Budd-Chiari Syndrome and Acute Portal Vein Thrombosis: Management by a Transjugular Intrahepatic Portosystemic Shunt (TIPS) and Portal Vein Interventions via a TIPS

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Acute portal vein thrombosis (PVT) is a devastating complication of Budd-Chiari syndrome (BCS). Conservative approach, anticoagulation, systemic or transarterial thrombolysis, and urgent liver transplantation were applied in this scenario but with poor results. We present and discuss an approach to treat BCS complicated by acute PVT. Two young female patients presented with acute liver failure, rapidly progressive tense ascites, renal- and respiratory failure. The diagnosis of chronic BCS complicated by acute PVT was confirmed with ultrasound Doppler. Initial treatment was supportive. Right portal vein localization was by transarterial portogram or by computed tomography–guided microcoil placement. Transjugular intrahepatic portosystemic shunt (TIPS) was performed and included Wallstents and a Jograft in one case and Viatorr stentgraft that was extended later with a Hemobahn stentgraft in another. Mechanical clot removal from the portal system was performed in the primary procedure and in a revision procedure in the following few days. Stents were placed precisely with no extension into the inferior vena cava or deeply into the main portal vein. Patients were fully anticoagulated and patency was assessed by ultrasound Doppler. The procedures were performed on days 5 and 10 following admission. In both cases, successful thrombectomies were reveised and maintained. Partial occlusion of the TIPS and reaccumulation of ascites were reversed with repeated procedure. Both patients were discharged without ascites and normal liver function. In conclusion, urgent TIPS and portal vein thrombectomy via TIPS are emerging therapeutic options that offer a safe and effective treatment to patients with BCS complicated by acute portal vein thrombosis. (J GASTROINTEST SURG 2006;10:417–421)

KEY WORDS: Budd-Chiari syndrome, portal vein thrombosis, hepatic failure, transjugular intrahepatic porto-systemic shunt, thrombectomy

Budd-Chiari syndrome (BCS) is a rare disease with a wide range of different clinical courses, depending upon the acuity and the extent of venous occlusion. More often than not, the clinical course of BCS in Western countries is acute, with rapid progression of liver disease and its sequelae over time periods ranging from weeks to months. Portal vein thrombosis (PVT) is the most common cause of presinusoidal portal hypertension. With the exception of systemic anticoagulation, the current treatment of a patient presenting with acute PVT is rather disappointing. A transjugular intrahepatic portosystemic shunt (TIPS) is the most commonly used treatment for the complications of portal hypertension caused by PVT,[1] but there are insufficient data on its ability to control acute cases of PVT. Both acute and chronic PVT may be found concomitantly with BCS in up to 25% of the patients.[2] This subgroup of patients (especially the majority, who have *chronic* PVT) has a much worse prognosis because neither liver transplantation nor shunt surgery are easily feasible any longer. The clinical

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appearance and the management of the two entities in a single patient have been published only in isolated case reports[3,4] so far. We describe two patients, both presenting with chronic/subacute BCS complicated by acute thrombosis of the portal venous system, and describe their management by the construction of a TIPS and mechanical evacuation of the portal thrombus via this route.

CASE REPORTS

Case 1

A 42-year-old woman was admitted to our department complaining of vague abdominal pain, abdominal distention, and worsening jaundice over the 2 weeks prior to her admission. She had been previously healthy and had not taken any medications. Physical examination revealed a fully alert and conscious patient with normal vital signs, icteric sclerae, distended abdomen with tense ascites, and mild pedal edema with neither respiratory distress nor other signs of chronic liver disease. Laboratory findings on admission were as follows: a normal complete blood count, activated partial thromboplastin time (APTT) of 32 seconds, international normalized ratio (INR) of 2.29, serum glucose of 110 mg/dl, sodium of 132 mEq/L, blood urea nitrogen of 44 mg/dl, and creatinine of 1.63 mg/dl. Levels of serum liver enzyme throughout the course of the disease are presented in Table 1. Serology for hepatitis A, B, and C viruses was negative. Serum levels of ceruloplasmin, ferritin, and transferrin as well as thyroid function tests were within the normal range. Doppler ultrasound study of the abdomen revealed chronic thrombosis of all the hepatic veins and acute thrombosis of the main portal vein extending into the right portal vein with slow hepatofugal flow in the left portal vein. Distally, a thrombus was seen extending into the splenic and superior mesenteric veins. Intravenous heparinization was promptly initiated, but a marked deterioration of the patient's status was observed during the next 48 hours. Disorientation, flapping tremor, oliguria, growing ascites, and respiratory failure ensued. The patient was intubated, and medical treatment for hepatic failure was initiated. An indwelling arterial catheter was placed in the superior mesenteric artery, and selective thrombolysis with urokinase was begun. During the following 48 hours, the patient lost consciousness and became anuric. She was then taken to the interventional radiology suite for a TIPS procedure. Before the TIPS insertion, the thrombosed right portal vein had been marked by a microcoil that was placed under computerized tomographic guidance. Left internal jugular access enabled easier entrance to the stump of the thrombosed right hepatic vein. Using the microcoil as a target, the portal system was accessed via the right portal vein. Venography revealed diffuse thrombosis of the splenic vein and the inferior mesenteric vein, occlusion of the upper superior mesenteric vein at the confluence and extensive non-occlusive thrombosis of the extrahepatic and intrahepatic portal tree. Pre-TIPS pressures were not measured. Mechanical declotting was carried out using the OASIS system (Boston Scientific, Natick, MA) with partial resolution of the thrombus (Fig. 1*A*). This was followed by the construction of a TIPS with a 10 × 80-mm Viatorr stent graft that was extended to the inferior vena cava with a 9 × 50 mm Hemobahn stent graft (both from Gore Medical, Flagstaff, AZ) (Fig. 1*B*). Dilatation of the shunt with an 8-mm balloon yielded a portosystemic gradient of 5 mm Hg.

Table 1. Liver function throughout the clinical course of patients 1 and 2

Patient	Time	AST (U/L)	ALT (U/L)	GGT (U/L)	Alk-P (U/L)	Total bilirubin (mg/dl)	Ammonia (μg/dl)
1	Admission	1468	1615	81	54	2.9	138
	Systemic heparinization	1392	2100	77	54	4.1	191
	Urokinase via SMA	880	1000	67	91	3.0	130
	TIPS and thromectomy	85	55	140	80	1.7	79
	Before revision of TIPS	192	322	108	110	2.5	112
	After revision of TIPS	68	49	112	90	1.3	67
2	Admission	880	910	130	110	2.7	99
	Systemic heparinization	990	844	130	122	4.2	131
	TIPS and thrombectomy	310	212	122	100	1.7	90
	Before revision of TIPS	166	196	100	269	2.8	100
	After revision of TIPS	84	69	100	133	1.7	87

AST = aspartate aminotransferase; ALT = alanine aminotransferase; GGT = γ-glutamyl transferase; Alk-P = alkaline phosphatase; SMA = superior mesenteric artery; TIPS = transhepatic portosystemic shunt.