Budd-Chiari syndrome in a patient of diabetic ketoacidosis

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ABSTRACT

We report a rare case of Budd-Chiari syndrome developing in a patient undergoing treatment for diabetic ketoacidosis. A 27-year-old female presented with newly detected Type 1 diabetes with sepsis in ketoacidosis. During the process of treatment, she developed pain abdomen, ascites, and pedal edema. Investigations revealed an alteration of liver function and imaging characteristics of acute on chronic Budd–Chiari syndrome. All known etiological factors for Budd–Chiari syndrome were negative. Diabetic ketoacidosis, being a severely dehydrated state often associated with sepsis, may precipitate an acute presentation of previously asymptomatic Budd–Chiari syndrome.

Key words: Budd-Chiari syndrome, dehydration, diabetic ketoacidosis

INTRODUCTION

Budd–Chiari syndrome is a rare disease (occurring in one in a million) characterized by hepatic venous outflow obstruction at any level from the small hepatic veins to the atrio caval junction. It presents with the classical triad of abdominal pain, ascites, and hepatomegaly. Here, we describe a case of Budd–Chiari syndrome developing during ketoacidosis in a patient of Type 1 diabetes mellitus.

CASE REPORT

A 27-year-old unmarried woman, not a known diabetic, presented to our outpatient department with osmotic symptoms for 1 month and several episodes of vomiting and fever with mild dyspnea for 3 days.

On examination, she was found to have a body mass index (BMI) of 14.22 kg/m², pulse of 115/min, and blood pressure (BP) of 110/70 mmHg (supine) and 80/50 (erect). Dehydration was marked and she had acidotic breath. Pallor was present, edema was absent, and jugular venous pressure (JVP) was normal. Gastrointestinal system examination was normal except for mild splenomegaly. Examination of other systems did not reveal any abnormality.

Initial laboratory data showed fasting blood sugar 494 mg/dl, postprandial blood sugar 568 mg/dl, glycated hemoglobin (HbA1c) 18.7%, urinary ketones strongly positive, serum creatinine 0.6 mg/dl; arterial blood gas showed acidosis (pH 7.29, pCO₂ 33, pO₂ 90, HCO₃ 15 mEq/l). Total leukocyte count was 21,000/mm³ with a left shift, Hb was 9 g/dl, and erythrocyte sedimentation rate (ESR) was 65 mm at the end of first hour. Chest X-ray showed a consolidation in the left lung. Anti-glutamic acid decarboxylase (anti-GAD) antibody was subsequently found to be positive.

She was diagnosed to be a case of Type 1 diabetes mellitus, left lung consolidation, and sepsis with diabetic ketoacidosis. She was started on the standard protocol for diabetic ketoacidosis along with antibiotics. After the initial treatment, her condition stabilized gradually. However, from the 5th day of admission, she started complaining of abdominal pain and distension of abdomen along with swelling of the feet. Physical examination...
now showed development of hepatomegaly, increase in splenomegaly, ascites, and pedal edema. Investigation revealed a deranged liver function: total protein 7.1 g/dl, serum albumin 2.7 g/dl, total bilirubin 1.1 mg/dl (indirect 0.8), aspartate transaminase (AST) 147 U/l, alanine transaminase (ALT) 99 U/l, serum alkaline phosphatase 1431 U/l, and gamma-glutamyl transpeptidase (GGT) 1075 U/l. Workup included negative hepatitis B and C viral serologies. Echocardiography, 24-hour urinary albumin, and thyroid stimulating hormone (TSH) were normal.

Contrast-enhanced computed tomography (CECT) scan of the abdomen showed nodular liver surface with attenuated hepatic veins and proximal inferior vena cava (intrahepatic and suprahepatic regions) with intrahepatic collaterals, dilated portal vein, splenic vein, dilated mesenteric venous channels, splenomegaly, and moderate ascites, indicating hepatic parenchymal changes with portal hypertension and compromised hepatic venous outflow suggestive of Budd–Chiari syndrome.

Gastroenterology consultation was taken, and as advised, the following investigations were done. Antinuclear antibody (ANA) and dsDNA were negative. Platelet count, prothrombin time (PT) and activated partial thromboplastin time (APTT) were normal. Antithrombin III, α-fetoprotein, protein C and protein S, and cardiolipin levels were normal. Analysis of ascitic fluid showed total protein 1.7 g/dl, glucose 95 mg/dl, lactate dehydrogenase (LDH) 145 U/l, WBC 46/mm3, and serum-ascites albumin gradient of 1.8 g. Myeloproliferative disease and hyperhomocysteinemia were ruled out. Upper gastrointestinal (GI) endoscopy was normal.

She was started on anticoagulation, sodium restriction, diuretic therapy, and paracentesis. She was advised transjugular intrahepatic portosystemic shunt, which she refused. There was resolution of the edema and ascites with medical management. She was discharged with insulin, warfarin, and diuretics, and advised to attend endocrinology and gastroenterology OPD regularly. At 6 months follow-up, there was no edema or ascites, but no alteration in the size of thrombus.

**Discussion**

Budd–Chiari syndrome was first described in 1845 by George Budd to describe the classic triad of abdominal pain, hepatomegaly, and ascites. This syndrome is characterized by hepatic venous outflow obstruction at any level from the small hepatic veins to the atrio caval junction. Twenty percent of cases of Budd–Chiari syndrome are idiopathic.[1] In others, it may be caused by thrombosis of the hepatic vein or compression of the hepatic vein externally. Hepatic vein thrombosis may be associated with myeloproliferative disease, sepsis, dehydration, polycythemia vera, pregnancy, and hepatocellular carcinoma.

Posthepatic obstruction leads to increased sinusoidal pressure, hepatomegaly, perisinusoidal necrosis of hepatocytes, and eventually to liver failure with portal hypertension and ascites. The clinical presentation of patients is governed by the extent and swiftness of hepatic outflow obstruction juxtaposed to the body’s ability to decompress the liver via development of collateral blood flow. According to Langlet’s clinicopathological classification, patients are divided into the following types: type I (acute injury), type II (chronic lesions, corresponding to sequelae of remote hepatic outflow obstruction), and type III (acute injury superimposed on chronic lesions).[4]

Considering that our patient had an acute presentation in the background of asymptomatic cirrhosis of the liver, we categorized her as type III.

Diabetic ketoacidosis is a state of absolute or relative insulin deficiency characterized by ensuing hyperglycemia, dehydration, and acidosis-producing derangements in intermediary metabolism. Biochemically, it is defined as a concentration of blood glucose level >250 mg/dl, and a blood pH <7.3 and ketosis. Hyperglycaemia results in osmotic diuresis leading to hyperosmolarity and severe dehydration. An orthostatic change in blood pressure is indicative of a fluid deficit.[3] Though arterial thrombosis is common with diabetes ketoacidosis, venous thromboembolism is rare,[4] and there appear to be no previous specific reports of hepatic vein thrombosis in this setting. The prothrombotic state due to severe dehydration in the presence of sepsis was thought to be instrumental in precipitating the acute presentation of Budd–Chiari syndrome in this patient.

The only other report of Budd–Chiari syndrome and Type 1 diabetes was by Okamoto Hideki *et al.* described in a case during treatment for Type 1 diabetes.[7] Associations of Budd–Chiari syndrome with other autoimmune diseases have been reported in many studies. 3% cases of Budd–Chiari syndrome are attributed to Behcet’s disease.[9] Danalioğlu *et al.* have reported an association of Budd–Chiari syndrome with celiac disease.[9] Mouelhi reported a triple association of Graves’ disease, antiphospholipid syndrome, and Budd–Chiari syndrome.[10] To our knowledge, this is the first reported case of Budd–Chiari syndrome developing during diabetic ketoacidosis in a patient of Type 1 diabetes mellitus. In conclusion, we suggest that in patients of diabetic ketoacidosis with ascites and abdominal pain, this association should be kept in mind.
REFERENCES

1. Rajani R, Melin T, Björnsson E, Broomé U, Sangfelt P, Danielsson A, et al. Budd-Chiari syndrome in Sweden: Epidemiology, clinical characteristics and survival - an 18-year experience. Liver Int 2009;29:253-9.

2. Amarapurkar DN, Punamiya SJ, Patel ND. Changing spectrum of Budd-Chiari syndrome in India with special reference to non-surgical treatment. World J Gastroenterol 2008;14:278-85.

3. Chung RT, Iafrate AJ, Armein PC, Sahani DV, Misrai J. Case records of the Massachusetts general hospital. Case 15-2006. A 46-year-old woman with sudden onset of abdominal distention. N Engl J Med 2006;354:2166-75.

4. Langlet P, Escolano S, Valla D, Coste-Zeitoun D, Denie C, Mallet A, et al. Clinicopathological forms and prognostic index in Budd-Chiari syndrome. J Hepatol 2003;39:496-501.

5. Wyckoff J, Abrahamsone MJ. Diabetic Ketoacidosis and Hyperosmolar Hyperglycemic State. In: Joslin’s Diabetes Mellitus. 14th ed. Philadelphia: Lippincott Williams and Wilkins; 2005. p. 887-99.

6. Gill GV, MacNamara G, English P. Diabetic ketoacidosis complicated by axillary vein thrombosis. Diabetes Res Clin Pract 2006;73:104-6.

7. Hideki O, Shigey Y, Minako M. Budd-Chiari Syndrome in a Patient with Type 1 Diabetes, Autoimmune Thyroid Disease, and an Eating Disorder. A Case Report. J Jpn Diabetic Soc 2003;46:667-71.

8. Carvalho DT, Oikawa FT, Matsuda NM, Evora PR, Yamada AT. Budd-Chiari Syndrome in a 25-year-old Woman with Behçet’s Disease. J Med Case Reports 2011;5:52.

9. Danalioğlu A, Poturoğlu S, Güngör Gülkoğlu M, Demir K, Beşişik F, Çakaloğlu Y, et al. Budd-Chiari syndrome in a young patient with celiac sprue: A case report. Turk J Gastroenterol 2003;13:262-5.

10. Mouelhi L, Chaieb M, Debbeche R, Salem M, Sfar I, Trabelsi S, et al. Association Budd Chiari syndrome, antiphospholipid syndrome and Grave’s disease. Tunis Med 2009;87:164-6.

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