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CASE REPORT

Case Report of Budd-Chiari Syndrome secondary to hepatic vein thrombosis

Mukesh Kumar Saphi [1 *], Sunny Kumar Yadav[1], Sai Vijaya Durga Yalla [1], Tejaswi Allu [1], Amit Kumar [2]

[1] *Pharm.D Intern, Department of Pharmacy Practice, Aditya College of Pharmacy, Surampalem, Andhra Pradesh, India*

[2] *Associate Professor, Department of Pharmacy Practice, Aditya College of Pharmacy, Surampalem, Andhra Pradesh, India*

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Abstract: Budd-Chiari syndrome is a rare disorder characterized by obstruction of hepatic venous outflow which can range from asymptomatic to severe manifestations including liver dysfunction. We present a case of a 29-year old male who presented with abdominal distension, breathlessness and dry cough. He had a history of Budd Chiari syndrome secondary to hepatic vein thrombosis for two years and portal vein thrombosis with cavernous transformation. Clinical examination revealed hepatomegaly and ascites. Laboratory investigations showed elevated liver enzymes and imaging revealed portal vein thrombosis with cavernous transformation. He was diagnosed with Budd-Chiari syndrome secondary to hepatic vein thrombosis. He underwent IVC venoplasty under fluoroscopy guidance using balloons with significant improvement seen post-procedure. Standard medical management including anticoagulation, antibiotics and supportive care was also provided. His liver function tests significantly improved after treatment. IVC venoplasty combined with medical management can be an effective treatment option for Budd-Chiari syndrome secondary to hepatic vein thrombosis.

Keywords: Budd-Chiari syndrome; Hepatic vein thrombosis; IVC venoplasty; Portal vein thrombosis; Liver function tests

1. Introduction

Budd-Chiari syndrome is a rare medical condition characterized by the blockage of blood outflow from the liver, primarily affecting the hepatic veins. This obstruction can manifest at different levels, including the small hepatic veins within the liver, the major hepatic veins as they exit the liver, or the inferior vena cava where the hepatic veins drain into. The consequence of this blockage is an increase in pressure within the liver veins, leading to blood congestion within the liver tissue. The clinical manifestations of Budd-Chiari syndrome vary widely based on the severity and duration of the venous outflow obstruction. The estimated global incidence of this syndrome is exceptionally low, with only 1-2 cases per million people, and it is slightly more prevalent in females. The condition is primarily associated with specific causes, with myeloproliferative neoplasms accounting for approximately 50% of cases. Around 30% of cases are linked to genetic or acquired conditions that heighten the risk of blood clot formation, while the remaining cases often result from structural or anatomical abnormalities that compress the hepatic veins.

Timely identification of the underlying cause is crucial in managing Budd-Chiari syndrome effectively. Early initiation of appropriate treatment is essential to prevent long-term complications and reduce mortality. Swift diagnosis helps mitigate progressive damage to liver cells, preventing complications such as liver failure and life-threatening bleeding from esophageal varices. The management of Budd-Chiari syndrome involves aggressive interventions aimed at reopening blocked vessels and preventing further obstruction.

We present a case of Budd Chiari syndrome secondary to hepatic vein thrombosis managed successfully with IVC venoplasty along with standard medical management. The objective is to discuss the clinical presentation, diagnostic evaluation, treatment approach and outcome of this case to add to existing literature on management of this rare disorder.

2. Case presentation

A 29-year-old male patient presented to the emergency department with complaints of abdominal distension and breathlessness for past 4 months and dry cough for 5 days.

* Corresponding author: Mukesh Kumar Saphi

2.1. Clinical history

On taking history, it was noted that the patient had a known case of Budd Chiari syndrome for past 2 years secondary to hepatic vein thrombosis. He also gave history of portal vein thrombosis with cavernous transformation for which he was on regular follow up. He had no history of alcohol consumption or drug abuse. There was no relevant family history.

2.2. Diagnostic Evaluation

On examination, the patient was conscious and oriented but dyspneic at rest. Vitals showed tachycardia and abdominal examination revealed distended abdomen with shifting dullness and moderate splenomegaly. Initial workup revealed normocytic anemia, thrombocytopenia and deranged liver function tests with elevated bilirubin, AST, ALT and alkaline phosphatase. Abdominal ultrasound showed features of portal vein thrombosis with cavernous transformation and altered hepatic vein Doppler study. Contrast enhanced computed tomography of abdomen confirmed the diagnosis of hepatic vein thrombosis with features of portal hypertension. Based on history, clinical examination and imaging findings, a diagnosis of Budd Chiari syndrome secondary to hepatic vein thrombosis was considered for the patient.

2.3. Imaging

Abdominal ultrasound revealed features suggestive of portal vein thrombosis with cavernous transformation. The hepatic veins could not be visualized clearly and there was evidence of collateral vascular channels. Contrast enhanced computed tomography (CECT) abdomen showed altered hepatic venous anatomy with evidence of thrombosis. There was cavernous transformation of portal vein with multiple collateral channels. Splenomegaly was also noted.

2.4. Laboratory investigations

Liver function tests showed elevated bilirubin of 2 mg/dL, serum glutamic-pyruvic transaminase (SGPT) of 80 U/L and serum glutamic-oxaloacetic transaminase (SGOT) of 90 U/L. Total leukocyte count was 8500/cumm and platelet count was 1,20,000/cumm. Prothrombin time was found to be 14 seconds. Renal function tests and other routine blood investigations were within normal limits.

2.5. Diagnosis

Based on the history of Budd-Chiari syndrome 2 years back, history of portal vein thrombosis, imaging findings of hepatic vein thrombosis and cavernous transformation of portal vein with features of portal hypertension on ultrasound and CECT abdomen along with deranged liver function tests, the patient was diagnosed as a case of Budd-Chiari syndrome secondary to hepatic vein thrombosis with portal vein involvement.

2.6. Treatment

Considering the patient's condition and history, it was decided to proceed with recanalization of the obstructed inferior vena cava via invasive procedure along with medical management.

2.6.1. Interventional procedure

The patient underwent IVC venoplasty under fluoroscopy guidance in interventional radiology department. Through the transfemoral route, an 8mm balloon catheter was positioned across the obstruction in IVC and inflated to achieve recanalization. Post procedure venography showed reperfusion of hepatic veins

2.6.2. Medical management

The patient was started on subcutaneous low molecular weight heparin and oral anticoagulant (Warfarin). He was also given diuretics, antibiotics and nutritional supplements

2.6.3. Outcome and follow-up

The patient improved clinically with resolution of symptoms over next few days. His liver function tests started improving after 1 week. He was discharged on anticoagulants and advised regular follow up. On follow up after 1 month, his LFTs normalized and ultrasound showed patent IVC and no new thrombus formation. The patient continues to do well on 6 months follow up.

3. Discussion

Budd-Chiari syndrome is a rare disorder resulting from hepatic venous outflow obstruction. [4] It can present with non-specific manifestations ranging from asymptomatic to severe complications like liver failure. [5] Various etiologies reported are myeloproliferative neoplasms, infections, genetic and acquired thrombophilia. [6]

In the index case, the underlying etiology was likely hepatic vein thrombosis secondary to his known history of portal vein thrombosis and hypercoagulable state. This led to features of Budd Chiari syndrome like hepatomegaly and ascites. Diagnosis was based on imaging findings and laboratory parameters suggestive of impaired hepatic function. The standard treatment for Budd Chiari syndrome involves recanalizing the obstruction to reduce venous pressure in the liver. [7] Options include transjugular intrahepatic portosystemic shunt (TIPS), open surgery and percutaneous transluminal angioplasty (PTA). [8] In this patient, IVC venoplasty was done which achieved recanalization as seen on post procedure venography. This led to clinical and biochemical improvement.

Medical management including anticoagulation plays an important role post intervention to prevent re-thrombosis. [9, 10] Warfarin was chosen due to its effectiveness and ease of administration orally on long term basis. Diuretics provided symptomatic relief from ascites. Antibiotics were administered to prevent any infections. The patient showed complete resolution of symptoms and improvement in liver function tests indicating success of the treatment approach followed. Regular follow up helped monitor his condition and compliance to anticoagulation therapy. With combined endovascular recanalization of obstruction and medical therapy, a good outcome can be achieved in properly selected patients of Budd Chiari syndrome as observed in this case. A multidisciplinary team approach involving radiologists and hepatologists is vital for management of this rare disorder.

4. Conclusion

This case highlights the successful management of Budd Chiari syndrome secondary to hepatic vein thrombosis with a combination of IVC venoplasty and standard medical therapy. IVC venoplasty helped in recanalization of the obstruction along with anticoagulation preventing further thrombosis. The patient showed significant improvement in liver function tests and resolution of symptoms post intervention. A multidisciplinary approach involving interventional radiology along with medical management can be effectively used for treating Budd Chiari syndrome secondary to hepatic vein thrombosis and prevent disease progression.

Abbreviations

BCS: Budd-Chiari Syndrome
IVC: Inferior Vena Cava
CT Scan: Computed Tomography Scan
PVT: Portal Vein Thrombosis
HV: Hepatic Vein
ALT: Alanine Transferase
AST: Aspartate Transferase
CBP: Complete Blood Picture

Compliance with ethical standards

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Conflict of interest statement

All authors declare that there is no conflict of interest.

Statement of ethical approval

If studies involve use of animal/human subject, authors must give appropriate statement of ethical approval. If not applicable then mention 'The present research work does not contain any studies performed on animals/humans subjects by any of the authors.

Statement of informed consent

Informed consent was taken from the patient.

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Author's short biography

Mukesh Kumar Saphi:

Pharm-D intern with a relentless commitment to transforming healthcare through pharmaceutical expertise and focused on merging clinical knowledge with a patient-centered approach to ensure optimal medication therapy outcomes. Dedicated to lifelong learning, innovation, and making a meaningful impact in the ever-evolving landscape of pharmacy practice. Attended Seminar on Various Approaches of Pharmacy and Pharmaceutical Field and Completed an ECG Interpretation Master certification course from CliMed

Sunny Kumar Yadav:

PharmD student with a fervour for the intersection of healthcare, pharmacy, and pharmaceutical industry. Grounded in a strong academic foundation, I aspire to apply pharmaceutical knowledge to enhance patient well-being. Committed to fostering a compassionate and evidence-based approach in pharmacy practice. I am eager to embrace challenges, learn, and contribute to the dynamic landscape of modern healthcare

Sai Vijaya Durga Yalla:

PharmD student dedicated to the art and science of pharmacy. Fueled by a passion for patient-centered care, I aim to leverage my academic background in pharmaceutical sciences to make a positive impact on healthcare outcomes. Eager to integrate clinical expertise with a compassionate approach, I strive for excellence in the evolving field of pharmacy practice

Tejaswi Allu:

Let people know you and your contribution in the area. A short biography of author. Font size 10pt, Cambria, Justified alignment. Do not include affiliation. This is already mentioned in first page. Write here mainly about research interest. It should be about authors academic degree, Research area, and Years of experience

Amit Kumar:

He is an accomplished professional in the field of pharmacy, holding a B. Pharmacy, M. Pharmacy, and had submitted his Ph.D. Currently serving as the Associate Professor and Head of the Pharmacy Practice Department at the NAAC A accredited Aditya College of Pharmacy in Surampalem. He has demonstrated his commitment to advancing pharmaceutical knowledge through his extensive publication record, with 33 articles published in various reputed Indian and international journals. His research contributions span a range of topics within the pharmaceutical domain, showcasing his expertise and dedication to the field.