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A Case Report on Budd Chiari Syndrome – Which Mimics Multiple Disorders

Niveditha Dileep, John Thomas, James Jisha, and Abhijith V

ABSTRACT

Background: **Budd Chiari Syndrome (BCS) is the king of disorders which will mimic other disorders like chronic biliary disease, constrictive pericarditis, sinusoidal obstruction syndrome and so on, as it be like that, it is less diagnosed and treated in many countries. The prevalence of BCS is one in one million population, so it is a very rare case therefore it should be treated properly because many disorders like hematologic or malignant disease are the complications of BCS [1], [2].**

Objective: **To access the clinical variants of BCS along with the similarities and differences in clinical presentation, diagnostic approaches, and general treatment pattern which mimic, BCS thus gives the physician a clear outline about those disorders.**

Method: **A man of 42 years old having BCS was taken for the study to carry out the differences in clinical features of BCS which distinguish the mimicking disorders. Clinical presentations were noted. Laboratory tests and diagnostic tests showed that the patient is having comorbidities including fatty liver with cholelithiasis, mild splenomegaly, liver parenchyma diseases, large esophageal varies with signs of recent hemorrhage, port hypertensive gastropathy and minimal ascites [3]. Patient get discharged after feeling better.**

Result: **This patient is having chronic BCS with DIPS dysfunction. The patient had no history of liver disease before diagnosing BCS. BCS is almost curable when it is diagnosed correctly as early as possible. If not diagnosed early and treated well, comorbidities will occur, and it will affect the patient quality of life.**

Discussion: **Early detection and proper treatment will help to control the disease up to an extent. This is depending on the physician's knowledge. Hence, this case study clearly explains the disorders which mimic BCS for better understanding.**

Keywords: Budd Chiari Syndrome, constrictive pericarditis, chronic biliary disease, veno occlusive disorder.

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I. Introduction

BCS is a rare disorder that is characterized by hepatic venous outflow obstruction without right heart failure [4]. Comparing with other chronic liver disorders, BCS has an acceptable prognosis. The obstruction can be thrombotic or non-thrombotic. The prevalence of BCS is one in one million population, hence it is a very rare case therefore it should be treated properly because many disorders like hematologic or malignant disease are complicated with BCS [5].

BCS is a king of disorders which will mimic much other disorder like chronic biliary disease, constrictive pericarditis, sinusoidal obstruction syndrome and so on (Table I) [6], [7]. This case report explains the condition of a patient having chronic BCS who underwent Direct Intrahepatic Portocaval Shunt procedure and also explains the other disorders having similar symptoms with BCS.

TABLE I: Disorders with Different Diagnostic Tests [21]

Diseases	Diagnostic tests
Chronic biliary disease	Blood test
	Liver function test
	ERCP
	CT
	MRI
	Liver biopsy
Budd Chiari Syndrome	Ultrasonography
	CT
	MRI
	Venography
	Biopsy
Constrictive pericarditis	Echocardiography
	CT
	MRI
	Invasive hemodynamic measurement
Venous occlusive disorder	Liver tests including GGT
	Doppler ultrasonography

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II. Methodology

A. Case Report

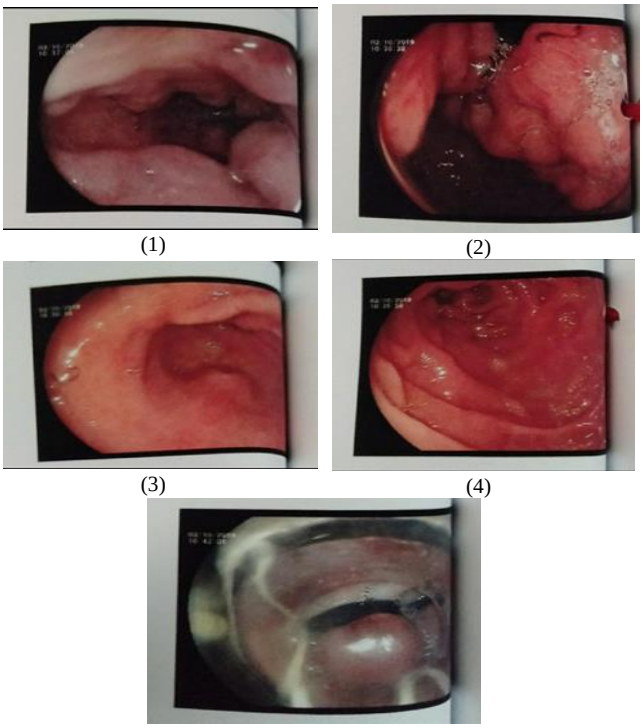
A man of 42 years old, admitted to the gastroenterology department over the complaints of lethargy, coffee coloured vomiting. He is a known patient of chronic BCS, who underwent Direct Intrahepatic Portocaval Shunt (DIPS) procedure two years back for recurrent variceal bleeding and was on anticoagulant since then. He was doing well until now when he developed multiple episodes of hemetemesis with melena for which an emergency Oesophago-Gastro-Duodenoscopy (OGD) was done, which revealed grade 2 esophageal varices with active bleeding. Endoscopic Variceal Ligation (EVL) was done, and hemostasis was achieved. The patient underwent repeat Contrast-Enhanced Computed Tomography (CECT) upper abdomen for stent review which was normal. Further DIPS demography and pressure gradient measures showed raised gradients suggestive of stent dysfunction.

During the course of admission, laboratory reports were checked and collected. Blood urea was found to be below than normal range as the patient is having low protein diet and abnormal liver function. Packed Cell Volume (PCV), Mean Corpuscular Volume (MCV), Mean Corpuscular Hemoglobin (MCH), total Red Blood Cells (RBC), platelet count and Hemoglobin were found to be decreased due to blood loss. Erythrocyte Sedimentation Rate (ESR) and C Reactive Protein (CRP) were found to be elevated as the patient is having an infection. Total bilirubin and Alkaline phosphatase were elevated due to hepatocellular dysfunction (Table II).

TABLE II: Abnormal Laboratory Report of the Patient

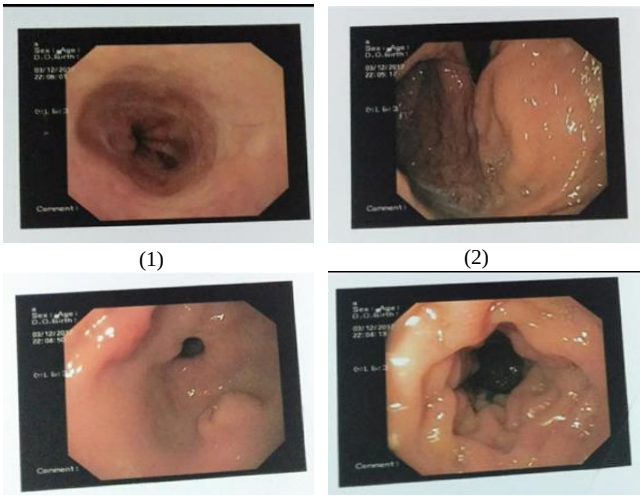
Test	Patient value	Normal value
Blood urea	8	15-45 mg/dl
PCV	26.2	35-49%
MCHC	31.7	32-36%
ESR	28	0-20mm/hr
MCH	25.7	26-32Pg
Total RBC	3.23	3.8-5.6 million/cu
Platelet count	119000	150-450*1000 c/cu
Hb	7.8	11-18g/dl
CRP	8.7	0-6mg/l
ALP	273	38-126 U/L
Total bilirubin	3.4	1-1.2mg/dl

UltraSound Scan (USS) abdomen/pelvis showed fatty liver with cholelithiasis. UltraSound Sonography test (USG) abdomen showed mild splenomegaly and liver parenchyma diseases. Endoscopic findings are large esophageal varies with signs of recent hemorrhage, port hypertensive gastropathy, Gastric Antral Vascular Ectasia (GAVE) (Fig. 1 and 2). CECT whole abdomen (triple phase) showed minimal ascites along with cholelithiasis and chronic liver parenchyma disease.



(1) (2) (3) (4) (5)

Fig. 1. Upper Gi endoscopy report of 2019.



(1) (2) (3) (4)

Fig. 2. Upper Gi endoscopy report of 2017.

III. Result

This case report is a 42-year-old male patient having chronic BCS with DIPS dysfunction. The patient had no history of liver disease. BCS is curable when it is diagnosed correctly as early as possible [9], [10]. Some diseases can mimic BCS and thus making the Physician under confusion. This case report mentioning those diseases along with common symptoms, major diagnostic tests, and general treatment options (Table I, III and VI). Differential diagnosis of BCS will confuse the Physician to make a final diagnosis. It is important that the Physician must be well-known about the other diseases mimicking BCS. So special considerations should be taken while handling BCS cases because early diagnosis will help the patient to recover immediately and can lead their normal life, but late diagnosis and treatment will lead to liver damage within 3 months and liver failure within 3 years.

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TABLE III: DISORDERS WITH GENERAL TREATMENT APPROACHES [15], [18]

Diseases	General treatment
Chronic biliary disease	Antibiotics Hepatoportoenterostomy ERCP Cholecystectomy
Budd Chiari Syndrome	Anticoagulants Thrombolytics Diuretics Ballon angioplasty TIPS Liver transplantation
Constrictive pericarditis	Pericardiectomy Diuretics NSAIDS Corticosteroids COX 2 inhibitors
Venom occlusive disorder	Low dose tissue plasminogen activator Anticoagulants Defibrotide Diuretics

IV. DISCUSSION

George Budd, a British internist, described three cases of hepatic vein thrombosis in 1845 and Hans Chiari, an Austrian pathologist, added the first pathologic description of a liver with "obliterating endophlebitis of the hepatic veins" in 1899, together named as "Budd Chiari Syndrome". The synonym of BCS is Rokitansky's disease [12].

This case report typically mentioning the diseases which mimic BCS, which include chronic biliary disease, constrictive pericarditis, venous-occlusive disease (Table VI). The clinical variants of BCS are mentioned in (Table IV). The prognosis widely depends on the level of bilirubin, creatinine, and ascites in the body.

TABLE IV: THE CLINICAL VARIANTS OF BCS [23]

Type	Characteristics
Acute and subacute	Rapid abdominal pain Ascites Hepatomegaly Jaundice
Chronic	Progressive ascites Jaundice is absent 50% of the patients will have renal failure.
Fulminant	Hepatic failure Ascites Tender hepatomegaly Jaundice Renal failure.

A score less than 5.4 is considered with a good prognosis. Here the patient is having 5.96 as prognostic index (as mentioned in Table V).

TABLE V: THE FORMULA USED TO CALCULATE THE PROGNOSTIC INDEX FOR BCS IS [23]

Prognostic index = (Ascites score × 0.75) + (pugh score × 0.28) + (age × 0.037) + (creatinine level × 0.0036)

The Disorders Sharing Common Characteristics Include:

- *Constrictive pericarditis*

Constrictive pericarditis is a chronic inflammation of the pericardium [19]. On examination, other than cardiovascular findings, constrictive pericarditis shows similar gastrointestinal findings with BCS [8], [11].

- *Veno occlusive disorder*

Veno occlusive disease also known as sinusoidal obstruction syndrome, happens when the small blood vessels that are leading into the lover become blocked [13]. BCS suffers from veno occlusive disease, based on the level of obstruction [14]. In veno occlusive disorder, the obstruction occurs at the level of sinusoids and terminal venules but in BCS, the obstruction occurs from hepatic veins to the superior end of inferior vena cava [17].

- *Chronic biliary disease*

Chronic biliary disease refers to diseases affecting the bile duct, gall bladder and other areas involved in the production and transportation of bile [16], [22].

TABLE VI: DISORDERS WITH COMMON SYMPTOMS [20]

Diseases	Common symptoms
Chronic biliary disease	Jaundice Abdominal pain Light brown urine
Budd Chiari Syndrome	Jaundice Ascites Hepatomegaly Splenomegaly Ankle edema Renal failure
Constrictive pericarditis	Tachycardia Fatigue Fever Ascites Peripheral edema
Venom occlusive disorder	Primary liver disease Hepatomegaly Ascites Jaundice Weight gain

This study will help to understand the difference between BCS and other mimicking disorders as it clearly explaining the common sign and symptoms along with diagnostic approaches and treatment options, which will help the Physician a clear cut knowledge in those diseases and thus they can provide a better therapy to their patients. Also, this case report is about a patient developed BCS without having a past history of liver diseases, thus Physicians must be aware about these kinds of rare diseases that are increasing day by day.

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REFERENCES

[1] Internal Medicine, 4th Ed.: Jay H. Stein, Editor-In-Chief; Mosby-Year Book, Inc., 1994. Pp. 632-33.
[2] Leevy CM, et al, eds. Diseases of the Liver and Biliary Tract: Standardization of Nomenclature, Diagnostic Criteria and Prognosis. New York, NY: Raven Press; 1994:161-62.
[3] Aydinli M, Bayraktar Y. Budd-Chiari syndrome: etiology, pathogenesis and diagnosis. World J Gastroenterol 2007; 13:2693–2696

[4] Blendis L. Budd-Chiari syndrome: a clinical model of the hepatorenal reflex? Gastroenterology 2006; 131:671–672
[5] Michl P, et al. Successful treatment of chronic Budd-Chiari syndrome with a transjugular intrahepatic portosystemic shunt. J Hepatol. 2000;32:516-20.
[6] Gupta S, et al. Comparison of ultrasonography, computed tomography and 99mtc liver scan in diagnosis of Budd-Chiari syndrome. Gut. 1987;28:242-47.
[7] Vons C, et al. Results of portal systemic shunts in Budd-Chiari syndrome. Ann Surg. 1986;203:366-70.
[8] Okuda K, Kage M, Shrestha SM. Proposal of a new nomenclature for Budd-Chiari syndrome: Hepatic vein thrombosis versus thrombosis of the inferior vena cava at its hepatic portion. Hepatology 1998;28:1191-8.
[9] Mukund A, Gamanagatti S. Imaging and interventions in Budd-Chiari syndrome. World J Radiol 2011;3:169-77.
[10] Boozari B, Bahr MJ, Kubicka S, Klempnauer J, Manns MP, Gebel M. Ultrasonography in patients with Budd-Chiari syndrome: Diagnostic signs and prognostic implications. J Hepatol 2008;49:572-80.
[11] Okuda K. Membranous obstruction of the inferior vena cava (obliterative hepatocavopathy, Okuda). J Gastroenterol Hepatol. 2001;16:1179-1183.
[12] Shin N, Kim YH, Xu H, Shi HB, Zhang QQ, Colon Pons JP, Kim D, Xu Y, Wu FY, Han S, Lee BB, Li LS. Redefining Budd-Chiari syndrome: A systematic review. World J Hepatol 2016; 8(16): 691-702 [PMID: 27326316 DOI: 10.4254/wjh.v8.i16.691].
[13] DeLeve LD, Valla DC, Garcia-Tsao G. Vascular disorders of the liver. Hepatology. 2009;49:1729-1764.
[14] McDermott WV, Ridker PM. The Budd-Chiari syndrome and hepatic veno-occlusive disease. Recognition and treatment. Arch Surg. 1990;125:525-527.
[15] Darwish Murad S, Plessier A, Hernandez-Guerra M, Fabris F, Eapen CE, Bahr MJ, Trebicka J, Morard I, Lasser L, Heller J. Etiology, management, and outcome of the Budd-Chiari syndrome. Ann Intern Med. 2009;151:167-175.
[16] Faraoun SA, Boudjella Mel A, Debzi N, Afredj N, Guerrache Y, Benidir N, Bouzid C, Bentabak K, Soyer P, Bendib SE. Budd-Chiari syndrome: a prospective analysis of hepatic vein obstruction on ultrasonography, multidetector-row computed tomography and MR imaging. Abdom Imaging. 2015;40:1500-1509.
[17] Rautou PE, Douarin L, Denninger MH, Escolano S, Lebrec D, Moreau R, Vidaud M, Itzykson R, Moucari R, Bezeaud A. Bleeding in patients with Budd-Chiari syndrome. J Hepatol. 2011;54:56-63.
[18] Ebrahimi M, Modaghegh MH, Esmaeilzadeh A. Presentation of hospital outcomes and different treatment methods of patients with budd-Chiari syndrome: a report from two tertiary hospitals in iran. Med Princ Pract. 2011;20:287-290.
[19] Arora, Anil M.D.; Tandon, Nikhil M.D.; Sharma, M. P. M.D.; Acharya, S. K. M.D. Constrictive Pericarditis Masquerading as Budd Chiari Syndrome, Journal of Clinical Gastroenterology: April 1991 - Volume 13 - Issue 2 - p 178-181.
[20] Valla DBudd–Chiari syndrome and veno-occlusive disease/sinusoidal obstruction syndromeGut 2008;57:1469-1478.
[21] DeLeve LD,Valla DC,Garcia-Tsao G, Vascular disorders of the liver. Hepatology (Baltimore, Md.). 2009 May.
[22] Tanaka M,Wanless IR, Pathology of the liver in Budd-Chiari syndrome: portal vein thrombosis and the histogenesis of veno-centric cirrhosis, veno-portal cirrhosis, and large regenerative nodules. Hepatology (Baltimore, Md.). 1998 Feb.
[23] [23] Zeitoun G,Escolano S,Hadengue A,Azar N,El Younsi M,Mallet A,Boudet MJ,Hay JM,Erlinger S,Benhamou JP,Belghiti J,Valla D, Outcome of Budd-Chiari syndrome: a multivariate analysis of factors related to survival including surgical portosystemic shunting. Hepatology (Baltimore, Md.). 1999 Jul.

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