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CASE REPORTS

"Budd Chiari Syndrome"- A Case Report

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Abstract:

"Budd Chiari syndrome (BCS)" is an uncommon condition induced by thrombotic or nonthrombotic obstruction to hepatic venous outflow. This disease can be potentially life-threatening but prognosis is more favorable in patient with IVC webs but is extremely poor in malignant and haematological cases. A cooperative collaboration of hepatologist and cardiologist can make the situation easy for diagnosis, even in treatment of some cases.

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Introduction:

Budd-Chiari syndrome (BCS) is a manifestation of hepatic venous outflow obstruction that was first described by Budd in 1845 and then expounded upon by Chiari, who presented 13 cases in 1899. The hepatic outflow obstruction usually occurs at the level of the inferior vena cava (IVC); the hepatic veins; and, depending on the classification and nomenclature, possibly at the venule level. Overall incidence of BCS is unknown.

Case Report:

A 29 yrs old man presented with abdominal swelling, occasional leg and scrotal swelling for last 15 months. He also developed yellow coloration of sclera and right upper abdominal pain for last 12 months. Physical examination demonstrated hepatic facies, mild jaundice, spider naevi on chest, gynaecomastia, ankle edema. Abdominal examination revealed engorgement of the superficial vessels of the anterior abdominal and chest wall with flow towards the superior venacava, Hepatomegaly with left lobe more enlargements, which was firm, mild tender. Also revealed splenomegaly, huge ascites, and scrotal edema, with soft nontender testis.

Laboratory studies demonstrated blood Hb level of 10 g/dl, ESR-15 mm in 1st hour, CBC-normal, S. albumin- 36g/L, S. bilirubin- 2.6mg/dl, SGPT-16u/L, PT-16sec, S.creatinine-98 micro mol/L, HBsAg and Anti HCV-negative, Ascitic fluid studies revealed protein 47g/L, Albumin-24g/L, AFB and Gm-stain-negative. BT, CT & PT were normal. Ultrasonogram of whole abdomen showed marked Hepatosplenomegaly with enlargement of the caudate lobe of the liver and huge ascites.

Doppler Ultrasonogram of HBS showed dilated inferior venacava with partial occlusion by thrombus, moderate portal hypertension, no evidence of intrahepatic venous obstruction. MRI of abdomen showed total occlusion in proximal inferior venacava with possible intraluminal thrombus in proximal Inferior Venacava (IVC), hepatomegaly, huge splenomegaly and ascites.

Upper GIT endoscopy showed grade 1 esophageal varises. Venography revealed right heart catheter could not be passed beyond the level of hepatic vein, complete obstruction in inferior venacava just 1 cm below the diaphragm. Superior venogram showed few millimeter of IVC. There was no indentation of caudate lobe.

About 1 year back he was diagnosed as a case of cirrhosis of liver, but now finally after doing venography he is diagnosed as a case of "Budd Chiari Syndrome" due to membranous obstruction of the Inferior Venacava (IVC) with Cirrhosis of Liver.

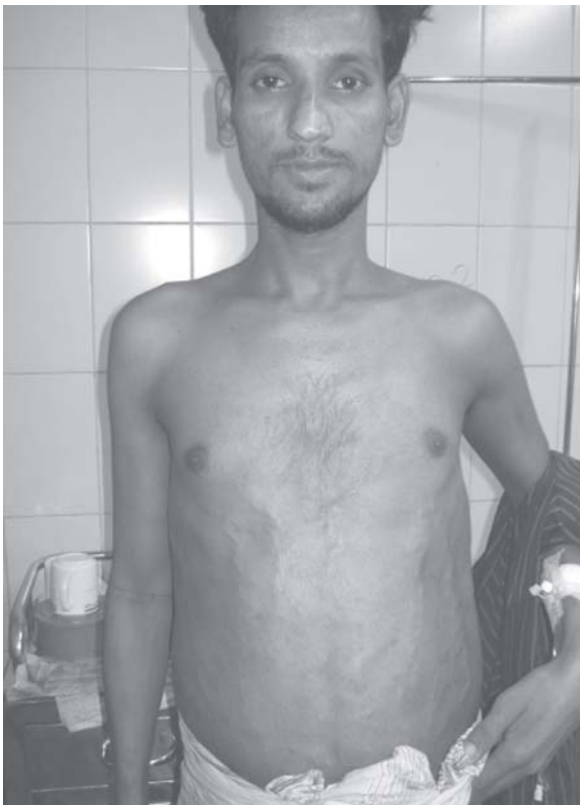

Fig.-1: *Patient of BCS.*

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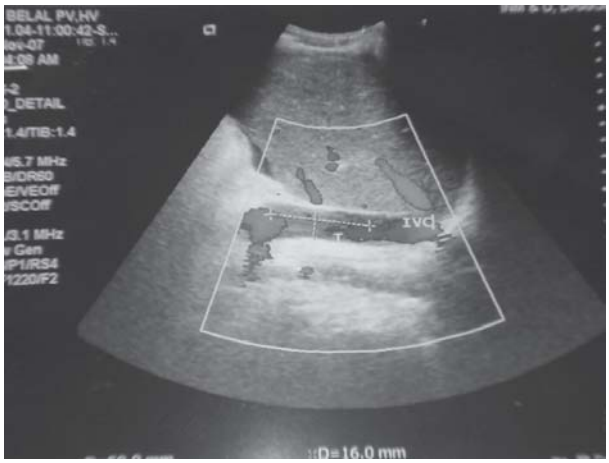

Fig.-2: *Dilated IVC, partial occlusion with thrombus.*

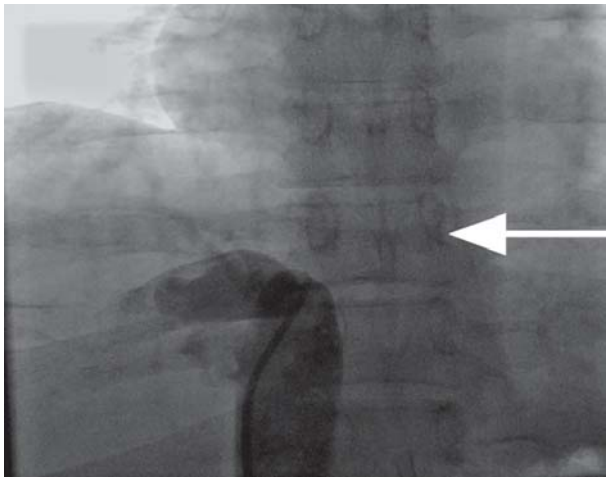

Fig.3: *Venography of IVC & Hepatic vein shows dilated hepatic vein with total cut in proximal IVC.*

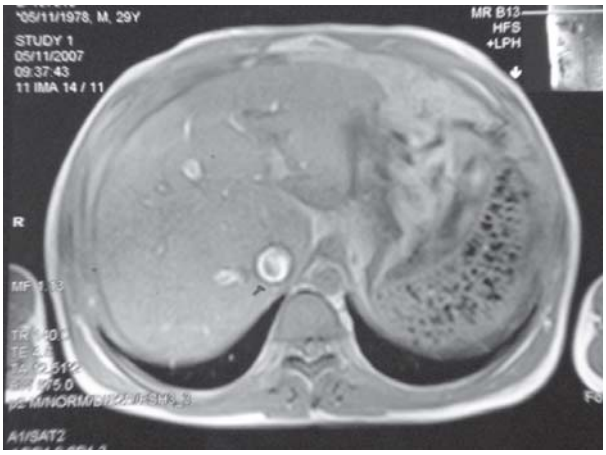

Fig.-4: *Thrombus in proximal IVC.*

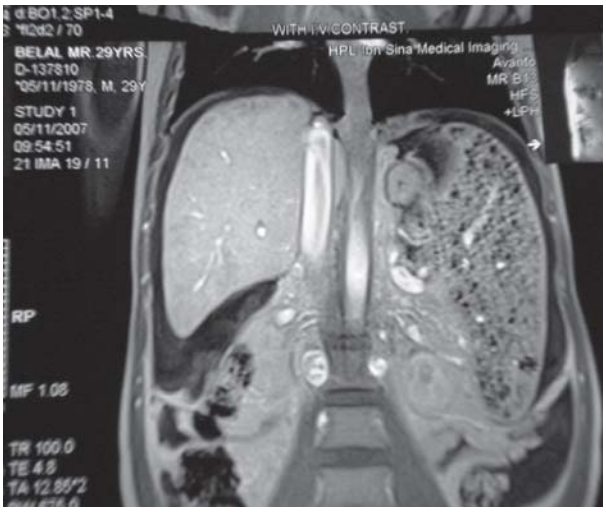

Fig.-5 : *IVC is total cut just after diaphragm.*

Discussion:

"Budd Chiari Syndrome" (BCS) is a disease of hepatic circulation caused by occlusion of the hepatic vein or IVC. The syndrome comprises classical triad of abdominal pain, ascites and hepatomegaly [1]. The syndrome may be primary, due to membrane in the proximal IVC or secondary causes. In practice, no identifiable cause is found in up to 70% of patients [2]. The principal cause of secondary BCS is thrombosis caused by a thrombophlic disorder in 75% cases. Primary myeloproliferative disorders are most common, but all genetically based prothrombotic tendencies (eg. Protin S, C, Factor 5 Leiden deficiencies or JAK2 mutation) can causes [3]. An important non genetic risk factor is the use of ostrogen-containing (combined) form of hormonal contraception. Other risk factors include the antiphospholipid syndrome, paroxysmal nocturnal haemoglobinuria, aspergillosis, Behcet's disease, dacarbazine, pregnancy, trauma. Another thrombotic state is in malignant conditions (eg. Adrenal, renal carcinoma, hepatocellular carcinoma, angiosarcoma, leimyosarcoma of hepatic vein) [4].

Obstruction to hepatic veins occurs at any site from the efferent vein of the acinus to the entry of the IVC in to the right atrium. Membranous obstruction of the IVC at the suprahepatic level is a rare but debilitating clinical problem [5]. According to presentation and treatment purpose BCS can be classified as fulminant, acute, subacute or chronic [5]. These depend on the speed of occlusion and the extent of the hepatic venous obstruction. In fulminant or acute cases necrosis spreading out from the central vein with massive ischemic necrosis. In chronic cases, the caudate lobe is enlarged and compresses the IVC as it passes posterior to the liver [5,6]. Chronic obstruction causes progressive increase portal vein and hepatic sinusoidal pressure as blood flow stagnated. This causes increased filtration of vascular fluid with the formation of protein

rich ascitic fluid. Flow through alternative veins leading to gastric varices, hemorrhoids and engorged vessels in abdominal wall. Spleen is also enlarged. Obstructed vein wall is thickened and there may be some recanalization with replaced by a fibrous strand or fibrous web. Obstruction also causes hepatic centrilobular necrosis, fibrosis due to ischemia. Portal vein involvement causes a veno portal cirrhosis, which happened in this patient. Unaffected areas of liver form nodular regeneration [5,6].

The fulminant or acute syndrome presented with rapidly progressive severe upper abdominal pain, mild jaundice, hepatomegaly, vomiting, ascites, elevated liver enzymes, encephalopathy and usually die with in 2-3 wks. In case of total hepatic vein obstruction, delirium, coma, hepato-cellular failure and death with in few days occurs [6,7].

In chronic cases patient presents with enlarged tender and palpable caudate lobe of the liver, ascites, mild or no jaundice, splenomegaly. Patient may progress to cirrhosis and show the sign of liver failure. An asymptomatic form is also recognized [6,7].

Initially diagnosis should be suspected from clinical background and an ascitic fluid tap with a high protein content, like this patient. Some supportive biochemical evidences like, serum bilirubin with in 2-3 mg/dl, slight rise in serum alkaline phosphatase level, reduce in serum albumin level in chronic cases but no features of hepatitis [6,7].

For diagnosis Ultrasonography with Doppler study is very important. Ultrasonogram may show hepatosplenomegaly, caudate lobe enlargement, ascites, hepatic parenchymal inhomogenesity(in chronic case), obliteration of hepatic vein, stenosis, spiderweb vessels, large collateral vessels or a hyperechoic cord replacing a normal vein. Intraluminal clot may be seen in the acute phase [6,7], but in chronic cases it can be occur due to long time stasis, like in this case. Doppler ultrasonography is usually sufficient to confirm the diagnosis, which shows altered, absent, reversed, turbulent direction of flow in the hepatic vein and retro hepatic IVC. Finding in the chronic phase include nonvisualiuzation of occluded vein and formation of collateral veins with in the liver or from the liver to either the left renal vein or the pericardiophrenic vein [8]. Portal vein is also thrombosed in 10% of patient and IVC in 20% cases, like in this patient.

Hepatic venography and Inferior venacavography are useful diagnostic tool. Hepatic venography may not be possible or may show narrow occluded veins. Typical spiderweb appearance in hepatic venography which is due to prominent collaterals. In inf.venocavography indentation (narrowing) may show due to enlarged caudate lobe[8]. Caval pressure measurement is needed. CT scan and MRI is sometimes employed although these methods are not as sensitive. But inthis patient, there was thrombosis in proximal IVC with total cut in proximal IVC in MRI.

Liver biopsy is nonspecific but it often shows diagnostic features of centrilobular necrosis and sinusoidal congestion, fibrosis in chronic cases. It is also used to exclude other diagnosis [9].

Treatment should be started with anticoagulation, especially in case of underlying hematological disorder as the cause of the BCS.

Long term anticoagulation is often needed. Surgical shunting is an alternative like's, heart-meso-atrial shunt or TIPS or side-to side portacaval shunt is the procedure of choice. But surgical shunt should not be used in acute cases. A 95% 5 years survival can be achieved in chronic cases. Liver transplantation is the preferred option where shunting is not possible or in fulminant cases. Where isolated webs are found in hepatic vein or in IVC, ballon dilatation, transcardiac membranotomy or percutaneous stent placement can be done by cardiologist [9,10,11] .

Conclusion:

Budd Chiari syndrome is a rare and confusing disease of the hepatic circulation. Sometimes it is misdiagnosed as a case of only chronic liver disease but a good venography can remove all kind of confusion like this patient.

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