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Budd Chiari Syndrome as a Manifestation of Systemic Lupus Erythematosus in 20-Year-Old Female - A Case Report

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Abstract: ***Budd-Chiari syndrome is defined as the obstruction of hepatic veins or terminal inferior vena cava[1]. There are many possible causes of Budd-Chiari syndrome. Only a few cases of Budd-Chiari syndrome as a manifestation of Systemic Lupus Erythematosus have been reported. This is a case of Budd-Chiari Syndrome a Cause of SLE.***

Keywords: budd chiari syndrome, Systemic lupus Erythematosus, APLA

1. Introduction

Budd-Chiari syndrome is defined as obstruction of hepatic veins or terminal IVC. Aetiology of Budd-Chiari syndrome includes myeloproliferative Disorder, Polycythemia rubra vera, membranous obstruction of IVC, filariasis, amoebic liver abscess, aspergillosis, schistosomiasis, Hepatocellular carcinoma, renal cell carcinoma, adrenal adenoma, leiomyosarcoma of IVC, and antiphospholipid syndrome[2, 3]

The spectrum of clinical presentation ranges from a completely asymptomatic disease to fulminant hepatic failure to chronic liver disease.4, 5 Major features include ascites, abdominal pain, and fever. All complications of chronic liver disease may occur, including GI bleeding, bacterial infections, hepatorenal syndrome, and encephalopathy. Systemic lupus erythematosus (SLE) is an autoimmune disorder being mediated by immunocomplexs and autoantibodies and INF-Alpha is the key mediator

2. Case Report

A 21 year old female patient was admitted with chief complaint of abdominal distention since last 1 month followed by bilateral pedal oedema since last two weeks. Patient also had mild fever with temperature of 100.4° Fahrenheit since last 4 days. Patient also had No history of recurrent miscarriages, rash over the face, seizures and body pain. No past history of previous surgery or prolonged medication. There was no history of menstrual irregularities and similar illness in the family members. On examination abdomen was distended with no hepatomegaly and spleen being not palpable. Shifting dullness was present with absence of fluid thrill. There was present of arthralgia in fingers of both hands since last 3 months, which was not associated with any deformity. Patient also had patchy non scarring alopecia on head since last 6 months.

On investigations –

Ascitic fluid analysis showed straw yellow fluid with lymphocytes, few polymorphs and no malignant cells. Ascitic fluid ADA was 10 u/l, glucose-132mg/dl, protein 1.6 g/dl albumin 0.7 g/dl, SAAG ratio was-1.4 and gram stain showed no bacteria or pus cells.

On USG of abdomen-Gross Ascitis, with coarse hepatic echotexture showing hepatic parenchymal disease and non visualisation of hepatic vein s/o-Budd-Chiari Syndrome.

CECT abdomen-showing Heterogeneous liver parenchyma, non-visualisation of portal vein, gross ascites s/o-Budd-Chiari Syndrome.

On ANA screening-It was found to be +ve with 4.4 units and Ds-Dna +ve with 38.69 IU/ml.

Lupus anticoagulant test was also done-Anticardiolipin and phospholipid antibody within normal range with

Lupus anticoagulant 1-44.6 seconds

Lupus anticoagulant 2-50.1 seconds.

3. Discussion

Budd-Chiari syndrome (BCS) is defined as the obstruction of hepatic veins or terminal inferior vena cava (IVC). Primary BCS arises from a venous anomaly, whereas secondary BCS arises from an initial lesion outside the veins. BCS is a rare disease. A territory corresponding to at least 2 major hepatic veins needs be obstructed before clinical manifestations of BCS develop.1, 4Increased hepatic venous and sinusoidal pressures translate into sinusoidal distension and congestion, predominantly in the centrilobular area, and cause ascites formation. Outflow tract obstruction reduces the low-pressure portal venous inflow. Due to stasis and an underlying prothrombotic condition, intra-and extrahepatic portal vein thrombosis is common. Decrease in hepatic blood flow causes ischemic coagulative necrosis and apoptosis, predominantly in the central parts of the lobules, leading to liver dysfunction or liver failure. Subsequently, the loss of hepatocytes results

in so-called parenchymal extinction, with replacement of liver cells with connective tissue.

The spectrum of clinical presentation ranges from a completely asymptomatic disease to fulminant hepatic failure to chronic liver disease[4, 5] Major features include ascites, abdominal pain, and fever. All complications of chronic liver disease may occur, including GI bleeding, bacterial infections, hepatorenal syndrome, and encephalopathy.4, 5

Antiphospholipid syndrome is characterized by the production of auto antibodies directed against phospholipids and is associated with multiple thrombotic events[6]. Thrombosis following antiphospholipid syndrome can occur anywhere or in any organs[7].

There are rare reports of association of antiphospholipid antibody syndrome (APLA) with Budd-Chiari syndrome. Most of the cases reported had a clinical manifestation of SLE before they were diagnosed with Budd-Chiari syndrome. In our case there is clinical manifestation of systemic lupus erythematosus before developing Budd-Chiari syndrome

Side to side porto-caval shunt is the most effective therapy for Budd-Chiari syndrome. Ortho topic liver transplantation is indicated only when there is a failure of porto-systemic shunt. This is because of the thrombosis of the portal vein, splenic vein, and superior mesenteric vein which results in un-shuntable portal hypertension [8]. In case of a thrombotic complication, patient should be on long tern anticoagulants like warfar in 20mg/day and INR should be kept at a level of 3 to 4 [9].

4. Conclusion

Budd-Chiari syndrome can be an initial manifestation of systemic lupus erythematosus, when a young female patient presents with features of Budd-Chiari syndrome always screen for antiphospholipid antibody syndrome even if the other clinical manifestations of SLE are absent.

Conflict of Interest

None

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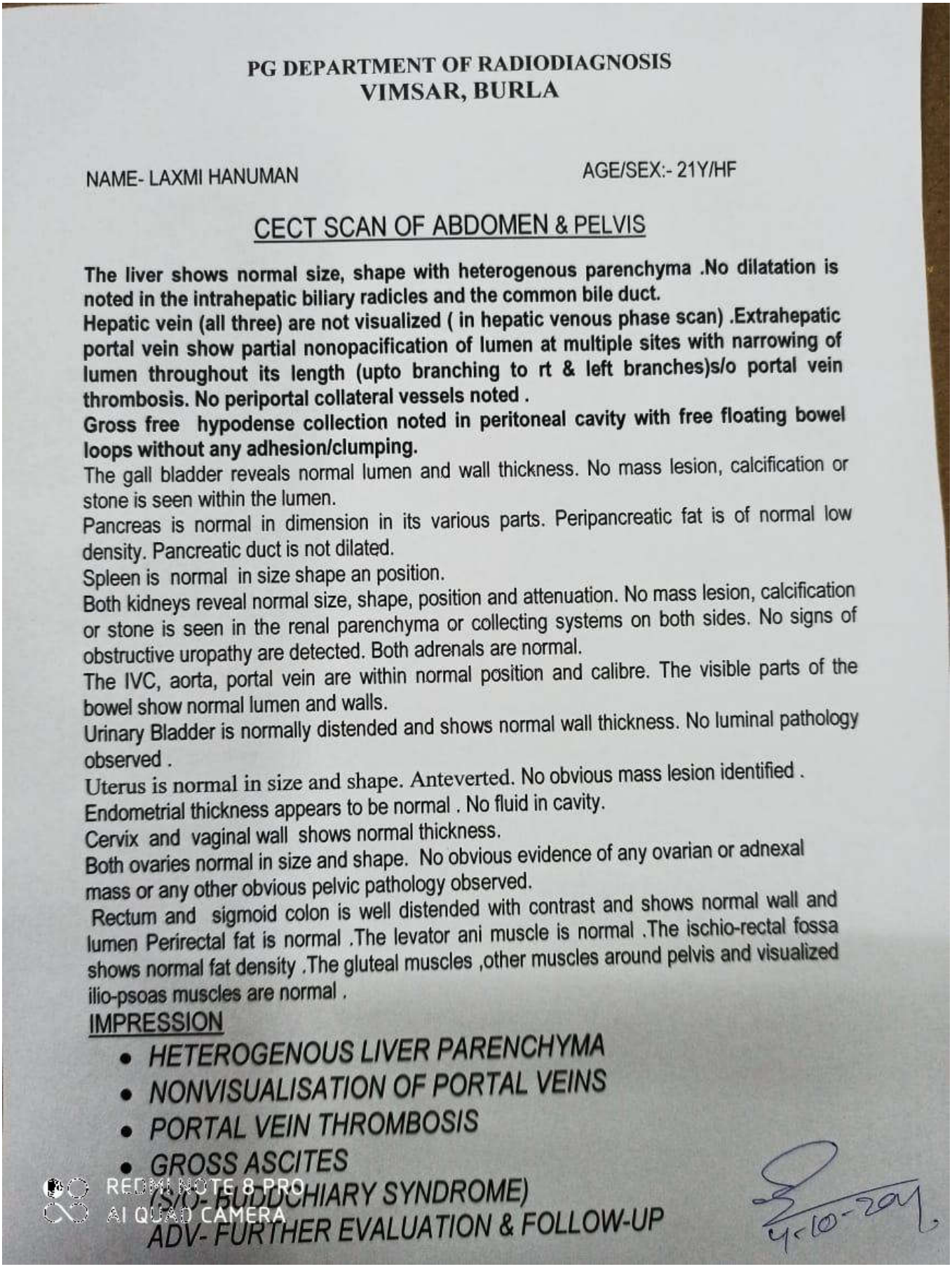

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CECT SCAN OF ABDOMEN & PELVIS

The liver shows normal size, shape with heterogenous parenchyma .No dilatation is noted in the intrahepatic biliary radicles and the common bile duct.
Hepatic vein (all three) are not visualized (in hepatic venous phase scan) .Extrahepatic portal vein show partial nonopacification of lumen at multiple sites with narrowing of lumen throughout its length (upto branching to rt & left branches)s/o portal vein thrombosis. No periportal collateral vessels noted .
Gross free hypodense collection noted in peritoneal cavity with free floating bowel loops without any adhesion/clumping.
The gall bladder reveals normal lumen and wall thickness. No mass lesion, calcification or stone is seen within the lumen.
Pancreas is normal in dimension in its various parts. Peripancreatic fat is of normal low density. Pancreatic duct is not dilated.
Spleen is normal in size shape an position.
Both kidneys reveal normal size, shape, position and attenuation. No mass lesion, calcification or stone is seen in the renal parenchyma or collecting systems on both sides. No signs of obstructive uropathy are detected. Both adrenals are normal.
The IVC, aorta, portal vein are within normal position and calibre. The visible parts of the bowel show normal lumen and walls.
Urinary Bladder is normally distended and shows normal wall thickness. No luminal pathology observed .
Uterus is normal in size and shape. Anteverted. No obvious mass lesion identified .
Endometrial thickness appears to be normal . No fluid in cavity.
Cervix and vaginal wall shows normal thickness.
Both ovaries normal in size and shape. No obvious evidence of any ovarian or adnexal mass or any other obvious pelvic pathology observed.
Rectum and sigmoid colon is well distended with contrast and shows normal wall and lumen Perirectal fat is normal .The levator ani muscle is normal .The ischio-rectal fossa shows normal fat density .The gluteal muscles ,other muscles around pelvis and visualized ilio-psoas muscles are normal .

IMPRESSION

- *HETEROGENOUS LIVER PARENCHYMA*
- *NONVISUALISATION OF PORTAL VEINS*
- *PORTAL VEIN THROMBOSIS*
- *GROSS ASCITES*

(S/O- BUDDCHIARY SYNDROME)
ADV- FURTHER EVALUATION & FOLLOW-UP

4-10-2021