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Case Study

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Buddchiari syndrome associated miscarriage - A case report

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ABSTRACT

Budd-Chiari syndrome (BCS) is described as blockage of the outflow of hepatic veins. This obstruction could be detected anywhere between the small hepatic veins to the suprahepatic inferior vena cava (IVC). BCS can be classified as two groups: primary and secondary. BCS is observed as primary when blockage of the outflow of hepatic veins is the outcome of an endoluminal venous lesion which could be either thrombus or web and its observed as secondary when blockage of hepatic veins originates from a lesion outside the venous system which could be a tumor or an abscess or cysts.

A pregnant patient of 26 years was admitted in the OBG department of Aster CMI hospital, Hebbal, Bengaluru with complaints of increased bleeding and abdominal pain who was diagnosed with Buddchiari syndrome 2 years back. This is her 2nd pregnancy and she had a history of spontaneous miscarriage at 5 weeks period of gestation(POG). On the 3rd day of admission, abortus fetus was delivered followed by placental delivery blood transfusion was also done.

With a prevalence of 1 in 1 lakhs to 5 lakhs, primary BCS is a rare disorder, majorly detected by imaging, whereas options of therapy consists of a step-wise approach with medication (anticoagulation), minimally invasive procedures like thrombolysis, percutaneous angioplasty, transjugular intrahepatic portosystemic shunting (TIPS), and surgical portosystemic shunting and orthotopic liver transplantation (LTX). BCS is rare and clinically challenging disease.

Keywords: buddchiari syndrome, pregnancy, miscarriage, spontaneous abortion, antepartum haemorrhage

INTRODUCTION

Budd-Chiari syndrome (BCS) is described as blockage of the outflow of hepatic veins. This obstruction could be detected anywhere between the small hepatic veins to the suprahepatic inferior vena cava (IVC). BCS can be classified as two groups: primary and secondary. BCS is observed as primary when blockage of the outflow of hepatic veins is the outcome of an endoluminal venous lesion which could be either thrombus or web and its observed as secondary when blockage of hepatic veins originates from a lesion outside the venous system which could be a tumor or an abscess or cysts. Usually patients

exhibit a basic prothrombotic disorder, and in a considerable set of patients various risk factors would be present.

As the typical patient in the case report is a young pregnant woman presenting with bleeding. Eventhough, pregnancy in women with BCS is exceptionally rare; pregnancy influences the prognosis of the disease in a deleterious way. The idea of a step-wise therapy has been entrenched, including anticoagulation, thrombolysis, percutaneous recanalization, transjugular intrahepatic portosystemic shunt, surgery and liver transplantation. Besides, an advanced use of TIPS has been put forward in chosen cases due to comparatively high mortality from BCS in patients who has gone through medical treatment alone.

CASE REPORT

A pregnant patient of 26 years was admitted in the OBG department of Aster CMI hospital, Hebbal, Bengaluru with complaints of increased bleeding and abdominal pain who was diagnosed with Buddchiari syndrome 2 years back. She is a k/c/o hypothyroidism and is on T. THYRONORM 50MCG. She has been treated with T. ACITROM 2mg and increased to 3mg after conception. The patient was started with INJ. CLEXANE 40mg the next day after the bleeding was started. The patient was on T. FOLIC ACID as well. This is her 2nd pregnancy and she had a history of spontaneous miscarriage at 5 weeks POG.

The vitals at the time admission were recorded as pulse rate of 122bpm, respiratory rate of 22cpm, blood pressure of 100/60mm/Hg and temperature was afebrile.

The lab values on the day of admission were, Prothrombin time(24.3 sec), INR Test(2.03), APTT Test(42.5sec), Lymphocyte count(13.3%), Hb(11.8g/dL), Haematocrit(34.4%), RDW(24.6%), Platelet Count(120 K/uL). On the 2nd day of admission the lab values were, PT time(17.7 sec), INR(1.39), Hb(10.5g/dL). On the 3rd day of admission the lab values were Hb(8.7g/dL), Platelet Count(74 K/uL). On the day of discharge the lab values were of Hb(9.4g/dL), Platelet Count(86K/uL).

She was treated with T. DUVADILAN(40mg) for 4 days, I. SUPACEF(1.5g), I. PAUSE(500mg), I. KABIMOL(1g), C.SUSTEN (200mg), T. CEFTUM (500mg) and T. CYCLOPAM (20/500) for 3 days and T. PAN(40mg), I. METROGYL(100mg) for 1 day and T. METROGYL(400mg), I. JUSTIN (75mg), I. EMESET(2mg/mL), I.SUSTEN (200mg), JONAC SUPPOSITORY (100mg), T. THYRONORM (50mcg), I. TRAMAZAC (50mg), I. CARBOPROST (250mcg) for 1 day. Also she was discharged with medications like T. CEFTUM (500mg) BD for 5 days, T. LIVOGEN BD to continue for 6 months, I. CLEXANE S/C OD to continue for a week.

On the 3rd day of admission, abortus fetus was delivered followed by placental delivery blood transfusion was also done.

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DISCUSSION

With a prevalence of 1 in 1 lakhs to 5 lakhs, primary BCS is a rare disorder. This can be majorly detected by imaging, whereas options of therapy consists of a step-wise approach with medication (anticoagulation), minimally invasive procedures like thrombolysis, percutaneous angioplasty, transjugular intrahepatic portosystemic shunting (TIPS), and surgical portosystemic shunting and orthotopic liver transplantation (LTX). To rebuild hepatic venous drainage, and liver transplantation in unresponsive patients. BCS is rare and clinically challenging disease.

Etiology and clinical presentation diverge between Western and certain Asian countries. Myeloproliferative neoplasms are shown by 35%–50% of European patients and are generally analogous to JAK2 V617F mutation. Non-invasive imaging (Doppler ultrasound, computed tomography, or magnetic resonance imaging) normally help in the accurate diagnosis. Liver biopsy must be carried out if small vessel BCS is doubted. This strategy provides a 5-year survival rate of around 90%. Long term result is determined by any underlying haematological condition and development of hepatocellular carcinoma.

Furthermore, the pregnancy-induced hypercoagulability, several changes inherent to pregnancy adds up to its injurious effect on BCS. This can be characterized by blood volume expansion and hypoproteinemia; the rise in intraabdominal pressure; pressure of the gravid uterus on the IVC and other intraabdominal vessels, including the lymphatic system; and the displacement of intraabdominal organs by the expanding uterus, resulting in changes in respective anatomical relationship. The incidence of thrombosis, in spite of full-dose corrected anticoagulant therapy emphasizes the increased thrombotic risk of Essential thrombocythemia patients during gestation.

There are no complete evidence to show that detrimental prognosis of the disease could have been avoided if patient had abstained from pregnancy or vice versa. The long time stability of the patient when not pregnant and when is pregnant has a greater variation and of course the pregnancy doesn't go hand in hand with BCS as well. The coagulation effect and hepatic dysfunction could be the major factor contributed to the miscarriage.