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.

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.
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 hematemesis 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

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

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 varices 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.

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.
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.

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).

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 liver 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].

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


Niveditha Dileep was born on 17.08.1998 in Cherthala, Kerala. She is pursuing my Pharm D at KVM College of Pharmacy (KUHS), Cherthala, Kerala, India. She has published articles like:

John Thomas was born on 19.02.1973 in Anikkad Kerala. He has completed his PhD in Pharmaceutical Sciences from Research Development and Innovation Centre (RDIC) C.U. Shah University, Gujarat, India in 2020 and completed his Master in Pharmacy in the field of Pharmaceutical Chemistry from JSS College of Pharmacy, Ooty under The Tamil Nadu Dr. MGR Medical University, Chennai, Tamil Nadu, India in 1999. Major field of study is design and development of life saving pharmacophores by synthesis and isolation of natural products, critical observations and evaluations on natural remedies.

He is currently working as academic director and professor at KVM College of Pharmacy Cherthala, Kerala, India. He is experienced as research associate in National Institute of Pharmaceutical Education and Research (NIPER), Mohali Punjab India and Senior Lecturer at Masterskill University College of Health and Allied Sciences, Kuala Lumpur Malaysia. He has published articles like:

Dr. Thomas, Academic Director and Professor of Department of Pharmaceutical Chemistry.

James Jisha was born on 25.05.1975 in Vennmoney Kerala. She is am pursuing her PhD in JITU, Rajasthan, India in 2020 and completed her Master in Pharmacy in the field of Pharmacutics from KM College of Pharmacy, Madurai affiliated to the Tamil Nadu Dr. MGR Medical University, Chennai, Tamil Nadu, India in 1999. Major Field of study is design and formulation of Novel Drug Delivery Systems by using natural ingredients.

She is currently working as HEAD and PROFESSOR in Department of Pharmaceutics at KVM College of Pharmacy Cherthala, Kerala, India. She was Senior Lecturer at Masterskill University College of Health and Allied Sciences, Kuala Lumpur Malaysia. She has published articles like:

Mrs. James is working as Head and Professor of Department of Pharmaceutics.

Abhijith V. was born on 21.09.1997 in Alappuzha Kerala. He is pursuing his M.B.A Hospital Management in Bharathari University Coimbator and completed my Bachelor of Pharmacy from KVM College of Pharmacy Cherthala affiliated to Kerala University of Health Sciences in 2019. He is working as Business Development Executive at Natco Pharma Pvt ltd India. He was pharmacist at Deepa Hospital karuvatta, Kerala India. He has published articles like Huntington Disease and Genetical Relation- A Case Study in Kerala, IOSR Journal of Pharmacy, 2020.

Mr. V. is working as Business Development Executive at Natco Pharma.