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Research Article

HEPATIC VENOUS OUTFLOW TRACT OBSTRUCTION ARE WE SEEING A CHANGE IN SPECTRUM; EAST TO WEST

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A B S T R A C T

Background and Aim: Hepatic Venous Outflow Tract Obstruction (HVOTO) is defined as an obstruction predominantly of venous process (thrombosis or phlebitis) or compression or invasion of the hepatic veins and/or the inferior vena cava, independent of the level or mechanism of obstruction, provided the obstruction is not due to cardiac disease, pericardial disease, or sinusoidal obstruction syndrome (veno-occlusive disease).The clinical features and etiology of Budd–Chiari syndrome (BCS) vary from region to region. The aim was to study its presentation at our Center.**Methods and Results:** 30 consecutive cases of BCS were evaluated during the period from April 2018 till October 2018 at Madras Medical College, Chennai. The diagnosis was made on the basis of MR venography and Doppler ultrasonography. The study population was made up of 16 females (mean age, 37.4 years) and 14 males (mean age, 46.5 years). Nine patients presented with acute disease, while 21 patients had a subacute presentation. Abdominal pain, jaundice, upper gastrointestinal bleed and tender hepatomegaly was the commonest presentation. Ascites and pedal edemawere seen only in 53% of the cases. BCS was caused by a hepatic venous obstruction in 63%, inferior vena cava in 6% and combined obstruction in 31%. Doppler ultrasound accurately detected the site of bock in 24 of 30 patients (80%). Etiologically, five had APLA syndrome, five secondary to extrinsic compression, three had protein C and protein S deficiency, three due to pregnancy and eleven (37%) was idiopathic.**Conclusion:** Hepatic venous outflow obstruction is a common problem; abdominal pain, tender hepatomegaly and UGI bleed was the common presentation, ascites was seen only in ½ (53%) of the cases showing a changing spectrum compared to various studies. Sub-acute presentation is more frequent and females are at an increased risk. Idiopathic and inherited/acquired hypercoagulable states were the most common etiology.

INTRODUCTION

Budd-Chiari syndrome (BCS) refers to hepatic venous outflow tract obstruction (HVOTO) starting from the level of small hepatic veins (HV) through large HV and inferior vena cava (IVC) to the junction of the IVC and right atrium[1-3]. This includes both hepatic vein thrombosis and IVC thrombosis or obliterative hepatocavopathy[4, 5]. There is increasing evidence that Budd Chiari syndrome occurs when acquired predisposing factor(s) affect a susceptible individual and geographical variations in disease pattern exist. While earlier studies from India reported isolated inferior vena cava (IVC) obstruction as the commonest disease type, this is a minority in more recent reports where a combination of IVC and hepatic vein obstruction is the commonest type.

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Longer duration of illness has been shown to be associated with IVC obstruction, and the recent change in disease profile in India may reflect an earlier diagnosis of Budd Chiari syndrome. Poverty, malnutrition, recurrent bacterial infections and filariasis have been previously suggested as a predisposing factor for IVC obstruction. Obstruction of the IVC, either thrombotic or nonthrombotic, was considered to be a major cause of BCS in Asia[5]. Two-thirds of IVC obstruction cases leading to BCS were due to membranous obstruction[6]. Dattaet alreported 40 cases of membranous obstruction of IVC (MOVC) while Victor *et al* [6] reported 17 cases of membranous obstruction. Though MOVC and membranous obstruction of HV was initially thought to be congenital, Okuda demonstrated these lesions to be an after-effect of thrombosis in the IVC or HV which were organised over a period of time[5].Over the years, many prothromboticstates leading to BCS have been described in India. A few differences in etiologies were noticed between India and the West i.e. peripartum occurrence of BCS was common in India

while oral contraceptive use was commonly implicated in the West. The majority of patients with BCS present with a chronic course, while only a small number of patients present with acute or fulminant forms. BCS is mostly encountered in the adult population and considered uncommon in children; when seen in children, the clinical presentation is similar[7]. Anti-thrombotic drugs and anticoagulants form the mainstay in the treatment of acute and chronic BCS. Percutaneous balloon angioplasty for membranous obstruction of the IVC and hepatic vein has also been used successfully in the treatment of BCS. Although TIPS has gained popularity in the treatment of BCS, it is rarely been performed in India[8].

METHODS AND RESULTS

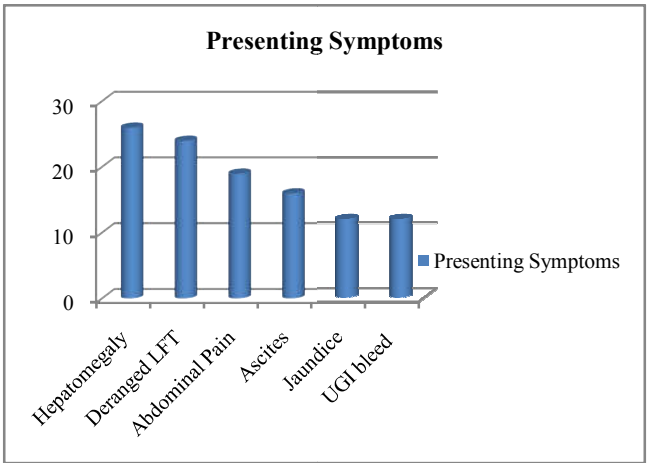
This was a cross-sectional study conducted on patients at Rajiv Gandhi Government General Hospital, Madras Medical College, from june 2107/december2017 to April 2018-October 2018.. This study was approved by the institutional review board. The study population consisted of consecutive patients diagnosed with Budd Chiari syndrome presenting with jaundice, hepatomegaly and ascites. Themedical chart of all patients was analyzed. Information regarding patient demographics, presentation, treatment efficacy and complications was assessed. All information was coded using SPSS software (version 22.0).Acute Budd Chiari was defined assymptom presentation less than <8 weeks duration, Sub-acute with symptom presentation of more than 3 months duration and chronic as those patients presenting with chronic liver disease.

The diagnosis was made on the basis of Doppler ultrasonography and confirmed by MR (Magnetic Resonance) venography in all the study patients. The study population was made up of 16 females (mean age, 37.4 years) and 14 males (mean age, 46.5 years). Nine patients presented with acute disease, while 21 patients had a subacute presentation. Abdominal pain, jaundice, upper gastrointestinal bleed and tender hepatomegaly was the commonest presentation. Ascites and pedal edemawere seen only in 53% of the cases. BCS was caused by a hepatic venous obstruction in 63%, inferior vena cava in 6% and combined obstruction in 31%. Doppler ultrasound accurately detected the site of bock in 24 of 30 patients (80%). Etiologically, five had APLA syndrome, five secondary to extrinsic compression (Hepatocellular carcinoma-3, Liver abscess-1, Hydatid cyst-1), three had protein C and protein S deficiency, three due to pregnancy and eleven (37%) was idiopathic. Only two patients in our study were diagnosed with underlying Myeloproliferative disorder with JAK2 mutations been positive in both these patientsshowing a different trend in etiologically spectrum to what is seen in the West.

Table 1 Patient Characteristics

Factors	N	%
Gender		
Male	14	46%
Female	16	54%
Age		
Median, range	42, 27-63	
Primary/ Secondary Etiology		
APLA	5	17%
Protein C and S deficiency	3	10%
Factor V, Prothrombin II gene mutation	1	3%
Extrinsic compression	5	17%
a) HCC	3	
b) Liver abscess/ Hydatid	2	
Pregnancy	3	10%
Myeloproliferative Disorders	2	6%
Idiopathic	11	37%
Stenosis Location		
Hepatic vein thrombosis	19	63%
Hepatic and IVC obstruction	9	31%
IVC obstruction	2	6%
Diagnosis		
MR venography	22[23]	96%
Doppler Ultrasound	24	80%

APLA- Anti Phospholipid Antibody Syndrome, MR- Magnetic resonance, HCC- Hepatocellular carcinoma



LFT- liver function test, UGI- upper gastrointestinal bleed

DISCUSSION

Our study is distinct from previous Indian reports in many aspects. It showed a predominance of HV thrombosis and identification of etiologies, the majority being hypercoagulable states. In our series, HV thrombosis represented the majority of cases (63%), which was followed in decreasing order by combined IVC/ HV thrombosis (31%), IVC thrombosis (6%). All previous Indian series, except two[9], describe the predominance of IVC up to 79.2%[10]. Isolated hepatic vein obstruction usually presents with ascites, hepatomegaly, jaundice and/oroesophagal/ gastric variceal bleeding. Difficult-to-control ascites and ascites out of proportion to pedal edema should raise clinical suspicion of hepatic vein obstruction. Abdominal pain is a prominent feature of acute presentation. Eliciting the abdominal-jugular reflux canbe employed to ascertain patency of hepatic venous outflow at the bedside. IVC obstruction in addition to the above presentation, manifests as distended veins over the abdominal wall (draining upwards), lower back and occasionally associated with stasis ulcers over the legs. Bilateral pitting pedal edema may be the sole presentation of chronic IVC obstruction. Infertility and varicocele are the other possible manifestations. Rarely, BCS may present for the first time as an acute liver failure with hepatic encephalopathy accompanying jaundice and ascites. Though BCS usually presents as symptomatic disease, the less commonly asymptomatic disease is detected on imaging done for some other indication[11].

Hepatic venous outflow obstruction is a common problem; abdominal pain, tender hepatomegaly and UGI bleed was the common presentation, ascites was seen only in ½ (53%) of the cases showing a changing spectrum compared to various studies. Sub-acute presentation is more frequent and females are at an increased risk. Idiopathic and inherited/acquired hypercoagulable states were the most common aetiology. MOVC was not seen among any of our patients probably due to early diagnosis and about 30% of patients presented with an acute presentation with severe right upper quadrant abdominal pain, jaundice, hepatomegalyand/or with minimal ascites. This

sudy also highlights acute presentation of BCS as a more common entity and warants prompt and clinical suspicion in patients with short duration of symptom(s) as well.

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