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Case Report **Biotechonology**

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AN INTERESTING CASE OF SUBACUTE BUDD-CHIARI SYNDROME

DR. SAKTHI SELVA KUMAR*[1], DR. VINOTH KUMAR[2], DR. BALAKISHNAN[3] AND DR.N.N.ANAND[3]

[1]Post graduate in Internal Medicine
[2]Assistant professor of Medical Gastro-enterology
[3]Professor of Internal Medicine Sree Balaji Medical College and Hospitals, Bharath University, Chromepet, India

ABSTRACT

The Budd Chiari syndrome is a heterogeneous group of disorders characterized by hepatic venous outflow obstruction at the level of the hepatic venules, the large hepatic veins, the inferior vena cava, or the right atrium[1]. Hepatic veno-occlusive disease refers to obstruction of hepatic venous outflow at the level of the central or sub lobular hepatic veins, or both We present a 21 year old female with no previous history who presented with sudden onset of abdominal pain and massive ascites. Ultrasound abdomen & Doppler study and finally an IVC venogram was done which showed occlusion in the hepatic veins & Inferior vena cava and was hence diagnosed to have Sub-acute Budd Chiari syndrome

KEYWORDS: **Budd Chiari syndrome, ascites, Hepatic veno-occlusive disease.**

DR. SAKTHI SELVA KUMAR
Post graduate in Internal Medicine

***Corresponding author**

INTRODUCTION

Budd-Chiari syndrome is obstruction of hepatic venous outflow that originates anywhere from the small hepatic veins inside the liver to the inferior vena cava and the right atrium. Manifestations range from no symptoms to fulminant liver failure. Diagnosis is based on ultrasonography. Treatment includes supportive medical therapy and measures to establish and maintain venous patency, such as thrombolysis, decompression with shunts, and long-term anticoagulation.

CASE DISCUSSION

A 21 year old female, a student presented with an acute onset of abdominal pain and abdominal distension of 15 days duration. No high colored urine, no hematemesis and no pruritus. Examination revealed: she was not icteric, not pale, no pedal edema, no scratch marks; vitals stable. Per abdomen: soft, fluid thrill positive (massive ascites ie more than 2 liters of free fluid), no hepato-splenomegaly and no dilated veins.

Laboratory investigations done

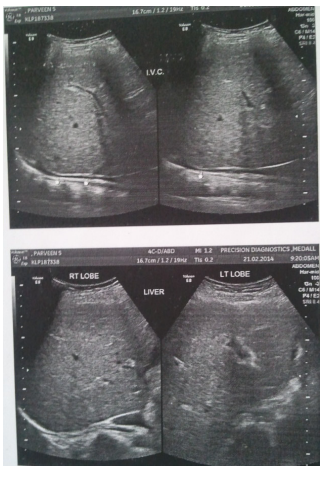
1) On 13/2/2014: hemoglobin 9g/dl, urea 26mg/dl, creatinine 0.7 mg/dl
Liver functions

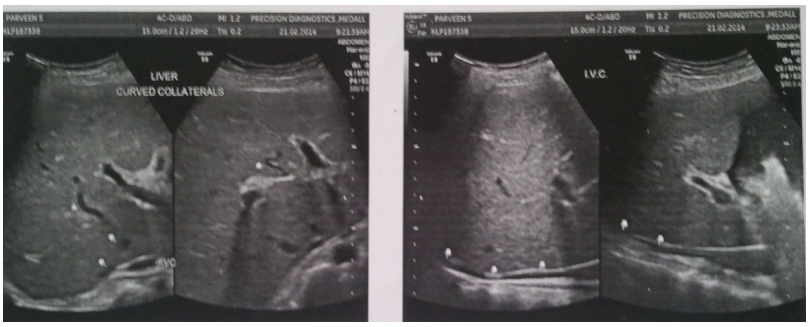
Total bilirubin	**0.6 mg/dl**
Direct bilirubin	0.3 mg/dl
Indirect bilirubin	0.3 mg/dl
SGOT	26 U/L
SGPT	20 U/L
ALP	191 U/L
Total protein	6.7 mg/dl
Serum albumin	3.8 mg/dl
Globulin	2.9 mg/dl
Prothrombin time(INR)	14.5

Ascitic fluid analysis revealed exudative type of fluid, ascitic protein 3.9 and Adenosine deaminase 21.

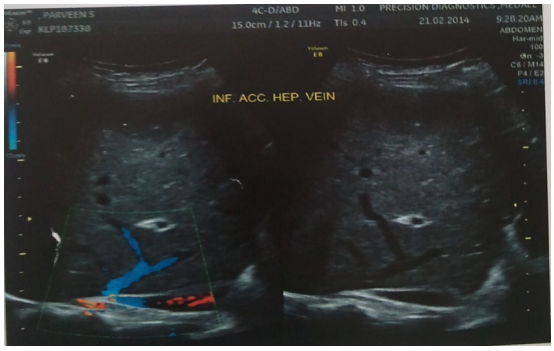
2) Ultrasound abdomen
(a) Gross ascites
(b) Liver parenchymal disease
(c) Splenomegaly

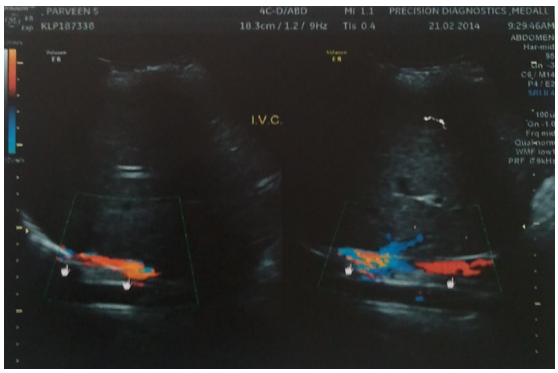
3) A week later repeat ultrasound with doppler study revealed:
4)

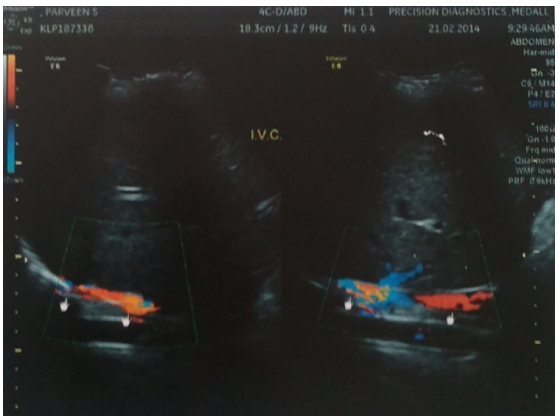


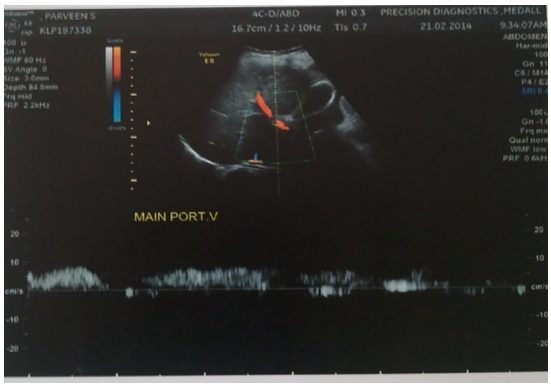


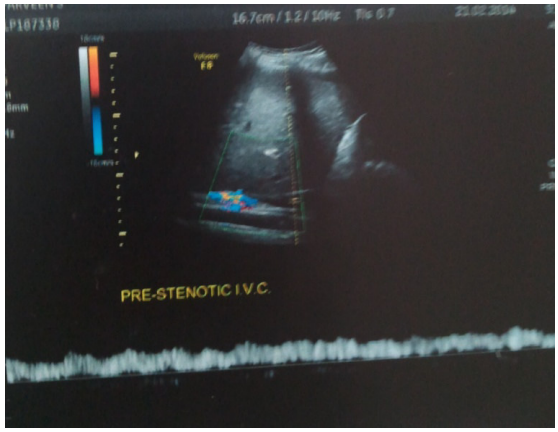
Doppler





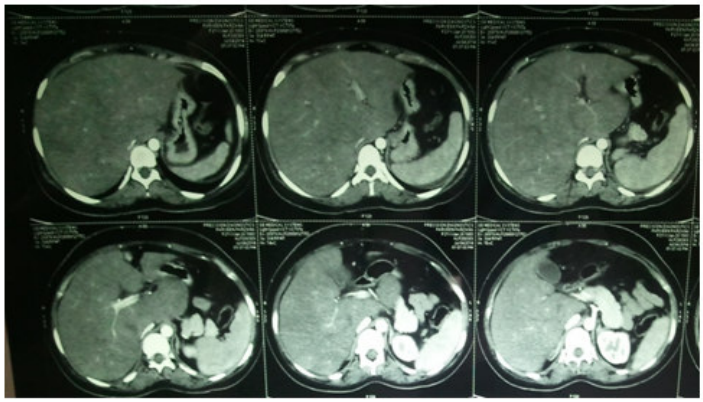




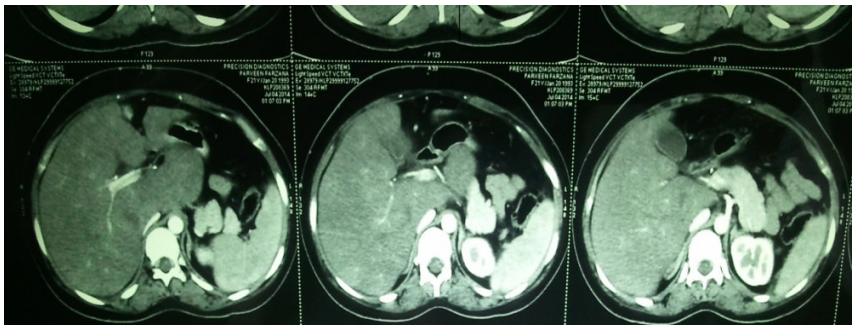


(a) Normal liver size & echo texture
(b) Distal hepatic segment of the IVC shows smooth narrowing but patent.
(c) All the hepatic veins are occluded in their distal segments and drain into the IVC through multiple collaterals and through an inferior accessory hepatic vein.
(d) Gross ascites.
(e) Main portal vein , splendid vein & superior mesenteric vein are normal sized & show normal flow pattern
(f) Spleen, gall bladder & both intra & extra hepatic bile tract are normal.
Features suggestive of acute Budd Chiari syndrome.

5) INR rose to 1.75
6) Upper Gastrointestinal endoscopy showed Grade 1 esophageal varices & antral erosions.
7) Multi slice CT scan of the abdomen with contrast:



1) **Gross Hepatomegaly**
2) **Splenomegaly**
3) **ascites**



(a) Hepatomegaly with diffuse heterogenous mottled enhancement (nutmeg liver). Significant hypertrophy of caudate lobe. Hepatic veins are not visualized on portal/hepatic venous phases- Budd Chiari syndrome
(b) Intrahepatic segment of IVC appears mildly compressed by enlargement of the liver.
(c) Small enhancing portal vein collateral in subcasular region of segment Iva extending towards the anterior abdominal wall.
(d) Splenomegaly
(e) Gross ascites

She was evaluated for hyper coagulable states

1)	Vitamin B12	193 pg/ml(normal)
2)	Folic acid	3.25 ng/ml (normal)
3)	Protein C	75 % activity(normal)
4)	Protein S	121% activity(normal)
5)	Anti phospholipid IgM	1.16 U/ml(normal)
6)	Anti phospholipid IgG	1.93 U/ml (normal)

Treatment given
Once Budd Chiari syndrome was suspected she was started on Heparin and Vitamin K antagonists and heparin was stopped after her INR rose to acceptable levels and was continued on vitamin K antagonist.

She was later subjected to an IVC venogram
- Imaging: right hepatic vein occluded with no flow in its entire length. Middle hepatic vein occluded in distal segment. LHV patent up to ostium. Flow could not be picked up across the confluence into IVC. Retro hepatic segment of IVC narrowed or occluded.
- Angiogram: though retro hepatic seg of IVC was narrowed-no occlusion/collaterals were noted. Pressure measurements across the narrowing was normal. Hepatic veins could not be cannulated from IVC. Left hepatic venogram was performed – abrupt cut off at the level of HV-IVC confluence with filling up of collaterals.

She was diagnosed with Sub acute Budd Chiari syndrome and referred to a surgical gastro-enterologist for Porto systemic shunt surgery.

DISCUSSION

Obstruction of the hepatic venous outflow tract results in increased hepatic sinusoidal Pressure and portal hypertension. In the early stages, portal venous perfusion of the liver is decreased, which may result in portal venous thrombosis[2]. The ensuing venous stasis and congestion lead to hypoxic damage to adjacent hepatic parenchymal cells. Furthermore, the ischemic injury to the sinusoidal lining cells results in the release of free radicals, and oxidative injury to the hepatocytes ensues[3].These mechanisms culminate in the

development of hepatocyte necrosis in the centrilobular regions, with progressive centrilobular fibrosis, nodular regenerative hyperplasia, and ultimately, cirrhosis of the liver[4]. However, if the hepatic sinusoidal pressure is reduced by the creation of a Porto-systemic shunt or by the development of a portal venous collateral system, liver function improves[5]. Factors that confer a predisposition to the development of the Budd–Chiari syndrome, including hypercoagulable states[6], both hereditary and acquired, and a variety of other causes can be identified in about 75 percent of patients.

Hypercoagulable states like

1) Polycythemia vera
2) Essential thrombocythemia and
3) Myelofibrosis
4) Paroxysmal nocturnal hemoglobinuria
5) Antiphospholipid syndrome[9]
6) And inherited deficiencies of protein C, protein S, antithrombin III.
7) Factor V Leiden mutation
8) Prothrombin-gene mutation
9) Women who use oral contraceptives, during pregnancy and in the immediate postpartum period.

Clinical manifestations

The clinical presentation of the Budd–Chiari syndrome[7, 9] depends on the extent and rapidity of hepatic- vein occlusion and on whether a venous collateral circulation has developed to decompress the liver sinusoids. The syndrome can be classified as

1. Fulminant
2. Acute
3. Sub acute
4. Chronic.

Patients with the fulminant form of the syndrome present with hepatic encephalopathy within eight weeks after the development of jaundice. This presentation is uncommon. Patients with the *acute* syndrome have symptoms of short duration, intractable ascites, and hepatic necrosis *without the formation of venous collaterals*. The *sub acute* form, which is the most common, has a more insidious onset; ascites and hepatic necrosis may be minimal, because the hepatic sinusoids have been decompressed by a portal and hepatic venous collateral circulation.

Clinical manifestations depend on the following factors

1. Rapidity of onset
2. Site of occlusion
3. Formation of collaterals

Based on site of occlusion Budd Chiari syndrome can be classified as that involving

1. Small hepatic veins
2. Large hepatic veins
3. IVC
4. Combined (large hepatic veins plus IVC)

Management

Diagnosis

1. Ascitic fluid analysis(a high SAAG ratio)
2. Ultrasound Abdomen and portal Doppler study

3. CT with contrast or MRI abdomen
4. IVC venography

Treatment

1. Anticoagulation
2. Angioplasty & stenting
3. TIPS (transjugular intrahepatic Porto systemic stent shunt)
4. Surgical shunt
5. Liver transplant

Our patient had a short history of about 15 days of symptoms with a high SAAG (serum ascites albumin ratio) ratio in ascitic fluid with occlusion of flow through the hepatic veins and IVC with developed venous collaterals as witnessed in the Doppler study & venogram, hence our patient had sub-acute Budd Chiari Syndrome of the combined type based on site of occlusion. We screened her for hypercoagulable states but couldn't find a specific cause and hence the etiology is idiopathic. IVC gram was done to delineate the site & extent of occlusion further intervention under radiographic guidance couldn't be done and hence the patient was referred for shunt surgery.

CONCLUSION

Budd Chiari syndrome should be suspected in a young patient with sudden onset of liver cell failure with no known previous ailments and should be evaluated for hypercoagulable states. This is imperative, for the patient can deteriorate very quickly if not diagnosed early and treated accordingly and the condition can be completely reversed if done so at the right time. Hence our reasoning for presenting this case report.

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