MEMBRANOUS OBSTRUCTION OF THE INFERIOR VENA CAVA OR HEPATIC VEIN WITH BUDD-CHIARI SYNDROME : A REPORT OF FOUR TURKISH CHILDREN

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SUMMARY : Membranous obstruction of the inferior vena cava and/or hepatic vein with Budd-Chiari syndrome is a rare condition in children. Four children with this lesion are described. Clinical features included hepatomegaly,splenomegaly,ascites and dilated superficial abdominal veins.Liver enzymes were slightly abnormal in most cases. Liver histology showed a wide range of changes, from almost normal to cirrhosis.Membranous web was demonstrated by angiography in all patients. The web was successfully dilated percutaneously by balloon catheters in two patients.

***Key Words:** Membranous Obstruction, Inferior Vena Cava, Hepatic Vein, Childhood, Budd-Chiari Syndrome.*

INTRODUCTION

Budd-Chiari syndrome (BCS), a syndrome of congestive hepatomegaly, portal hypertension, ascites and varying degrees of liver failure, results from occlusion of hepatic venous drainage channels.'Caval web' associated with varying degrees of thrombosis and obstruction affecting hepatic veins and the inferior vena cava is the major cause of the syndrome in the Orient and India (1). The lesion is thought to be of congenital origin (2-5) however, it has rarely been described in children.

PATIENTS AND METHODS

This paper reports four children with BCS due to membranous obstruction of the inferior vena cava (IVC) and/or hepatic veins. The children presented with abdominal distension, hepatosplenomegaly, dilated superficial abdominal veins, and hemathemesis.

Laboratory investigations included blood count, prothrombin time and determination of serum proteins, biluribin, transaminases, ceruloplasmin, serum and urinary copper, α antitrypsin, Hepatitis B surface antigen, hepatitis C antibody, protein C, S, antithrombin III, Factor V Leiden and prothrombin gene mutation. Ascitic fluid was analysed when present. Echocardiography was carried out to exclude pericardial disease. All patients had abdominal ultrasound and Doppler ultrasound. In all patients, the esophagus was examined endoscopically. Transfemoral inferior cavography was performed in all cases. In patients 1, 2 and 3, transhepatic angiography was also performed. Percutaneous liver biopsy was done by Menghini technique in all patients.

RESULTS

Clinical Presentation:

Four male children, ranged in age from 9 to14 years. Patients 1, 3, and 4 had an enlarged liver 3-6 cm below the costal margin, one had ascites and peripheral edema, and three had dilated superficial abdominal veins.

Laboratory studies: Routine laboratory tests (Table 1) revealed abnormal prothrombin time, albumin and transaminases in three patients. Hepatitis B surface antigen and anti hepatitis C antibody were negative in all patients. Serum ceruloplasmin, serum and urinary copper, α antitrypsin and sweat electrolyte levels were within normal ranges. Serum levels of antithrombin III, protein C, protein S were normal and antiphospholipid antibodies were negative. Factor V Leiden mutation and prothrombin gene mutation were absent.

Table 1 : Laboratory values of the patients.

Patients	Albumin(g/dl)	AST (IU/L)	ALT(IU/L)	Total biluribin(g/dl)	Prothrombin time (seconds)
1	2.5	65	70	0.8	44
2	2.8	44	36	0.9	47
3	4	86	88	0.4	52
4	3.7	51	47	0.7	39

Radiographic studies: Portal system duplex Doppler Ultrasonography demonstrated patent splenic and portal veins in all patients. There was an anastomosis between hepatic veins in patient 1 and 3. The caudate lobe was hypertrophic in all patients.

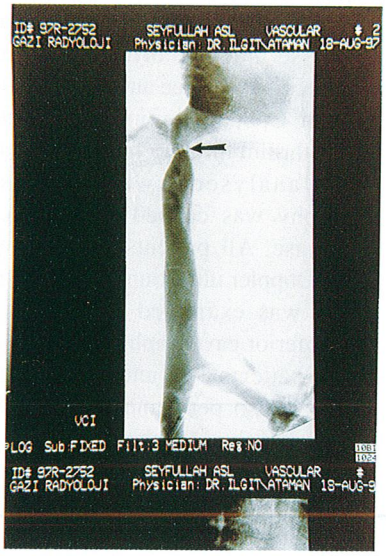

Fig - 1 : Patient 4. Inferior vena cavogram showed constriction of the hepatic IVC.

Inferior vena cavography and percutaneous hepatogram were carried out in patients 1, 2 and 3. Patency of IVC was demonstrated in patient 1 and 3. Inferior venacavogram showed a tapered constriction of the hepatic IVC in patient 4 (Fig. 1) and IVC could not be filled with dye in patient 2. Injection of dye into the intrahepatic veins showed venous drainage via the collateral veins in the abdominal wall in this patient (Fig. 2). Membranous web was demonstrated between the middle hepatic vein and IVC in patients 1 and 3 by percutaneus transhepatic angiography (Fig. 3).

Histology :

Cirrhosis was diagnosed at liver biopsy in patient 1, 2 and 3. Liver biopsy showed mild chronic congestion, central necrosis, dilated sinusoids and sparse portal inflammatory infiltrate in patient 4.

Endoscopic examination:

Endoscopic examination of esophagus revealed

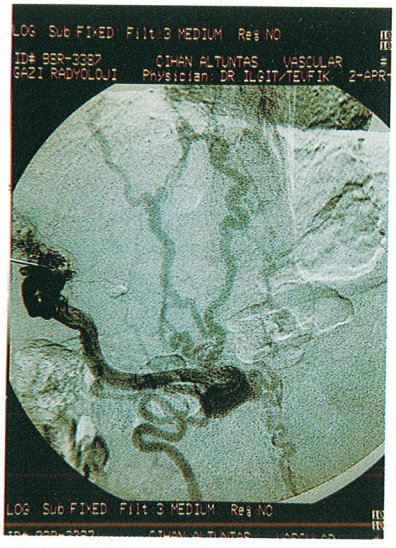

Fig - 2 : Patient 2. Injection of dye into the hepatic veins showed venous drainage via the collateral veins on the abdominal wall.

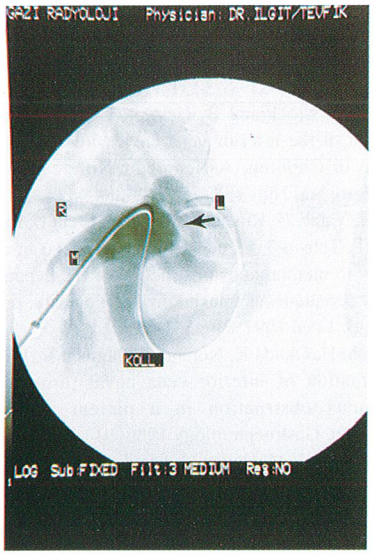

Fig - 3 : Patient 3. Transhepatic venogram documenting occlusion.

grade II and III varices in patients 1, 2 and 3.

DISCUSSION

The causes of BCS are myeloproliferative disorders, thrombotic conditions, malignant neoplasms, or membranous obstruction of the hepatic vein and inferior vena cava. The latter is particularly infrequent, except in the Orient (2, 4). The aetiology is controversial. As one group favours the theory of thrombosis and organization, others favour a congenital origin, based on non-union or malunion of veins during the development of hepatic veins and cava (6, 7). In our cases, clinical presentation in a young age group and exclusion of potential causes of thrombosis imply that the origin of the pathology was congenital.

The male predominance reported in other series (2-4) was also supported by our small number of patients. The typical clinical picture in children consists of hepatomegaly, dilated superficial abdominal veins and ascites, but they are not invariably present in all patients. A high awareness is needed to make the diagnosis. Histologic changes vary from almost normal to severe chronic congestive changes. Chronic by nature, BCS can progress to liver cirrhosis as in our 3 patients.

Liver function tests were slightly abnormal in most cases. None of our patients had HbsAg positivity, in contrast to other series (3). Ultrasound visualisation of the hepatic veins and inferior vena cava is essential for diagnosis.

Ultrasonography, which is the most reliable noninvasive screening test, was diagnostic in our patients, but hepatic and/or inferior venocavogram also should be made for the therapeutic management.

Treatment options include conservative treatment, a variety of surgical vascular by-pass, interventional radiological techniques, decompression procedures, and liver transplantation. Treatment method should be individualized according to the site of obstruction and the extent of liver failure (2). The most common operation performed for the true membranous

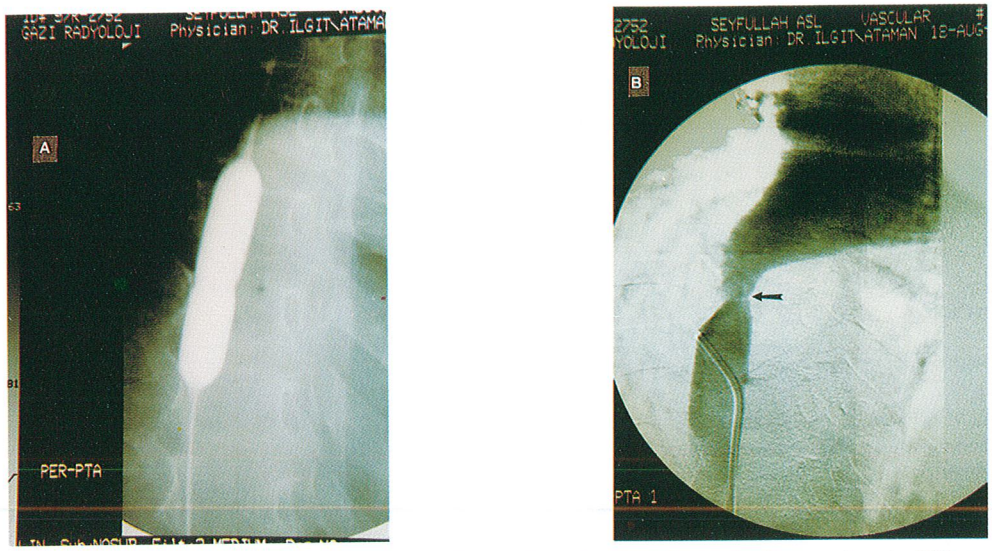

Fig - 4 : Patient 4. The obstructed segment was dilated using balloon catheter (A), immediately after the procedure, the IVC was shown to be sufficiently dilated (B).

obstruction is transcardiac membranotomy (9-11). Poor results are obtained with this procedure if one or both hepatic veins are occluded (12). Recently interventional radiological procedures have gained popularity in the treatment of BCS. Percutaneous membranectomy with a balloon catheter is an alternative procedure in cases with a thin membrane containing a central pore (5, 13). We successfully used this procedure in children with BCS due to membranous web in the hepatic vein in patient 1 and inferior vena cava in patient 4 (Fig. 4 A-B). Follow-up ultrasound examination revealed a patent hepatic vein and inferior vena cava. Unfortunately, balloon dilatation could not be carried out in patients 2 and 3. Prophylactic beta-blocker was started and liver transplantation was planned for these patients.

In conclusion, BCS due to membraneous obstruction of the IVC and/or hepatic vein may not be rare in children. A high level of awareness is needed for the diagnosis.

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