CASE REPORT
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Twenty three years old Women with Budd Chiari Syndrome

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ABSTRACT

Bud-Chiari Syndrome (BCS) is a progressive and diffuse obstruction of hepatic venous drainage system characterized by abdominal pain, hepatomegaly, ascites and hepatic histology showing zone three sinusoidal distension and pooling. The syndrome may be triggered by the prothrombotic state which is a variety of conditions ranging from membranous webs in the supra hepatic segment of the inferior vena cava (IVC), to constrictive pericarditis or right heart failure. Hepatic vein thrombosis can be caused by several thrombotic illnesses, including antithrombin III deficiency, Polycythemia rubravera (PRV), Systemic lupus erythematosus (SLE), myeloproliferative disorders and the use of oral contraceptives. There have been reports of thrombosis in the IVC resulting from renal or adrenal cancer, as well as blockage from hepatocellular carcinoma and angiosarcoma. Actual cause of BCS is unknown in one fourth of cases. We illustrate a case with BCS that is manifested with dysponea, abdominal pain, bilateral pleural effusion and ascites.

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INTRODUCTION

Budd-Chiari Syndrome (BCS) is a rare disorder caused by either thrombotic or, non-thrombotic obstruction of the hepatic vein. It is two types: Primary BCS, when obstruction originates in the vein and thrombosis is the main cause; or secondary BCS, when there is external compression of the vein (such as from an abscess or tumor).[1] The presentation of BCS ranges from asymptomatic to fulminant hepatic failure, passing through the development of acute (rapid) or chronic (progressive) symptoms in a period of weeks to months prior to the diagnosis. The following circumstances are indicative of BCS: i) simultaneous presentation of ascites, hepatomegaly and upper abdominal pain, ii) massive ascites with mildly altered liver function tests, iii) sinusoidal dilation upon liver biopsy, without heart disease, iv) fulminant hepatic failure in association with hepatomegaly and ascites and v) unexplained chronic liver disease following exclusion of diagnoses of alcoholism, autoimmunity, chronic viral hepatitis B or C, Wilson's disease, iron overload and α-1 antitrypsin deficiency.[2,3]

As a rare case, it is worthwhile to report this case in order to raise awareness of the illness. This study discussed a 23-year-old woman with BCS presented with bilateral pleural effusion and dysponea.

The Case

A 23-year-old woman was admitted in the departmentof medicine of North Bengal Medical

College Hospital (NBMCH), Sirajganj, because of new-onset breathlessness, abdominal pain and distention of abdomen for the previous two weeks. She had a history of nausea, recurrent vomiting and orthopnea. Four years back she had a history of taking oral contraceptives for two years but at present she is not taking any medications. The rest of the history was unremarkable. Physical examination showed an ill-looking, mildly icteric and pyrexial, but conscious. Abdominal examination showed moderate ascites with hepatomegaly, but the spleen was not palpable. There was bilateralleg edema up to thigh. The rest of the examination was noncontributory. Initial laboratory tests revealed grossly abnormal liver function test results, with markedly elevated total bilirubin and liver enzymes. Serum albumin was low 1.20 gm/dl. The complete blood count was normal, but the prothrombin time was prolonged (more than twice the control).

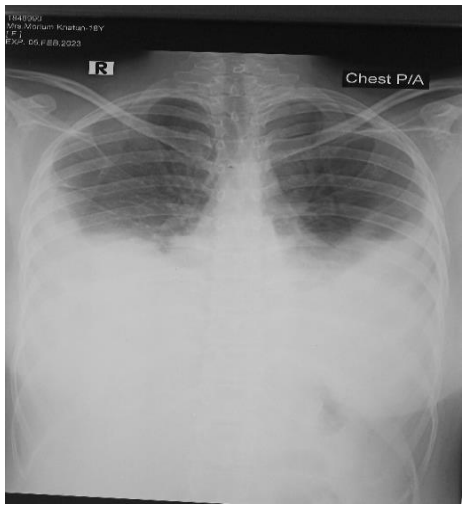

Figure 1: Chest X-ray P/A view showing bilateral pleural effusion

Analysis of pleural fluid revealed protein content of 1.9 gm%, RBC 120/cmm, WBC 1500/ccm with 91% lymphocyte, Adenosine deaminase (ADA) was negative. Cytology for malignant cells was negative. Other relevant blood tests were normal, and those included viral hepatitis screen, autoantibody profile and serology titers for RA, ANA, ANCA were negative.

X-ray chest P/A view showed bilateral pleural effusion (Figure 1). Abdominal ultrasound scan confirmed the presence of hepatomegaly and ascites, with the evidence of enlargement of spleen and bilateral pleural effusion. The liver appearance was not suggestive for fatty liver and cirrhosis. Endoscopy of upper GIT showed congestive gastropathy. She was kept under observation and under conservative management. Liver biopsy cannot be carried out. A repeat abdominal ultrasound scan showed that the liver was markedly enlarged with enlargement of the caudate lobe. This appeared to be compressing the inferior vena cava. Computed tomography (CT) scans of hepatobiliary system showed poorly visualized hepatic veins with poor filling of contrast, compressed IVC, hepatomegaly with coarse echo texture and multiple enhancing nodules, moderate ascites (Figure 2).

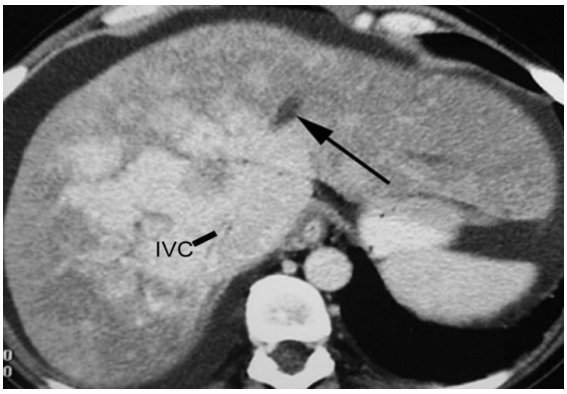

Figure 2: CT scan image of hepatobiliary system

The patient continued to deteriorate despite the best possible therapy with diuretics, vitamin K, low salt/high calorie diet, and salt-poor albumin infusions and other supportive management. She developed hepatic encephalopathy; her liver function did not improve and she became progressively more edematous and was refereed to Sheikh Russel Gastro liver hospital, Dhaka, Bangladesh for better management but she expired subsequently.

DISCUSSION

Budd in 1845 and Chiari in 1899 reported the occurrence of hepatic vein occlusion as well as the genesis of this syndrome.[4] The etiological

factors include: pregnancy, underlying malignancy, congenital webs in the inferior vena cava and/or hepatic veins, abdominal trauma, thrombotic events and use of oral contraceptives, cytotoxic chemotherapy, herbal tea and herbal medicines.[5,6] In a large number of cases, however, the cause remains unknown. In a series of 164 cases the etiology was either unidentified or inadequately established in 70.1%.[7] Most common manifestations of Budd-Chiari Syndrome (BCS) are tender hepatomegaly, ascites with or without abdominal pain. Previous studies revealed, a very few cases of BCS were reported with hepatic hydrothorax and bilateral pleural effusion. A retrospective review by McCarthy et al.[8] showed thirty patients with documented BCS with an overall mortality of 57%.

In our study patient, the most relevant etiological factors are the exposure to oral contraceptives. The mechanism of causation of the syndrome by oral contraceptives remains not clear. Time interval between the use of oral contraceptives and occurrence of BCS varies and ranging from two weeks up to five years. In summary, the combination of the rarity of BCS itself, the relatively low incidence of hepatic vein thrombosis related to contraceptive use, and the need for multiple risk factors makes a case of BCS in a female patient with a history of contraceptive use particularly uncommon.

Medical treatment for BCS alone was found to beassociated with 86% mortality; hepatic failure was the most common cause of death. The most widely applied surgical treatment for BCS at present is Porto systemic shunting, the site of the shunt is largely determined by the level of occlusion. For those with inferior vena cava obstruction and/or high IVC pressure shunts to the right atrium have been advocated.[8] However, shunts like these do not last longer because of their length. When the IVC is patent, then a portocaval or preferably mesocaval shunt would be more appropriate.[9-10] Surgical measures like orthotopic liver transplantation is the only curative treatment for this condition and this is best performed early in the clinical course of illness.[11]

CONCLUSION

Early recognition and treatment of BCS with systemic anticoagulants can prevent a fatal outcome, such as pulmonary embolism or liver failure, and may reverse hepatic functional abnormalities. Most patients with BCS are now treated by endovascular intervention, which has improved survival rate in those afflicted by this disease.

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Conflict of Interest: None

REFERENCES

1. Mancuso A. An update on management of Budd-Chiari syndrome. Ann Hepatol. 2014; 13: 323–326.
2. Aydinli M, Bayraktar Y. Budd-Chiari syndrome: Etiology, pathogenesis and diagnosis. World J Gastroenterol. 2007; 13: 2693–2696.doi:10.3748/wjg.v13.i19.2693.
3. DarwishMurad S, Plessier A, Hernandez-Guerra M, Fabris F, Eapen CE, Bahr MJ, et al. Etiology, management, and outcome of the Budd-Chiari syndrome. Ann Intern Med. 2009; 151: 167–175.doi: 10.7326/0003-4819-151-3-200908040-00004.
4. Lalonde G, Théoret G, Daloze P, Bettez P, Katz SS. Inferior vena cava stenosis and Budd-Chiari Syndrome in a woman taking oral contraceptives. Gastroenterol. 1982; 82(6): 1452-1456.
5. Bhattacharya SC, Anand R, Radhakrishnan AP, Murthy G. Budd chiari syndrome with hepato hydrothorax. Med J Armed Forces India. 1999; 55(4): 357-358.
6. Valla D, Le MG, Poynard T. Risk of hepatic vein thrombosis in relation to recent use of oral contraceptives. Gastroenterol. 1986; 90: 807-811.

7. Hoyuma AM, Schiff L, Helfman EL. Budd-Chiari syndrome in women taking oral contraceptives. Am J Med. 1971; 50: 137-139.
8. McCarthy PM, Van Heerden JA, Adson MA. The Budd-Chiari syndrome: medical and surgical management of 30 patients. Arch Surg. 1985; 120(6): 657-662.
9. Putmen CW, Porter K A, Weil R. Liver transplantation for Budd-Chiari syndrome. JAMA. 1976; 236: 1142-1143.
10. Starzl TE, Koep L, Porter KA, Schroter GP, Weil R, Hartley RB, et al. Decline in survival after liver transplantation. Arch Surg. 1980; 115(7): 815-819.
11. Kumana CR, Ng M, Lin HJ. Herbal tea induced hepatic veno-occlusive disease: quantification of toxic alkaloid exposure in adults. Gut. 1985; 26 (1): 101-104.