

## CASE REPORT

# Budd Chiari Syndrome

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### ABSTRACT

*Budd Chiari syndrome is a rare hepatic disorder characterized by obstruction of hepatic blood flow caused due to lesion formation in the liver. This report refers to a 16-year-old female who presented to the hospital with chief complaints of abdominal distension and discomfort for 10 days and dry cough in the past 5 days. She had similar complaints one year back. Based on her clinical presentations, ultrasonography, computed tomography, and venography results, she was diagnosed with Budd Chiari syndrome which develops as a result of lesion formation, and was treated with the following medications such as diuretics, anticoagulants, broad-spectrum antibiotics, and hepatoprotective agents and surgical procedures such as direct intrahepatic portosystemic shunt was also performed. The patient showed improvement in the symptoms after the surgery. Budd Chiari syndrome, if left untreated can be fatal.*

**Keywords:** - Budd Chiari syndrome, lesion, Abdominal distension, Direct intrahepatic portosystemic shunt.

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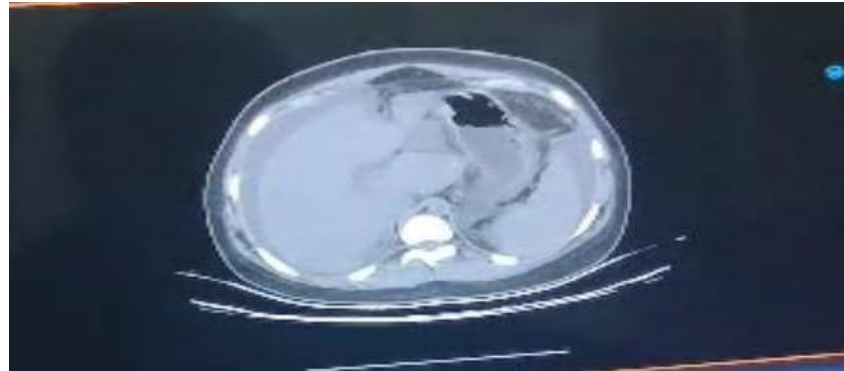
### INTRODUCTION

Occlusion of the hepatic outflow between the junction of the inferior vena cava with the right atrium and small hepatic veins characterizes the rare and possibly fatal illness known as Budd-Chiari syndrome (BCS).[1] It is an uncommon condition characterized by hepatomegaly, pain in the upper right portion of the abdomen, and ascites. This syndrome is observed in developing nations like China, India, Nepal, and South Africa affecting typically 1 in 100,000 people. [2,3] Budd-Chiari syndrome is more prevalent in women in Non-Asian nations during the third or fourth decade of their life. In contrast, it is more prevalent in men of Asian descent with a median age of 45 years old. [4] We here report a case of Budd-Chiari syndrome presenting with abdominal distension and discomfort, treated with surgical intervention.

### CASE REPORT

A 16-year-old female patient from a rural backward area of the central part of Andhra Pradesh was presented with complaints of abdominal distension and abdominal discomfort for 10 days without any association of food intake and dry cough for 5 days. She had similar type of symptoms 1 year back and visited a hospital in her home town where ascitic fluid analysis was done with TLC=100 of which 95% lymphocytes and gram stain and AFB of ascitic fluid was negative. A complete blood picture of the patient showed high peripheral white blood cells count 10,700 cells/cu<sup>3</sup> and increased total bilirubin 1.4 mg/dl and direct bilirubin 0.4 mg/dl and decreased serum proteins: 6.2 g/dl and increased prothrombin time to 16.1 sec and increased INR: 1.40. Ultrasonography scan showed a diffuse altered attenuation within the homogenous mottled enhancement on contrast administration giving nutmeg appearance and hypertrophy of caudate lobe. Venography revealed mild narrowing in intrahepatic IVC. A computed Tomography scan showed a Grade III-defined isodense lesion noted in segment II of live measuring 1.0×1×0.6cm (CC×AP×T rans) (Figure 1). On contrast administration, the lesion is showing moderate heterogenous enhancement on the arterial phase and no washout in the venous phase. A gross amount of free fluid noted in the

abdomen and pelvis is also seen. Hence, she was diagnosed with Budd chairi syndrome and referred to a direct intrahepatic portosystemic shunt she underwent the procedure. Medications prescribed were lasilactone 50mg 1 tab OD, apixaban 5mg BD, Magnex forte 1.5g BD and Udiliv 300 BD. Post procedure her liver function tests were normal and appeared to be asymptomatic.



**Figure 1: CT SCAN: Isodense lesion in segment -II of liver**

### **DISCUSSION**

Hypercoagulable state is the complication of Budd-Chiari syndrome, which develops in 80% of cases. The main factor for hepatic vein blockage is thrombosis. Vascular Compression results from an infection or a lesion that occupies space in the liver, forming hepatic cysts, adenomas, cystadenomas, invasive aspergillosis, and aortic aneurysm seen in these patients. [5] Twenty percent of the cases are idiopathic. About 20% cause are by use of oral contraceptives and pregnancy, which can cause a hypercoagulable state. [6] Blockade of hepatic veins lead to an increase in sinusoidal pressure reducing sinusoidal blood flow. Sinusoidal dilation and increased lymphatic drainage causing congestion of the liver, abdominal pain and ascites resulting in hypoxia of hepatocytes, increased portal pressure, and decreased hepatic perfusion. Diagnosis is based on ascites, hepatomegaly, increase serum bilirubin levels and decrease in serum proteins moderately, Ultrasonography, CT SCAN. Anticoagulation therapy, angioplasty, TIPS, and liver transplantation are recommended for the management of Budd-Chiari syndrome. [7] In the case of whole hepatic venous thrombosis, TIPS is sometimes not technically feasible so, Direct Intrahepatic Portosystemic Shunt (DIPS) with access to the portal vein from the IVC is a potential approach. [8]

### **CONCLUSION**

Budd chairi syndrome is a rare and life-threatening disorder with complications of hepatic encephalopathy, variceal haemorrhage, complete blockage of hepatic veins and death. In this patient isodense lesion is seen in the segment II of the liver characterized by Caudate lobe hypertrophy and mild narrowing in intrahepatic IVC which leads to ascites and Budd Chiari syndrome. When left untreated can be life threatening but can be managed by Medications and surgical intervention using procedures such as DIPS, TIPS and Liver transplantation.

### **ABBREVIATIONS**

DIPS: Direct intrahepatic portosystemic shunt; BCS: Budd-Chiari syndrome; AFB: Acid fast bacteria; TLC: Total leukocyte count; INR: International normalized ratio; IVC: Inferior vena cava; CC:AP:TRANS: Craniocaudal, Anteroposterior, Transverse; OD: Once daily; BD: twice daily, CT SCAN: Computed tomography; TIPS: Trans jugular intrahepatic portosystemic shunt

### **INFORMED CONSENT**

Informed consent form from the patient was taken.

### **COMPETING INTEREST**

The authors have declared that no competing interest exists.

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