Gutfile

Budd-Chiari syndrome with portal, splenic, and superior mesenteric vein thrombosis treated with TIPS: who dares wins

PATIENT/HISTORY

A 20 year old man presented with severe ascites and malnutrition due to Budd-Chiari syndrome (BCS) with portal vein (PV), and splenic and mesenteric vein (SMV) thrombosis in the proximal 3 cm. He had received regular and more frequent paracenteses of up to 17 litres each for eight months. He had a poor BCS prognostic index of 8.4¹ (table 1). After referral, despite full anti-coagulation and diuretic drugs, he deteriorated over three weeks. No thrombophilic disorder was found.

DIAGNOSTIC PROCEDURES/INTERVENTION

He underwent transjugular intrahepatic portosystemic stent shunt (TIPS) under general anaesthesia. As all hepatic veins were completely occluded, a direct inferior vena cava (IVC) puncture was made. No patent major PV tributaries were demonstrated; two capsular punctures were embolised. Eventually, a small PV tributary was cannulated and a catheter advanced into the SMV. Because of extensive thrombosis, no PV pressure was recordable; right atrium (RA) 12 mm Hg, IVC 21 mm Hg. The parenchymal tract was dilated and two overlapping 12 mm Memotherm stents deployed. The SMV and PV thrombus was partially mechanically disrupted by an angiographic pigtail catheter. Anti-coagulation was not performed due to the risk of developing intrahepatic haematoma. Two days later, further thrombus was again mechanically disrupted and one Memotherm stent was overlapped in the upper and one in the lower end of the TIPS tract. Good flow was re-established and pressures were (mm Hg): PV 33, IVC 22, portosystemic gradient 11, and RA 6. Heparin was started. A widely patent TIPS shunt with excellent flow was present a week later with an RA pressure of 13 mm Hg and PV 23 mm Hg. Warfarin was commenced.

OUTCOME

The patient improved progressively, only one paracentesis was needed, ascites resolved completely, and diuretics were stopped after 45 days. At 60 days he had

improved dramatically, with no ascites, and body weight increasing by 12 kg (table 1).

DISCUSSION

BCS can be associated with splanchnic venous thrombosis in up to 20% of patients, and this association worsens prognosis.² If the thrombus extends down the SMV, both radiological and surgical intervention, including liver transplantation, are contraindicated in many centres. In this setting, successful treatment with transcaval liver resection and hepatoatrial anastomosis (Senning's procedure) has been reported.³ However, this surgery requires sternotomy and laparotomy, and cannot be performed in severely ill patients such as ours who was severely malnourished and with worsening hepatic function. In contrast, TIPS has been reported in single BCS patients not responding to medical therapy and with portal vein thrombosis alone.⁴

Our patient shows that extensive splanchnic venous thrombosis in patients with BCS is not an absolute contraindication for TIPS and is worth attempting even when SMV is thrombosed. There is only one previous case report in which TIPS was used in this situation: TIPS was successful and the patient was doing well after 18 months.⁵

In conclusion, we believe that TIPS should be attempted in patients with BCS even when splanchnic veins are

extensively thrombosed. It can provide the only therapeutic avenue, improvement can be spectacular, and liver transplantation can be avoided.

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Table 1 Comparison of laboratory and clinical data before and two months after transjugular intrahepatic portosystemic stent shunt (TIPS)

Parameter (normal values)	Before TIPS	After TIPS
Weight (kg) (height 170 cm)	45	56
Haemoglobin g/l (11.5-15.5)	8.2	11.6
White blood cells ($\times 10^9/l$) (3.2-11)	12.2	5.5
Platelets ($\times 10^9/l$) (140-400)	260	308
International normalised ratio (0.9-1.2)	2.3*	2.3*
Alanine aminotransferase (u/l) (5-40)	24	27
Aspartate aminotransferase (u/l) (5-40)	46	37
Bilirubin ($\mu mol/l$) (5-17)	66	51
Albumin (g/l) (35-50)	27	39
Creatinine ($\mu mol/l$) (60-120)	66	45
Ascites	Yes	No
Encephalopathy	No	No
Child-Pugh class	C	B
Diuretics	Yes	No
Budd-Chiari syndrome index >5.4	Yes	No

*Under warfarin.