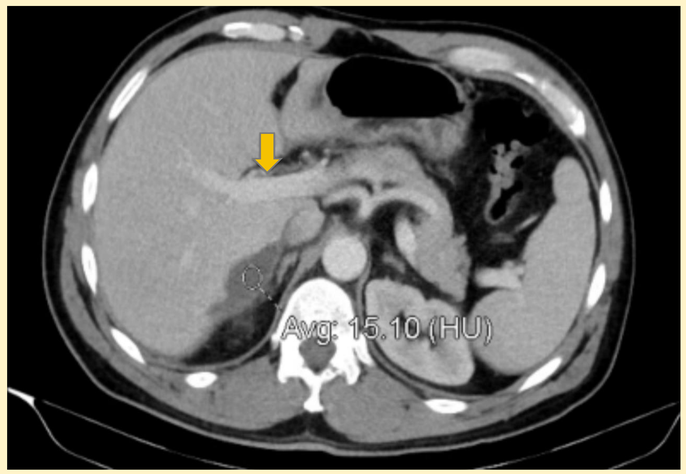

Fig.1: CT Abdomen showing patent portal vein (yellow arrow)

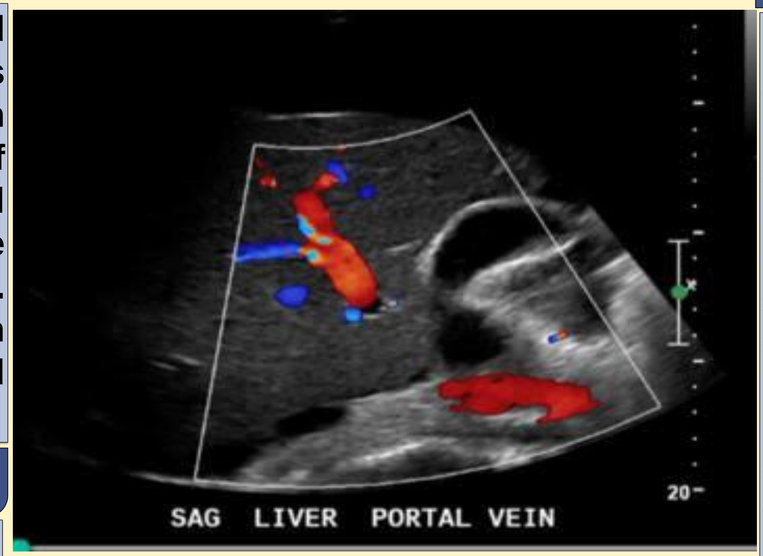

Fig. 2: Ultrasound doppler showing patent portal vein

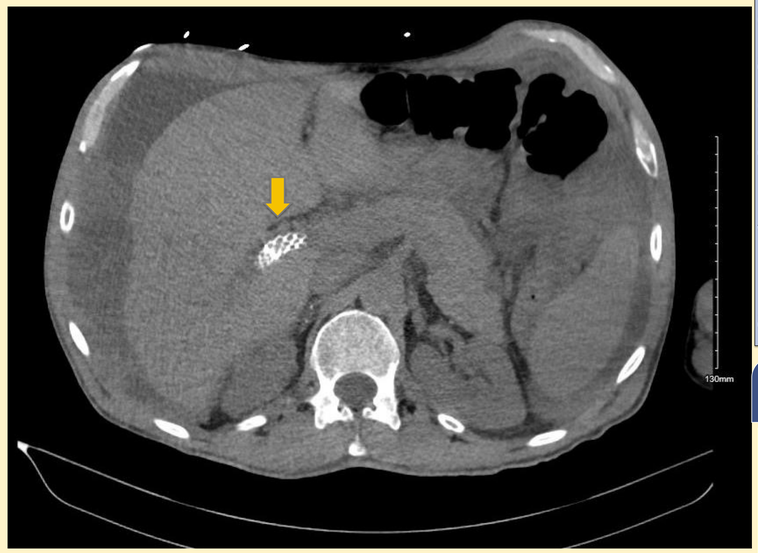

Fig. 3: CT Abdomen showing successful placement of TIPS (yellow arrow)

DISCUSSION

NCPH is a rare disease that typically presents with complications of PHT, such as ascites or variceal bleeding, and is commonly misdiagnosed as cirrhosis. The pathophysiology is unknown but both intrahepatic vascular obstruction and increased splanchnic blood flow have been suggested to explain NCPH. At minimum, the diagnosis of NCPH requires the presence of portal hypertension, the absence of cirrhosis, the presence of advanced fibrosis or other causes of chronic liver diseases, and the absence of thrombosis of the hepatic or portal veins on imaging. NCPH is a diagnosis of exclusion and an extensive work-up is typically recommended to first evaluate for other causes of liver disease, such as alcoholic and nonalcoholic steatohepatitis, autoimmune hepatitis, and viral hepatitis. For confirmation, liver biopsy findings can be associated with NRH and OPV, which are two causes of NCPH. Different conditions have been associated with NRH and OPV, including immunological disorders such as Lupus or APS, viral infections such as HIV, and immunosuppressive medications such as azathioprine.

CONCLUSION

TIPS is proven to be beneficial for symptomatic relief in refractory ascites. But definitive treatment in the setting of presence of autoimmune disease markers and role of immunosuppressants as well as anti-coagulation need more research data and investigations to reach on definitive treatment modalities.