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Case Report

Internal Medicine Section

Budd- Chiari Syndrome as an Initial Manifestation of Systemic Lupus Erythematosus

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ABSTRACT

Budd- Chiari syndrome is caused by obstruction of hepatic venous outflow. There are numerous causes for Budd-Chiari syndrome. One of the causes is systemic lupus erythematosus due to antiphospholipid antibodies. Only few cases have reported Budd-Chiari syndrome as an initial manifestation of systemic lupus erythematosus (SLE). This is a case report of Budd-Chiari syndrome due to SLE.

Keywords: Antiphospholipid antibodies, Hepatic vein, Inferior vena cava (IVC)

CASE REPORT

A 24-year-old female was admitted with complaints of breathlessness and abdominal distension for 4 months. Patient also complained of fever for 4 days. There was a history of facial puffiness and pedal oedema for 4 days. 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 there was bilateral pitting pedal oedema. Inspection of the abdomen showed distension with dilated veins over the anterior abdominal wall. Palpation of the abdomen showed splenomegaly with free fluid [Table/Fig-1].

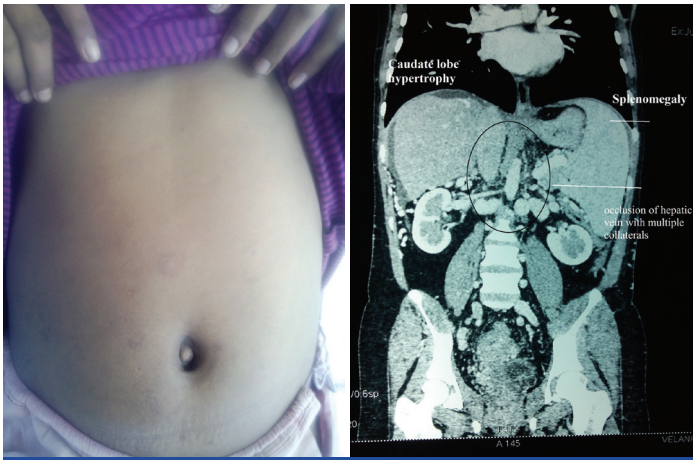
Investigations showed normal haemogram and renal function test. Liver function test showed total bilirubin -1.08mg/dl, AST- 68U/L. ALT-33U/L. Serum viral marker was negative. Coagulation profile was normal. Ultrasound abdomen showed splenomegaly with free fluid. Contrast CT showed occlusion of the right hepatic vein and the distal middle and left hepatic veins with multiple intra hepatic collaterals [Table/Fig-2]. Compressed intra hepatic segments of IVC with multiple retroperitoneal and abdominal wall collaterals. There was an evidence of free fluid with splenomegaly [Table/Fig-3]. Upper gastro intestinal endoscopy showed grade I varices. Patient was screened for Antiphospholipid Antibodies, Antinuclear Antibody, Antinuclear Antibody Anti-Sm and Anti-dsDNA antibody and was found to be positive. Based on the above investigations patient was diagnosed as anti phospholipid antibody syndrome due to systemic lupus erythematosus. Patient was initially treated with low molecular weight heparin, followed by oral anticoagulant warfarin. Later patient was prepared for thrombolysis with urokinase. Thrombolysis failed to improve the condition. So patient underwent Transjugular intrahepatic portosystemic shunt (TIPS). Patient followed up for one year. There was significant improvement with warfarin and steroids.

DISCUSSION

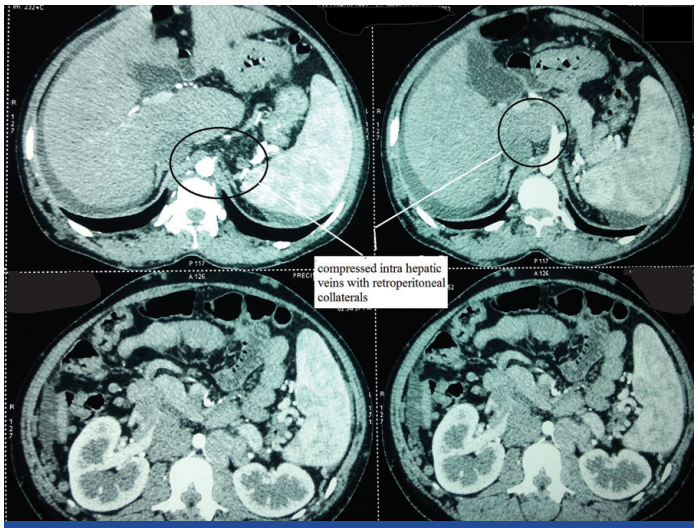
Budd-Chiari syndrome is caused by the obstruction of hepatic venous outflow which produces intense congestion of the liver. In Budd-Chiari syndrome there is thrombosis of all three major hepatic veins. The smaller hepatic veins that are usually draining the caudate lobe are often spared [1]. Aetiology of Budd- Chiari syndrome includes 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].

Clinical features of Budd- Chiari syndrome includes abdominal pain, abdominal distension, weakness, anorexia, jaundice, massive ascites, hepatomegaly, splenomegaly, abdominal venous distension and oedema of thighs, legs and feet [4]. Antiphospholipid syndrome is characterised by the production of auto antibodies directed against phospholipids and is associated with multiple thrombotic events [5].

Thrombosis following antiphospholipid syndrome can occur anywhere or in any organs [6]. These thrombosis causes gangrene of the limbs, pulmonary embolism, myocardial infarction, renal



[Table/Fig-1]: Inspection of abdomen showed distension with dilated vein over anterior abdominal wall. Palpation of abdomen showed splenomegaly with free fluid.
[Table/Fig-2]: Contrast CT showed occlusion of the right hepatic vein and the distal middle and left hepatic veins with multiple intra hepatic collaterals.



[Table/Fig-3]: Compressed intra hepatic segments of IVC with multiple retroperitoneal and abdominal wall collaterals with splenomegaly

artery thrombosis, avascular necrosis of bone and Budd-Chiari syndrome. Most important feature of Budd-Chiari syndrome on contrast CT is hypertrophy of caudate lobe, ascites, and splenomegaly [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. All those reported cases initially had recurrent fetal loss with portal vein thrombosis [8], mesenteric thrombosis [9], ileofemoral thrombosis [10] followed by Budd-Chiari syndrome. But in our case there is no clinical manifestation of systemic lupus erythematosus before developing Budd-Chiari syndrome. Similar findings was reported by Espinosa G et al., and Ilkgül O et al., showed Budd-Chiari syndrome as an initial manifestation [11,12].

Budd-Chiari syndrome as a manifestation of secondary antiphospholipid syndrome in SLE is rare. Patient with secondary antiphospholipid syndrome should be on long term antiplatelet drugs to prevent arterial occlusion and on low dose aspirin to prevent venous thrombosis. In an established case of Budd-Chiari syndrome the thrombolysis of the hepatic vein is largely ineffective because the window for effective clot lysis is only 2-3 weeks and most of the patients present only after that [13].

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 unshuntable portal hypertension [14]. In case of a thrombotic complication, patient should be on long tern anticoagulants like warfarin 20mg/day and INR should be kept at a level of 3 to 4 [15].

CONCLUSION

Budd-Chiari syndrome can be an initial manifestation of systemic lupus erythematosus, even before the appearance of other manifestations. 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. Before diagnosing idiopathy as a cause for Budd-Chiari syndrome screen for antiphospholipid antibody syndrome in young females because prior anticoagulation therapy and steroids will avoid future miscarriage.

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