A Rare Case of Budd Chari Syndrome

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Introduction

Budd Chiari syndrome is a rare disorder resulting from hepatic venous outflow tract obstruction anywhere from the small hepatic veins to the suprahepatic inferior vena cava. The obstruction can be due to various causes, but all of which results in either reduction or obstruction of hepatic venous outflow. BCS can be further classified as primary BCS, where the obstruction is due to venous disease, such as thrombosis or phlebitis or secondary BCS where the compressions or invasions by a lesion originating outside the veins. We are presenting a patient with a primary Budd Chiari Syndrome.

Case Presentation

A 21 year old woman with tobacco abuse, history of two miscarriages presented with flu like illness six week prior followed by two weeks history of abdominal pain and abdominal swelling. Patient during the two week period complained of early satiety followed by right upper quadrant abdominal pain. The pain was described as dull, non radiating and constant 5-8/10 in intensity. The pain was aggravated by movement and alleviated somewhat by pain medication. The patient was initially evaluated at another hospital and was found to have ascites. Work-up for her ascites including two paracentesis, and imaging studies failed to reveal a cause for her ascites thus the patient was transferred to our tertiary facility.

On physical exam, patient had pale conjunctiva, no icterus or lymphadenopathy. Abdominal exam revealed distended abdomen with nonmoveactive bowel sounds, positive fluid wave and shifting dullness, increased liver span and no guarding or rigiditiy or rebound tenderness. Patient also had bilateral pedal edema without any cyanosis, clubbing. Genitourinary and pelvic exams were unremarkable.

On laboratory analysis, patient was leucopenia with WBC of 3.3x10^9/L, anemia with Hemoglobin of 11.2 and hematocrit of 34 with an MCV of 80 and RDW of 18%. Patient also had total bilirubin of 2.1 mg/dL and PT of 20.8 sec, INR of 1.7, and PTT of 68.9 sec. Evaluating for the cause of ascites numerous other labs were draw: negative acute viral hepatitis, negative HIV testing, Negative EBV titers, CMV IgM and IgG, Negative pregnancy test, ANA, SS-A, Anti-smooth muscle antibody, Anti-Mitochondrial Ab, SBE/UEP, Negative Gonorhea and Chlamydia screening. A CT scan of abdomen and pelvis was done, and it revealed hepatomegaly, ascites, abnormal appearance of intra-hepatic veins, and intra-hepatic portion of IVC. A liver biopsy and hepatic venogram was done and it showed a clot in the right hepatic vein with spider web collaterals and edematous mass effect on the hepatic veins. Patient was stared on anticoagulation with IV heparin therapy. Hypercoagulable work-up revealed low protein C of 37 and borderline low anti- cardiolipin, and prothrombin gene G20210A mutation, Factor V Leiden. Work up revealed low protein C of 37 and borderline low anti-cardiolipin, and prothrombin gene G20210A mutation, Factor V Leiden. Lupus anticoagulant was sent home with Hematology and Hepatology and was sent home with Hematology and Hepatology and Hepatology. She was started on long term anticoagulation with Coumadin and was seen in Budd Chiari Syndrome. Patient’s abdominal pain and distention improved greatly during this hospital course. She was started on long term anticoagulation with Coumadin and was seen in Budd Chiari Syndrome.

Discussion

Budd Chiari Syndrome (BCS) refers to the pathological resulting from obstruction or reduction in normal flow of blood out of the liver. It is a rare condition with incidence of 1 in 2.5 million persons per year. Underlying cause of BCS includes myeloproliferative disorder, malignancy, infections and benign lesion of the liver, oral contraceptives, pregnancy, post-partum state and hypercoagulable states. The obstruction of hepatic outflow tracts seen in BCS leads to increase in sinusoidal pressure and portal hypertension. The venous stasis leads to hepatic congestion which decreases hepatic perfusion. This can cause ischemic injury to the hepatocytes leading to hepatic necrosis, which eventually leads to hepatic fibrosis and cirrhosis. Patient can present at any stage of this continuum, thus clinical presentation of BCS varies depending on the acuteness of presentation and degree of obstruction. These patient can present with acute, even fulminant hepatic failure, subacute or chronic hepatic failure. BCS present mainly in the third and fourth decade and more commonly in women than men. Clinical presentation of BCS can vary from absence of symptoms to fulminant hepatic failure. Up to 20% of the patient may not have any symptoms, while very low <1.5% of all acute liver failure is accounted for by BCS. The two most commonly associated symptoms of BCS are ascites and abdominal pain. Other classical signs and symptoms include hepatomegaly, fever, abdominal pain, lower extremity edema, gastrointestinal bleeding, and hepatic encephalopathy.

Diagnosis of BCS can be established noninvasively using Doppler ultrasonography, Computed Tomography (CT) scan or Magnetic Resonance Imaging. The most prominent feature noted on noninvasive imaging is hepatomegaly and caudate lobe hypertrophy. Although, Doppler US, CT, MRI are commonly used to diagnose BCS, the gold standard is venography. Liver biopsy can help diagnose BCS, though both procedures can and it also can help decide upon treatment options. The treatment for BCS includes medical, radiological and surgical treatments. Medical treatments include anticoagulation to prevent recurrent thrombosis and extension of existing thrombosis. Local thrombolysis can also be used for acute and subacute BCS of less than four weeks duration. Ascites can be treated with low sodium diet, and diuretics. Treating underlying causes whenever possible is also essential to relieve symptoms. Radiological and surgical interventions include angioplasty, Transjugular intrahepatic portosystemic shunting (TIPS), Shunting, Shunting procedures and liver transplant. The treatment is individualized to the particular patient. The treatment is individualized to the particular patient.

Discussion Con’t.

Independent variables predicting a worse prognosis include the presence of encephalopathy, ascites, an International Normalized ratio of ≥2.3, older age at diagnosis, higher bilirubin concentration, more severe liver failure, and presence of right upper quadrant pain. Overall 1 year survival was 90% 6 months, 82% at 1 year, 82% at 2 years and 80 % at 5 years. The main cause of death for patients with BCS are liver failure, multiforgan failure, and gastrointestinal bleeding.

Conclusions

Budd Chiari syndrome is a rare disorder resulting from hepatic venous outflow tract obstruction anywhere from the small hepatic veins to the suprahepatic inferior vena cava. Underlying cause of BCS includes myeloproliferative disorder, malignancy, infections and benign lesion of the liver, oral contraceptives, pregnancy, post-partum state and hypercoagulable states. Clinical presentation of BCS can vary from absence of symptoms to fulminant hepatic failure. The two most commonly associated symptoms of BCS are ascites and abdominal pain. Diagnosis of BCS can be established noninvasively using Doppler ultrasonography, Computed Tomography (CT) scan or Magnetic Resonance (MR) imaging. The gold standard is venography. Liver biopsy can help diagnose BCS in acute or subacute cases and it can also help decide upon treatment options. The treatment for BCS includes medical, radiological and surgical treatments. The treatment is individualized to the particular patient.

Reference