INTRODUCTION

Transjugular intrahepatic portosystemic shunt (TIPS) is a new treatment modality in patients of intractable ascites with Budd-Chiari syndrome. We report a young lady who presented with intractable ascites and encephalopathy and diagnosed to have Budd-Chiari syndrome. She was treated with TIPS and on follow-up after 12 months she is free of ascites, encephalopathy and able to perform her daily work.

CASE REPORT

Twenty five year old lady presented with progressive distension of abdomen and swelling of feet since one year. On presentation she had massive ascites, not responding to medical therapy. There was no history of hematemesis, melena and jaundice in the past. There was history of premature delivery three months prior to the development of abdominal distension. There was no past history of blood transfusion or any other major illness. On examination she was drowsy, there was mild icterus, bilateral pedal edema, liver was enlarged 3 cm below the right costal margin, firm in consistency, spleen was not palpable and there was free fluid in the abdomen.

On investigation her hematological parameters were normal. Liver function tests showed bilirubin on 1.5 mg/dl, direct bilirubin being 1.2 mg/dl, SGOT, SGPT were mildly raised, serum protein was 5.6 gm%, albumin 2.6 gm%, globulin 3 gm%, INR was 2.1. Renal function test was within normal limits. Ascites fluid analysis showed protein of 2.8 gm/dl, cell count of 24 cells/cumm, ADA of 4 units/L. Echocardiogram of heart and chest X-ray were within normal limits. Viral markers like HBeAg, Anti HCV and HIV were negative. Ultrasonography with color Doppler of abdomen showed mild hepatomegaly with nodular surface and mild splenomegaly, portal vein diameter being 12 mm with hepatofugal flow. Right and middle hepatic veins showed narrowing with very poor flow. Left hepatic vein was showing good flow. Intrahepatic collaterals were seen. There was narrowing of intrahepatic IVC, but there was no thrombus within it. Upper gastrointestinal endoscopy revealed 3 column of Grade-1 oesophageal varices.

With the above Doppler finding the diagnosis of Budd-chiari syndrome (BCS) with refractory ascites was established. Procoagulant workup for protein C, protein S, antiphospholipid antibody, factor V leiden mutation failed to reveal any positive result. Considering the resistant ascites and deteriorating liver function she was referred for angiographic study and interventional radiological treatment. IVC gram revealed narrowing of the intrahepatic IVC due to caudate lobe enlargement, without any evidence of thrombus or web. Right jugular vein was canalized and on advancement of the catheter into the suprahepatic IVC, stump of right hepatic vein was opacified. Percutaneous hepatic venogram was suggestive of venous collaterals within irregular right and middle hepatic vein. Left hepatic vein was smooth, but small and not draining freely into the IVC and due to its small caliber was not suitable for hepatic venous angioplasty. The extensive intrahepatic thrombosis precluded recanalization of middle and right hepatic vein, thus TIPS was considered. A tract was created between the hepatic venous stump and the portal vein bifurcation. Portal venogram showed mild dilatation of portal vein and retrograde filling of the left gastric vein with a portosystemic gradient (PSG) of 28 mm Hg. The tract was dilated and a self expandable metal stent (wall stent, Boston Scientific) was put within the tract. Following the procedure, portosystemic gradient dropped to 9 mmHg. She was started on heparin followed by oral anticoagulants to maintain the INR between 2.5 and 3. Her ascites regressed completely within two weeks and liver function improved with bilirubin of 1.2 mg/dl, albumin 3.6 gm/dl. She was followed up for prothrombin time every weekly and for stent patency by Doppler initially monthly for three months and then three monthly. Nine months after TIPS, she developed ascites and on Doppler the stent was found to be blocked. She underwent balloon dilatation and effective recanalization of the stent. Now at 12 months of followup, she is free of ascites and able to perform her daily work. She did not have single episode of encephalopathy.

DISCUSSION

Most common presentation of BCS is subacute liver disease complicated by ascites, portal hypertension and varying grades of hepatic failure. They require treatment with portal decompression surgery or liver transplantation. Surgical shunts are associated with high mortality and morbidity. TIPS has been used recently as a treatment modality in BCS, which is less invasive, has less morbidity.
Recent reports have evaluated the long term effect of TIPS in BCS.\(^3\) Patients without cirrhosis have excellent prognosis with decompressive procedure and TIPS has allowed time for development of extensive collateral, suffice to maintain the liver decompression despite TIPS stenosis.\(^4\) It has shown to improve the liver function, controls the ascites and reduces the transaminase level in BCS not responding to medical treatment.\(^4\) In acute BCS, TIPS helps in decompression as well as thromblytic therapy.\(^4\) Common problems encountered in TIPS are recurring TIPS occlusion, TIPS dysfunction which can be taken care by balloon dilatation and restenting. Preliminary results of new expanded-polytetrafluoroethylene covered stent grafts have shown improved primary and secondary patency rates.\(^6\) TIPS with these stents might help in better stent patency in BCS.

TIPS in Budd Chiari syndrome requires greater skill as hepatic vein thrombosis adds difficulty to the procedure. In India where liver transplantation is still not well established, TIPS is a viable option in BCS not responding to medical therapy as there is not much alternative. However patients need close monitoring of coagulation parameters and Doppler assessment for stent patency.

**REFERENCES**


