Inferior Mesentericocaval Shunt - an Efficient Therapeutical Alternative in Budd Chiari Syndrome Associated with Portal and Splenic Vein Thrombosis in a Teenager

Lucia Burac, Constantin Ciuce, Zeno Spârchez, Genel Sur, Cristina Mureșan, Nicolae Miu

1) 2nd Pediatrics Clinic. 2) 1st Surgical Clinic. 3) 3rd Medical Clinic, University of Medicine and Pharmacy, Cluj-Napoca

Abstract

The authors present the case of a 17 year old girl admitted to hospital for poor general state, mild scleral jaundice, deficient nutritional state, oliguria and massive ascites. She was diagnosed with Budd-Chiari syndrome: thrombosis of the left suprahepatic vein and nonocclusive thrombosis of the inferior vena cava at the level of the 12th thoracal and the lumbar vertebrae. The specific feature of the case was the association of portal and splenic vein thrombosis. A mesentericocaval shunt with external jugular grefon was performed. The evolution at 20 months after surgery has been favorable. She has no ascites, the nutritional state has normalized and hepatic laboratory findings have returned to normal values. There still persists a high consistency splenomegaly, but without hematological hypersplenism. Even though the mesentericocaval shunt is not without complications, it represents an efficient alternative for the treatment of Budd-Chiari syndrome, when endovascular techniques are not available.

Key words

Budd-Chiari syndrome - portal vein thrombosis - splenic vein thrombosis - mesenterico-caval shunt

Introduction

The incidence of the Budd-Chiari syndrome, defined as the thrombotic or nonthrombotic occlusion/stenosis of the suprahepatic veins (starting with the venules) up to the inferior vena cava (the hepatic portion of the vein) (1), is unknown. Both genders are equally affected. The highest incidence is between 30 to 40 years, but the syndrome has been identified in children and also in older people.

The classical but unspecific triad consists of: hepatomegaly, abdominal pain and ascites and can be found in the majority of the patients (2). There are four evolutive forms: acute, subacute, chronic (the most common) and fulminant. The fulminant evolution is uncommon (4,5) and consists of markedly elevated aminotransferase level, painful hepatomegaly, ascites and renal failure. The specific features of the chronic form comprise accumulation of ascites over a period of two months or longer, mild elevation of aminotransferase level and prolonged prothrombin time. It is not associated with jaundice and some patients present alterations of the renal function. There are also asymptomatic patients, in whom the hepatic sinusoids are decompressed by the portosystemic and intrahepatic collaterals (4).

Case report

A 17 year old female patient was admitted to the 2nd Pediatrics Clinic in Cluj, in April 21, 2004 for the sudden onset of ascites and oliguria, two weeks prior admission. In the patient’s history, the most important events were an appendectomy in 2002 and a first grade atrio-ventricular block. Splenomegaly was first mentioned in 2002, without any further investigations to detect the cause.

The physical examination at admission evidenced a mild alteration of the general state, pallor, mild scleral jaundice, important increase of the abdomen volume due to ascites, visible collateral circulation on the abdomen and chest. The liver and the spleen could not be palpated.

The erythrocyte sedimentation rate, the hemoleucogram and the renal function tests were normal. The antinuclear and anti DNA antibodies were negative. The urinary copper was normal (50.9 μg/24 hours). Serum markers for hepatitis B and C were negative. The laboratory findings showed alterations of all hepatic tests: alanine aminotransferase 226 U/l, aspartate aminotransferase 142 U/l, gamma-glutamyl transpeptidase 105 U/l, total bilirubin 1.78 mg/dl with conjugated bilirubin 0.71 mg/dl, the prothrombin time 20.8 seconds, serum albumin level 2.9 g/dl, blood ammonia...
concentration 64.4 μmol/l (normal values 10-47 μmol/l). Abdominal Doppler ultrasonography (Figs.1-3) and magnetic resonance imaging (MRI) revealed right suprahepatic vein thrombosis associated with portal and splenic vein thrombosis. The MRI could not visualize the superior part of the inferior vena cava, so a cavography was performed. Cavography described the narrowing of the inferior vena cava at the entrance to the left atrium, through the extrinsic compression by the hypertrophied hepatic caudate lobe, and nonocclusive thrombosis of the inferior vena cava at the level corresponding to the 12th thoracic and the lumbar vertebrae. The final diagnosis was: Budd-Chiari syndrome associated with portal and splenic vein thrombosis.

Considering the magnitude of the thrombotic process, some prothrombotic coagulopathies were taken into account. Antithrombin III (AT III) deficiency was detected. The upper digestive endoscopy revealed gastric varices on a portal gastropathy.

Treatment with diuretics, human albumin, plasma transfusions, hepatoprotective drugs, vit K and antibiotics (i.v. ceftriaxone followed by oral ciprofloxacin) was administrated. On May 31, 2004 a surgical intervention was performed: inferior mesentericocaval shunt with external jugular grefon. Anticoagulant therapy was administered, enoxaparin followed by acenocumarine, the last one still continuing to be administered. A hepatic biopsy was performed during surgery, which showed the hyalinization of the walls of the intrahepatic vessels some of them with thrombosis and recanalization, marked congestion in zones 2 and 3 and moderate fibrosis in the portal space.

The evolution after the shunt was slowly favorable. The controls performed at 9, 12 and 20 months after surgery showed the disappearance of the ascites, a normal nutritional state and the normalization of some of the biochemical parameters: aminotransferase level, albuminemia, GGT, bilirubin level, blood ammonia. Splenomegaly is still present (5 cm under the costal border), with an increased consistency but without hematologic hypersplenism. The actual treatment consists of spironolactone associated with ethacrinic acid (two days per week), and acenocumarine.

**Discussion**

In Budd-Chiari syndrome, obstruction of the hepatic veins leads to the increase of pressure in the sinusoids and then in the portal vein. In the first stages, the liver perfusion from the portal vein is low, and could favour the portal vein thrombosis (6). Therefore, obstruction of the portal vein (7) is associated in 10-20% of patients, as in our patient. Another specific feature of this case is the association of the splenic vein thrombosis. The caudate lobe hypertrophy can determine the obstruction of the inferior vena cava, as was found in our patient by means of cavography.

The treatment objective is to obtain as soon as possible the decompression of the congested liver and to reduce the portal hypertension before the development of cirrhosis. The repermeation of the obstructed vein can be obtained through thrombolytic therapy, which is efficient in recent thrombosis (this was not our case) or through endovascular techniques, which are minimally invasive: percutaneous angioplasty with stent placement and transjugular intrahepatic portosystemic stent-shunt.
When the hepatic obstruction is associated with the cava vein obstruction, the classical approach is hepatic transplantation or a mezointeratrial shunt. The modern endovascular techniques allow recanalization and placement of a stent, first in the vena cava, followed by the attempt of recanalization of the origins of the hepatic vein. Since these techniques were not available, a surgical shunt was attempted, which is efficient regarding hemodynamics but has a higher rate of complications.

The shunt types are: portacaval, splenorenal and mesentericocaval (3). In our case, an inferior mesenterico-
vagal shunt with exterior jugular grefon was performed. Although evolution to cirrhosis might take place even after the successful performance of the shunts (8), the evolution of our patient was favorable one year after the shunt was completed.

**Conclusion**

Although the mesentericocaval shunt may result in complications, it represents an efficient alternative in the treatment of Budd Chiari syndrome, especially when endovascular techniques are not available.

**References**