NYC HEALTH+ HOSPITALS

Collapsing Alcoholic Cirrhosis Presenting as Functional Budd Chiari Syndrome.

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INTRODUCTION

Budd-Chiari syndrome (BCS) is a rare disease described as hepatic venous outflow obstruction involving the hepatic veins or inferior vena cava (IVC), devoid of cardiac origin. It is diagnosed in 1 per 2.5 million people in the general population without any predilection to gender. Majority of cases are associated with hypercoagulable state including, myeloproliferative disorders, malignancy and pregnancy or oral contraceptive pills. Clinical presentation depends on the acuity and severity of the case, however, commonly seen is acute liver failure, refractory ascites, hepatomegaly and abdominal pain. Diagnosis is made using ultrasonogram (US) with doppler (primarily) and triphasic CT scan or Magnetic Resonance Imaging (MRI) can be used as confirmatory tests. The complications are severe, so prompt and effective treatment is necessary with anti-coagulation and thrombolysis, with consideration of Transjugular Intrahepatic Portosystemic Shunt (TIPS) as an adjunct to treat associated portal hypertension. We present a case of advanced alcoholic cirrhosis that on initial imaging with US showed signs suspicious for flow obstruction but BCS was ruled out using triphasic CT, highlighting the value of using confirmatory tests and initiating appropriate intervention to minimize complications associated with anticoagulaton therapy in cirrhotic patients.

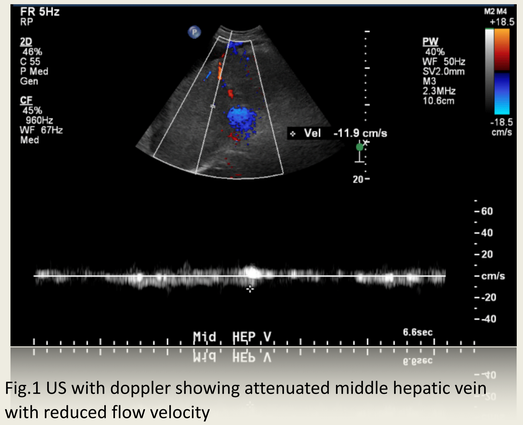

Fig.1 US with doppler showing attenuated middle hepatic vein with reduced flow velocity

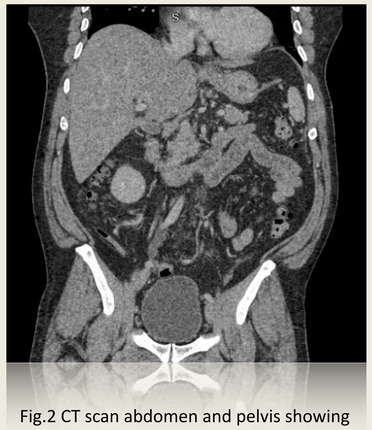

Fig.2 CT scan abdomen and pelvis showing hepatomegaly nd nodular liver contour suggestive of cirrhosis.

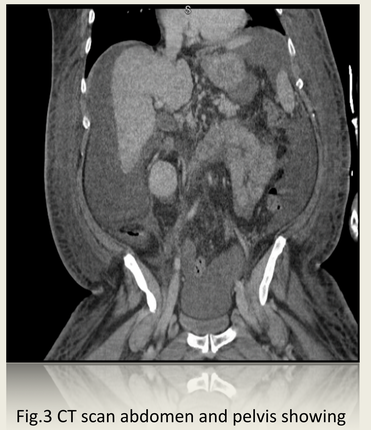

Fig.3 CT scan abdomen and pelvis showing Significantly shrunken liver with modularity And ascites 1 year after image in Fig.2.

CASE DESCRIPTION

A 49 year- old male with history of alcohol use disorder, alcoholic liver cirrhosis (Child-Pugh class C) with MELD- Na of 20 and grade I esophageal varices presented to the emergency department with worsening bilateral lower extremity edema with associated pain, right and left upper quadrant pain, shortness of breath and abdominal distention. Vital signs significant for hypertension at 168/ 86 on arrival but were otherwise within normal limits. Physical examination was significant for scleral icterus, jaundice, abdominal distention with positive fluid wave, 3+ bilateral lower extremity edema to the groin and tenderness.
Serum studies showed normocytic anemia hemoglobin of 10.2, thrombocytopenia with platelet count of 83, hypokalemia 3.3, ALK 268, AST 44, ALT 23, Direct bilirubin 1.4 and ammonia 48. The patient was admitted for decompensated liver cirrhosis. Antibiotic prophylaxis for spontaneous bacterial peritonitis (SBP) was initiated, as was diuresis with furosemide and aldactone and paracentesis was performed removing 5L of serous fluid. Fluid analysis showed SAAG score > 1.1 and Neutrophil count < 250, indicating portal hypertension. Ultrasound with doppler flow of the abdomen was obtained, showing hepatic veins with attenuated caliber and discontinuity between hepatic veins and IVC and low velocity bidirectional flow of the IVC. Confirmation study with Tri-phasic CT of the abdomen showed a shrunken liver, negative for clots in the hepatic veins or IVC and lacked compensatory hypertrophy of the hepatic sinusoids, ruling primary Budd chiari syndrome out.
Patient was assessed for intervention for refractory ascites and was not deemed to be a suitable candidate for TIPS given MELD-na of 20, severely shrunken hepatic architecture and lack of transplant availability should the procedure be unsuccessful. Patient was discharged on medical management and follows in hepatology clinic.

DISCUSSION

This case of alcoholic cirrhosis shares clinical and physiologic similarities with BCS. Differentiating between the two is key to initiating correct intervention and minimizing complications. This patient has atrophied hepatic architecture that serves as an outflow obstruction similar to that seen in BCS. Imaging with doppler US, CT or MRI looks for direct and indirect signs of BCS; here, no diagnostic signs were observed, despite a defect of venous outflow on doppler US. Had confirmatory tests not ruled out BCS, AC likely would have been initiated, exposing him to unnecessary risks. This highlights a patient with advanced alcoholic cirrhosis presenting with signs and symptoms suspicious for BCS. Initial imaging was also concerning for BCS, but confirmatory tests proved otherwise. This illustrates the value of confirmatory triphasic CT scan in a patient with US findings that are suspicious for, but not confirmatory of, BCS, guiding treatment away from AC and its risks and complications.

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