
Polycythemia Vera Presenting with Fulminant Hepatic Failure Due to Acute Budd-Chiari Syndrome: Case Report

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ABSTRACT

We describe a 38 year-old woman with polycythemia vera who presented with fulminant hepatic failure due to acute Budd-Chiari syndrome. She had a history of abdominal pain and distention for 4 days. Laboratory and clinical findings showed fulminant hepatic failure due to acute Budd-Chiari syndrome. Diagnosis was confirmed with abdominal ultrasonography and doppler ultrasonography showing ascites, hepatomegaly, portal hypertension and total occlusion of hepatic veins. Complete blood count and other clinical findings were compatible with polycythemia vera. The patient was successfully treated with urgent administration of continuous heparin infusion, repeated phlebotomies and hydroxyurea. We emphasize that early diagnosis and effective treatment in such fulminant cases can be life saving.

Key Words: Budd-Chiari syndrome, Polycythemia vera, Hepatic failure.

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INTRODUCTION

Budd-Chiari syndrome (BCS) is a rare syndrome resulting from obstruction of hepatic vein or inferior vena cava and presenting with painful hepatomegaly and ascites. Clinical presentation varies depending on the time and degree of the obstruction^[1,2]. If the occlusion is total and rapid, the patient may present with fulminant hepatic failure.

Mostly encountered causes of BCS are myeloproliferative diseases, especially polycythemia vera (PV), paroxysmal nocturnal hemoglobinuria, and other hypercoagulability states^[4-11]. Although the prognosis of acute BCS is poor, with early diagnosis and effective treatment, the outcome may be favorable. The case described here, is an acute form of BCS presented with fulminant hepatic fa-

ilure, treated successfully with anticoagulant treatment, and diagnostic and therapeutic approach is discussed.

CASE REPORT

A 38 year-old female was admitted to our hospital with rapidly progressive abdominal distention and pain. She had no personal or family history of thrombotic episodes. Physical examination revealed that she was confused and severely ill. She had ascites, painful hepatomegaly with a total span of 20 cm, massive splenomegaly, edema of the legs, and flapping tremor. Laboratory investigation on admission showed: Hemoglobin 20 g/dL, hematocrite 60.8%, MCV: 72 fL, white blood cell count $3.2 \times 10^9/\mu\text{L}$, platelet count $1.3 \times 10^9/\mu\text{L}$. Arterial blood gas values were within normal limits. Coagulation tests revealed elevated PT and aPTT; 24 and 45 seconds respectively. INR was 1.8. Liver enzymes were as follows: Aspartate aminotransferase (AST): 345 U/L, alanine aminotransferase (ALT): 450 U/L, gamma-glutamyltranspeptidase (g-GT): 342 U/L, alkaline phosphatase (AP): 452 U/L. Vitamin B12 level was 1511 pg/mL. Renal function tests were normal. Two units of phlebotomy was done in the emergency room. Ultrasonographic examination of the abdomen confirmed ascites, hepatomegaly and splenomegaly. On doppler ultrasonography no flow could be observed in hepatic veins. Inferior vena cava was patent. Bone marrow biopsy showed significant hypercellularity with marked increase of the number of megakaryocytes. At presentation increased hematocrite, white blood cell and platelet counts, splenomegaly, normal arterial oxygen saturation and bone marrow biopsy findings were compatible with polycythemia vera. In the light of clinical and laboratory data, the patient was diagnosed as fulminant hepatic failure due to acute BCS secondary to hyperviscosity caused by PV. Heparin infusion and hydroxyurea 1 g, 3 times per day was started promptly.

On the second day of the treatment, clinic and laboratory parameters of the patient improved. Ascites and edema of the lower extremities decreased and liver tests improved. On the 5th day of the treatment, liver transaminases and prothrombin time were within normal limits. On the 6th day

of admission coumadin was started and heparin was stopped. Now, 4 months after the thrombotic attack, the patient is on treatment with alpha-interferon and coumadin. She had no ascites and with normal complete blood counts and liver enzymes. On follow-up doppler ultrasonography, left hepatic vein was patent and left liver lobe was hypertrophic. No flow was observed in right hepatic vein, and right hepatic lobe was atrophic.

DISCUSSION

BCS is an uncommon disorder resulting from obstruction of hepatic veins or inferior vena cava. Most patients are young women. Diseases causing BCS are myeloproliferative diseases, paroxysmal nocturnal hemoglobinuria, protein C, antithrombin III deficiencies, and Factor V Leiden mutation^[3,4,6,7]. BCS is also defined in the clinical course of systemic lupus erythematosus antiphospholipid syndrome, disseminated intravascular coagulation, and Behçet's disease^[8,10-13]. BCS may be seen in oral contraceptive drug using women and pregnant women^[5,9]. Rarer causes of BCS are adrenal, renal and hepatic cancers invading inferior vena cava^[14-16]. In 30% of cases an underlying disease can not be found^[17].

Myeloproliferative diseases, especially PV is responsible from the majority (60%) of BCS^[3]. In some patients due to portal hypertension and hypersplenism, blood counts may be normal or low which may obscure the underlying myeloproliferative disease. Therefore in such patients in vitro endogenous colony formation should be performed which is pathognomonic to PV^[3].

Clinical presentation may change due to occlusion time and degree of the obstruction in the hepatic veins^[1,2]. If occlusion is total and rapid as in our case, the patient may present with fulminant hepatic failure. The clinician should not waste time with unnecessary invasive procedures as venography. Clinical findings and doppler ultrasonography are usually enough to have a diagnosis^[18]. If BCS is diagnosed early enough, fibrinolytic treatment may be worthwhile with variable results^[19]. Heparin should be administered at presentation as continuous infusion. For underlying PV, hematocrite should be reduced below 50%

with repeated phlebotomies, with simultaneous administration of hydroxyurea or a similar cytotoxic agent for thrombocytosis, if present.

This case was a fulminant form of BCS and we emphasise that, in this clinical setting, early diagnosis and appropriate treatment, both for BCS and underlying PV, may be life saving. Otherwise due to hepatic failure and its complications, coma and death is inevitable.

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