Case Report

Budd chiari syndrome: A case report on a rare disorder

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ABSTRACT

Budd Chiari syndrome (BCS) is a rare disease that threatens life due to a hepatic vein blood flow obstruction. As per the literature, thrombosis is a significant factor for hepatic venous obstruction. It can treat BCS based on the severity of the patient's condition by three methods like hepatic vein angioplasty, interventional radiology stenting with direct intrahepatic portosystemic shunt, or liver transplantation. We describe the case of a 34-years-old female patient who presented with abdominal distension and generalized weakness with a history of BCS. A cooperative collaboration of hepatologists and interventional radiologists helps in effectively treating the syndrome.

Key words: Budd Chiari Syndrome, Hepatic venous outflow, Intrahepatic portocaval shunt, Thrombophilia

Budd Chiari syndrome (BCS) is a rare or uncommon disorder characterized by obstruction of the hepatic venous outflow which can be either thrombotic or non-thrombotic. It has a heterogeneous clinical disease that can be treatable or possibly fatal [1]. Initially, Budd reported the disease in 1845, and in 1899, Chiari expanded on it by presenting 13 cases. The majority of cases result from thrombosis within the hepatic veins [2]. BCS has predicted to affect one in every 2.5 million people in Western nations each year [3]. It has a high prevalence of one in the millions of the general population. BCS has known to affect all age groups, but it is more common in people between 30 and 40 years and mostly in women [4]. The blockage of the hepatic vena flow can be at any level irrespective of the cause of obstacle from the small hepatic veins to the inferior vena cava end. It develops into sinusoidal congestion, hypoxic injury to the parenchyma, centrilo-based fibrosis, and ultimately cirrhosis [3]. It is a serious, rare, life-threatening condition of the liver and it requires medical procedures immediately and aggressively [4].

CASE REPORT

A 34-years-old female patient presented to the tertiary care center with complaints of abdominal distension and generalized weakness for 1 month. Also, there was continuous pain and swelling in the upper abdomen (may be caused by excess fluid) along with swelling in the legs. The patient had her first pregnancy 10 years back at the age of 25 years. At that time, the patient was advised for further evaluation, after which, the doctor identified that she has a rare condition called BCS. Two hepatic veins out of three were blocked entirely, and there might be a chance for blocking the third vein, so they suggested aspirin. She followed the therapy for 8 years, later she discontinued the treatment. After 2 years of discontinuation, she came to the hospital with the above symptoms.

On examination, the patient had a low-grade fever of 99.8°F with stable vitals.

Abdomen Doppler was done and gave the impression of hepatomegaly, splenomegaly, and moderate ascites. Hepatic veins were not visualized with a thrombosed collateral vein in the liver. Triphasic contrast-enhanced computed tomography (CECT) suggested BCS with non-visualized hepatic veins. Multiple arterial phase hyper-enhancing foci showing persistent enhancement in the later phases in both lobes of the liver. She has thrombosed intrahepatic venous collaterals and significant narrowing of the hepatic segment of the inferior vena cava. CT whole abdomen suggested multiple arterial phase hyper-enhancing foci showing persistent enhancement in the later phases in both liver lobes and bilateral minimal pleural effusions with underlying basal lung passive atelectasis. Cirrhosis secondary to BCS manifests as hepatomegaly with caudate lobe enlargement and an uneven nodular shape (Fig. 1).

Ascites in the abdomen had removed by pigtail drainage procedure, and they took an interventional radiologist opinion for stenting. Stenting was not possible, including direct intrahepatic portosystemic shunt (DIPS). The transplant versus surgical shunt

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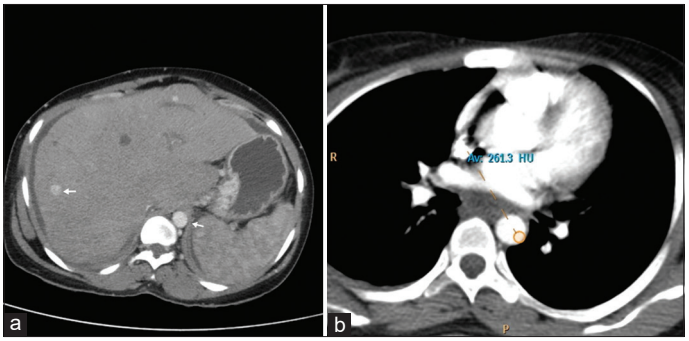

Figure 1: (a and b) Computed tomography scan shows hepatomegaly with caudate lobe hypertrophy and irregular nodular contour (cirrhosis) secondary to Budd-Chiari syndrome

procedures was discussed with the patient and relatives. They agreed to shunt surgery, and doctors suggested that she requires a liver transplant in the future if the shunt does not work. After the pre-anesthetic assessment, fitness, and consent, she underwent mesocaval shunt surgery and the Tru-cut gun liver biopsy. The surgery went uneventful.

Postoperative, she was treated with IV antibiotics, intravascular fluid, albumin, and other supportive measures. The patient improved well. Initially, they treated her ascitic drain output with injection albumin and diuretics, and gradually, drain production was decreased. She was put on tablet acinocoumarol and overlapped with injection enoxaparin for 3 days. The patient gradually recovered over the past 2 days. At discharge, she was hemodynamically stable, comfortable, ambulating, and tolerating a soft diet.

DISCUSSION

BCS is characterized by obstruction of hepatic venous outflow anywhere between the small hepatic veins and the supra-hepatic lower vena cava. It affects women of the reproductive age group in particular. The etiology and disease did not fully comprehend [5].

BCS has been classified into primary and secondary. It is regarded as primary if the blood flow had obstructed because of the primary venous entity. Thrombosis with primary hematological disorders and conditions of hypercoagulation usually has been deemed as secondary causes of the disease as the blood flow blocks compression or invasion of a hepatic venous outflow tract lesion, mainly due to benign malignant tumors, abscess, and cysts. Blocking hepatic venous flux, regardless of the cause, prevents normal blood drainage. Portal hypertension and intrahepatic and extrahepatic vessels are common in patients with BCS [3]. The venous blockage induces sinusoidal dilation and fluid leaking into the interstitial spaces by increasing hydrostatic pressure in the portal capillaries. When the lymphatic drainage capacity had surpassed, the liquid flowed through the Glisson capsule, increasing portal pressure and decreasing hepatic perfusion, causing cellular damage resulting from hypoxia [6].

The most common causes of BCS include inherited, acquired, and coagulable states. Inherited causes include factor 5 Leiden mutation, protein C and S deficiencies, antithrombin 3 deficiency, and prothrombin G20210A mutation which results in hepatic venous flow thrombosis and further precipitating BCS. Acquired causes include myeloproliferative disorders like polycythemia vera, paroxysmal nocturnal hemoglobinuria, essential thrombocytosis, and myelofibrosis. Other conditions reported as risk factors for the development of BCS include antiphospholipid syndrome, hypereosinophilic syndrome, Behcet disease, and ulcerative colitis [7].

Abdominal discomfort, ascites, hepatomegaly, jaundice, and leg swellings are some of the clinical symptoms. BCS affects up to 20% of asymptomatic people. The chronic type of BCS has characterized by the development of portal hypertension with ascites [1,8]. In our case report also, the patient had the same symptoms as in a chronic condition. Ultrasonography (USG) with Doppler study is essential for the diagnosis of BCS. Hepatic venography and inferior venography are valuable diagnostic tools. Apart from this, CT scan and MRI are also recommended. Similarly, our patient was screened by USG abdomen and Doppler, CT scan of the stomach, and Triphasic CECT scan to detect abnormalities in the liver. Liver biopsy is non-specific but often shows diagnostic characteristics of centrilobular and sinusoidal necrosis, chronic fibrosis, and other diagnoses [9].

The therapy of BCS has been classified in many different methods. Anticoagulant therapy and interventional therapy, such as surgical shunts, TIPS, balloon dilation angioplasty, and liver transplantation are the treatments of choice for most BCS patients. The cause of BCS determines the most appropriate treatment option. The obstruction's location, length, and the patient's physical condition can achieve a 95% 5-year survival in chronic cases by surgical shunt [10]. Tomita *et al*. reported three cases of BCS, one of which had subacute and handled with the transjugular intrahepatic portosystemic shunt [11]. In our case, there was no way to include DIPS in a stent, so our patient underwent mesocaval shunt surgery. Another patient in that case series had a factor 5 Leiden mutation that leads to progressive liver dysfunction which requires liver transplantation [11], similarly, our patient was also suggested for transplantation. Moreover, the third case used oral contraceptives that lead to BCS, but in this case, the patient did not use any contraceptive pills [11]. When shunting is not practicable or in fulminant situations, liver transplantation is the best alternative [10]. Stenting was not possible for our patient including DIPS. The patient was explained about the possibilities of liver transplantation and shunt surgery to the patient attendants, but they agreed to shunt surgery.

CONCLUSION

BCS is a rare disorder in the hepatic circulation that requires accurate diagnosis and immediate therapy. The patient should be counseled clearly about the illness and treatment to overcome the discontinuation of medications. Early detection of symptoms helps in early diagnosis and regaining the hepatic blood flow, either by angioplasty or radiology stenting before it affects the functioning of the liver.

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