

Management of Pregnancy in a Rare Case of Budd–Chiari Syndrome: A Case Report

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ABSTRACT

Budd–Chiari syndrome is a rare liver disorder in which there is obstruction of hepatic veins causing liver cirrhosis and fulminant hepatic failure. It is a non-specific liver disorder of pregnancy, if it occurs in pregnancy, it should be managed by a multidisciplinary team approach involving an obstetrician, gastroenterologist, and interventional radiologist. We present a case of Budd–Chiari syndrome managed by right hepatic vein angioplasty and stenting, was on oral anticoagulants and became pregnant after that, and was managed in our hospital and had successful outcome of pregnancy.

Keywords: Case study, Pregnancy, Upper gastrointestinal symptoms.

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INTRODUCTION

Budd–Chiari syndrome is an uncommon illness characterized by obstruction of the hepatic venous outflow pathway. Between the tiny hepatic veins and the supra-hepatic inferior vena cava, an obstruction might occur.

Pregnancy with Budd–Chiari syndrome is rare, and its management is entirely different from normal pregnancy according to the status of the liver and its pathology during pregnancy. We present a case of successful management of pregnancy in a known treated case of Budd–Chiari syndrome.

CASE REPORT

A 21-years-old female, second gravida, with previous history of one abortion, with a history of 6 months of amenorrhea, LMP – 18/3/2021 and EDD – 25/12/2021, had been referred to us for antenatal management in September 2021. She was a known case of chronic liver disease and diagnosed as a case of Budd–Chiari syndrome in December 2020. Initially, the patient had a history of hematemesis for which basic ultrasonography has been done, which showed altered echotexture and hypertrophy of liver and splenomegaly, upper GI endoscopy showed grade 1 oesophageal varices, liver fibro scan – 10.2 KPA suggestive of significant fibrosis, AFP and CA 19-9 were normal, which rules out hepatocellular carcinoma, antinuclear antibody, antimitochondrial antibody, and anti-smooth-muscle antibodies are negative, and the probability of autoimmune hepatitis was ruled out. Followed by triphasic, contrast enhanced computed tomography (CECT) had been done, which showed chronic parenchymal liver disease with thrombosis in the right and middle hepatic vein for which right hepatic vein stenting was done in January 2021 and was on oral, anticoagulant therapy.

During her regular follow-up visit, she was found to be pregnant and was referred to us for further management of pregnancy.

Routine antenatal investigations like complete blood count, liver function test, renal function test, viral markers, urine routine and microscopy, and baseline antenatal sonography were done, which showed single live intrauterine pregnancy corresponding to

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24 weeks of gestation. The patient was advised to stop T. warfarin and started on inj. Clexane 0.4 mg once a day subcutaneously. A liver function test and coagulation profile monitoring were done once in 15 days.

Throughout her antenatal period, liver function test and coagulation profile were within normal limits. At 36 weeks of pregnancy, she was admitted and liver Doppler was done, which showed infra-, intra-, and supra-hepatic IVC normal and no evidence of thrombus, portal vein Doppler was normal, and liver showed chronic parenchymal disease. The stent in the right hepatic vein showed normal color flow, left hepatic vein was normal.

She had labor pains. The morning dose of clexane was withheld. She delivered vaginally, the birth weight of the baby was 2.7 kg, and the postnatal period was uneventful. The next day, the patient started on injection clexane 0.4 mg and tablet acitrom 4 mg, liver function test (LFT) and coagulation profile monitoring was done for warfarin bridging, and discharged on postnatal day 5.

DISCUSSION

Budd–Chiari syndrome is a rare condition defined as thrombotic or nonthrombotic obstruction of the hepatic venous outflow tract. The obstruction may be located from small hepatic venules up to the entrance of IVC into the right atrium.

Hepatic outflow tract obstruction related to cardiac disease, pericardial disease, and sinusoidal obstruction syndrome is excluded from this definition. It is a rare condition. Prevalence is 1 in 100000–500000.¹

There are 3 types. In type I – there is IVC occlusion with or without occlusion of hepatic veins. In type II – there is occlusion of major hepatic veins and in type III – occlusion of small centrilobular veins. It is also classified as primary or secondary BCS. Primary Budd–Chiari syndrome is defined as an obstruction that is predominantly due to thrombosis of hepatic veins. In contrast, secondary Budd–Chiari syndrome is defined as compression or invasion of the hepatic veins and/or the inferior vena cava by a lesion that originates outside of the vein (for example, a malignancy).²

Budd–Chiari syndrome should be suspected in patients with ascites, hepatomegaly, and presence of upper abdominal pain simultaneously, patients with chronic liver disease with intractable ascites and mildly altered LFT, and in case of idiopathic chronic liver disease.

Budd–Chiari syndrome is caused by thrombus formation in the hepatic veins, which blocks the hepatic outflow, resulting in hepatic venous congestion, increased sinusoidal pressure, cellular hypoxia, and eventually liver cirrhosis and portal hypertension. In acute cases, this can lead to liver failure, while in chronic cases, it can lead to ascites and hepatomegaly with preserved liver function. Later, fibrosis develops, leading to cirrhosis.³

The complications of portal hypertension include the formation of portosystemic shunts, which leads to oesophageal varices, caput medusae, and rectal hemorrhoids, ascites, and splenomegaly.

The causes of Budd–Chiari syndrome include inherited causes like factor V Leiden mutation, prothrombin gene mutation, protein-C and -S deficiency, and antithrombin-3 deficiency, acquired causes include antiphospholipid syndrome, myeloproliferative disorders, and oral contraceptives, and might also be idiopathic. An important nongenetic risk factor is the use of estrogen-containing (combined) forms of hormonal contraception. Antiphospholipid syndrome, Behcet's disease, aspergillosis, pregnancy, dacarbazine, and trauma are all risk factors.

Diagnosis is based on radiological evaluation. Ultrasonography and color-flow Doppler have a sensitivity and specificity of 85–90% in diagnosing BCS. Common findings of Budd–Chiari syndrome include the presence of inferior vena cava (IVC) webs and thrombi, decreased IVC diameter, presence of hepatic venous thrombosis, increased caudate lobe size, presence of ascites, presence of intrahepatic or extrahepatic collaterals, stenosis in the IVC, or hepatic veins.⁴ Triphasic CT scan and MRI are a non-invasive modality.

Liver fibro scan is a noninvasive modality that is used to measure the degree of fibrosis in the liver, the normal value is 2–7 kPa (kilo Pascal). If more than 10, it is suggestive of severe cirrhosis of the liver.

Markers like AFP, CA 19-9, antinuclear antibody, anti-smooth muscle antibody, and antimitochondrial antibodies should be done to know the cause of cirrhosis of the liver. In BCS – all these markers are normal and the cause of liver cirrhosis in BCS is due to hepatic cellular hypoxia and increased venous congestion. AFP and CA 19-9 are elevated in hepatocellular carcinoma, antinuclear antibody, anti-smooth-muscle antibody, and antimitochondrial antibodies are elevated in autoimmune hepatitis.

Management depends on size of the thrombus and type of presentation, acute or chronic. It may be surgical or nonsurgical. It includes systemic thrombolysis, catheter-directed thrombolysis, angioplasty and stenting, and TIPSS.

CONCLUSION

Pregnancy is a hypercoagulable state, and patients are more prone to develop Budd–Chiari syndrome in pregnancy, but it is not a contraindication to pregnancy. If diagnosed during pregnancy, management is based on size of thrombosis and the extent of liver injury. Serial LFT monitoring should be done, warfarin should be stopped and changed to low-molecular-weight heparin, and continued through the antenatal period till delivery, coagulation profile monitoring, and liver Doppler should be monitored.

If Budd–Chiari syndrome is treated promptly in pregnancy, the disease is well controlled, and the prognosis of pregnancy, maternal, and fetal outcomes is good and successful.

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