CASE REPORT

Percutaneous intervention averted the need for liver transplantation

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Received 17 February 2012; accepted 7 May 2012
Available online 18 July 2012

KEYWORDS
Budd–Chiari; IVC stenosis; TIPSS complications; Percutaneous intervention; Balloon dilatation

Abstract
A patient with Budd–Chiari Syndrome who had Trans-jugular Intrahepatic Portosystemic Stent Shunt (TIPSS) performed with no improvement and was consequently put on the liver transplant list. A non-obstructed channel up to the right atrium was created by opening the misplaced TIPSS stent and dilating supra-hepatic inferior vena cava stenosis. Marked clinical improvement occurred with the patient losing ten kilograms of weight and both his lower limb edema and ascites disappeared 6 weeks after the procedure. In this case percutaneous intervention averted the need for liver transplantation.

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1. Introduction
Budd–Chiari syndrome is a clinical condition caused by obstruction of the venous outflow of the liver. Its pathogenesis is complex, usually involving an acquired thrombotic stimulus associated with a genetic clotting abnormality producing occlusion of the hepatic veins. Several treatments are available for the different stages and clinical manifestations of the disease with liver transplantation being offered after more conventional therapy, such as anticoagulation, diuretics or porto-systemic shunts, had failed as was the case in this patient.

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Peer review under responsibility of Egyptian Society of Cardiology.

2. Manuscript
A 20 year old male presented with a history of long standing bilateral lower limb edema accompanied by progressive abdominal swelling. He was thoroughly investigated and an abdominal CT scan revealed significant narrowing in all hepatic veins with a web obstruction extending into the supra-hepatic inferior vena cava (IVC) (Fig. 1). He was diagnosed as a case of Budd–Chiari Syndrome due to heterozygous Factor V leiden mutation. Transjugular Intrahepatic Portosystemic Stent Shunt (TIPSS) was performed, and a 10 cm 8 French stent was placed between the portal vein and IVC with no improvement of the patient’s condition. An abdominal CT scan performed six months later revealed that the stent was misplaced as it was seen extending horizontally across the IVC lumen and the already present supra-hepatic IVC narrowing itself had not been tackled (Fig. 2).

On examination, the patient had bilateral wasting of temporal muscles and mild pallor. Abdominal examination revealed tense ascites with dilated veins visible over his flanks.
(Fig. 3) that drained upward by milking test. There was bilateral, pitting lower limb edema up to the level of his knees.

Laboratory investigations revealed microcytic hypochromic anemia, with an International Normalized Ratio (INR) of 1.1. His serum creatinine level was 1.1 mg/dL, total bilirubin was 1 mg/dL, albumin was 2.6 mg/dL, ASOT was 55 mg/dL, and ALT was 66 mg/dL.

The patient was listed for liver transplant after failure of the TIPSS procedure with persistence of symptoms. He had required repeated abdominal paracentesis nearly every week and he was on diuretic therapy. We approached the patient with the plan of percutaneous (transfemoral) intervention to relieve the two levels of obstruction; the first caused by the endothelialized, misplaced TIPSS stent and the second in the supra-hepatic portion of the IVC before its entry into the right atrium. At first the patient refused, but because of the long waiting list, he finally gave his approval to perform the procedure after 6 months.

2.1. Procedure

We aimed to create a non-obstructed channel up to the right atrium. This would be achieved by opening a channel in the misplaced TIPSS nitinol stent and dilating the suprahepatic inferior vena cava (IVC) stenosis. We planned to deploy balloon expandable stents at both sites.

After inserting a femoral sheath, venography revealed obstruction of the IVC above the level of the renal veins.
We started off by passing a zip wire through the IVC obstruction and through the TIPSS stent to RA. After repeated trials, we also managed to pass a PTCA wire. The main difficulties we faced during crossing the wires were caused by the many tributaries and the TIPSS nitinol stent that crossed horizontally in the IVC.

A 3.5 × 20 mm coronary balloon was then passed and inflated at a pressure of 20 atmospheres to open a track. After that we were able to pass a 4 × 30 mm peripheral balloon that was inflated at a pressure of 14 atmospheres. Minimal flow improvement occurred (Fig. 5). At that point, we decided to exchange the femoral sheath with an 11 French long sheath with which we were able to introduce just below the level of the TIPSS stent struts. This enabled us to dilate using a 10 × 30 mm balloon after which we were able to introduce the long sheath past the TIPSS stent struts, thus enabling the passage of a 20 × 30 mm balloon which was repeatedly inflated both at the level of supra-hepatic obstruction and at the level of the TIPSS nitinol stent (Fig. 6), thus partially crushing it.

We finished off with having full dilatation of the original supra-hepatic IVC stenosis and to a lesser extent at the level of the TIPSS stent (Fig. 7). It was decided to stop at this stage and follow up the patient without stenting.

Rapid marked clinical improvement occurred. The patient lost 10 kg and both his lower limb edema and ascites disappeared just 6 weeks after the procedure (Fig. 8). Abdominal

**Figure 5** Venography showing that still no satisfactory results were seen.

**Figure 6** Maximal inflation with the 20 × 30 mm balloon showing constriction at the level of TIPSS stent.

**Figure 7** Final outcome with restoration of venous flow without significant obstruction.

**Figure 8** Scaphoid abdomen 6 weeks after second intervention indicating relief of ascites.
CT scan three months after the procedure revealed satisfactory results with disappearance of the collaterals indicating relief of obstruction (Fig. 9).

At present, after nine months of follow-up, the patient did not require at any point to receive diuretic therapy and has no lower limb edema or ascites.

3. Discussion

Studies have shown that TIPSS dysfunction was mostly related to stent restenosis or complete occlusion by thrombosis which was corrected by angioplasty or restenting. There was also a report of a stent being dislocated with its subsequent placement into the right iliac vein.

In our reporting of this case we aimed to display how percutaneous intervention averted the need for liver transplantation in a patient with Budd–Chiari Syndrome with two levels of obstruction; one caused by the disease itself and the other iatrogenically caused by a previously misplaced TIPSS stent.

References