BUDD CHIARI SYNDROME

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INTRODUCTION

Budd Chiari Syndrome (BCS) is an uncommon condition first described by Budd and then expounded upon by Chiari. In this syndrome there is hepatic venous outflow obstruction, which usually occurs at the level of inferior vena cava, the hepatic veins and depending on the classification and nomenclature, possibly at the venule level. There are two types of BCS: acute and chronic [1]. The acute form results from an acute thrombosis of the main hepatic veins or the inferior vena cava (IVC). The chronic form is related to fibrosis of the intrahepatic veins, presumably related to inflammation. The classical presentation is with ascites, hepatomegaly and abdominal pain.

We present a case of Acute Budd Chiari syndrome in a young patient with classical symptoms and signs reporting in our hospital.

CASE REPORT

A 22 years old male labourer, presented in medical OPD with complaints of generalized abdominal pain of two weeks duration. The pain was more marked in the epigastric and right hypochondrial region. Along with pain the patient also noticed distention of abdomen. On examination of the patient, he was afebrile but was in distress due to pain. The abdomen was distended and tender. Abdominal wall veins were prominent, fluid thrill was positive indicating ascites. Liver was palpable 6 cm below right costal margin. No jaundice was seen clinically. The patient was admitted in medical intensive therapy centre (ITC) and immediate Laboratory and radiological investigations were advised.

His LFT's were deranged with raised bilirubin, alkaline phosphatase and alanine transaminase (ALT). Prothrombin time was 25 seconds. Platelets count was within normal limits. Ascitic fluid examination showed raised proteins. Mantoux test was negative.

Ultrasound examination revealed hepatosplenomegaly with gross ascites. On Color Doppler evaluation of IVC and hepatic veins, there was a large echogenic thrombus of post hepatic part of IVC occupying and almost completely obliterating it with no evidence of Doppler flow in the region of IVC thrombus (fig.1). IVC proximal to the thrombus was collapsed. Distal IVC showed slow flow on Doppler examination. Intrahepatic veins and their drainage confluence could not be visualized.

CT scan examination was carried out subsequently, in the CT scan department. There was a large homogenous and hypodense thrombus in the post hepatic IVC almost completely filling it with failure of visualization of intrahepatic veins. On post Intravenous contrast examination, there was generalized non enhancing hepatic parenchyma with patchy enhancement of caudate lobe in comparison to well enhanced splenic parenchyma. On the basis of these clinical and radiological findings, diagnosis of Acute Budd Chiari Syndrome was made (fig.2).

Patient was placed on antibiotic therapy, anticoagulants including tablet warfarin 5 mg and injection claxan with pain killer injections. Ascitic fluid tapping was done on regular intervals, enough to reduce the distress of the patient.

Patient remained in the hospital ITC for approximately 10 days, but showed no clinical improvement with deranged LFTs and coagulation profile.

Other Laboratory investigations estimation of Protein C, Protein S, antithrombin III, Anticardiolipin antibodies could not be carried out due to lack of availability of these investigations, hence exact cause leading to this syndrome could not be determined.

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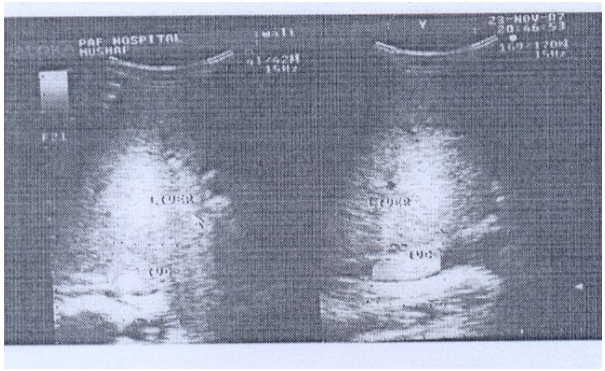

Fig. 1: Color doppler ultrasonography thrombus

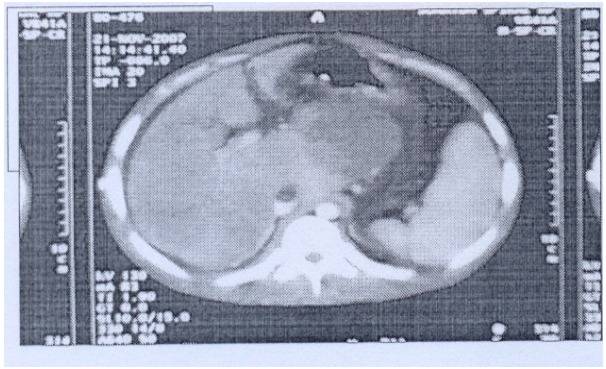

Fig. 2 Budd shiari syndrome patient with thromb us and generalized reduction in hepatic parenchymal echogenicity on post contrast enhanced CT

Prothrombin time of the patient progressed to 59 seconds from 25 seconds. The patient was later on referred to a tertiary care hospital in Lahore for further expert opinion and possible surgical intervention.

DISCUSSION

Budd Chiari syndrome has mainly three types, in type I: there is occlusion of IVC with or without the involvement of hepatic veins, in type II: there is occlusion of major hepatic veins with or without the involvement of IVC, in type III: there is occlusion of small centrilobar veins. Main causes are idiopathic (50-65%), Thrombotic seen in hypercoagulable state [2], Injury to vessel wall and Non thrombotic obstruction.

Patients of acute Budd Chiari syndrome present with rapid onset of abdominal pain, intractable ascites and hepatomegaly. Chronic Budd Chiari syndrome patients usually present with insidious onset of jaundice, intractable ascites and variceal bleeding. A high index of suspicion is needed to confirm the diagnosis.

Radiological evaluation plays a vital role in early and prompt diagnosis of the syndrome. Ultrasound, color Doppler imaging, CT scan, MRI and Nuclear imaging can be performed for this purpose.

The radiological examination play very important role in the diagnosis and management. Common radiological tecniques used are CT scan, magnetic resonance imaging, ultrasonography and color doppler imaging (which was useful in this case also)

Ultrasonography is noninvasive and has high sensitivity and specificity. Conventional US may show gallbladder wall thickening, ascites, patchy liver echo pattern, splenomegaly, hypertrophied caudate lobe, and ascites. Color-flow Doppler (CFD) images demonstrate abnormality of the anatomy or

flow [4]. Visualization of thrombus or tumor or echogenic membrane in IVC.

Nuclear immaging in BCS, 99mTc sulfur colloid uptake is increased (i.e., hot) in the caudate lobe at the expense of the rest of the liver, in which uptake may be normal, reduced, absent, or patchy.

Treatment options for Budd Chiari Syndrome are both medical and surgical. Biopsy analysis of a specimen from the liver may be required in some patients for histological confirmation. Optimum treatment or palliation of BCS depends on the cause [5].

The Transjugular intrahepatic portosystemic shuuts (TIPS) procedure may benefit selected patients [6]. Liver transplantation may be considered for advanced cases. Although most of the procedures are surgical they do require accurate radiological assessment of the IVC, liver, portal vein, and hepatic veins prior to intervention.

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