CASE REPORT

Innovation in direct intrahepatic portosystemic shunt; a developing country experience

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Abstract

Anticoagulants are the first-line treatment option for patients with Budd-Chiari syndrome (BCS); however, intervention is required when this fails. Although, the ultimate treatment is liver transplant, other radiological procedures are for the management of the disease and bridge to definitive therapy. TIPS (trans jugular intrahepatic portosystemic shunt) is a method used by interventional radiologists to create a shunt between portal vein and hepatic vein. At times it is technically not possible, in such cases, direct intrahepatic portosystemic shunt (DIPS) is performed. This patient underwent a successful DIPS procedure for BCS along with balloon dilatation (venoplasty) for inferior vena cava (IVC) stenosis.

Keywords: Budd-Chiari syndrome (BCS), Trans jugular intrahepatic portosystemic shunt (TIPS), direct intrahepatic portosystemic shunt (DIPS), inferior vena cava (IVC).

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Introduction

Budd-Chiari syndrome (BCS) also known as hepatic venous outflow obstruction (HVOO), occurs when hepatic veins are partially or completely blocked. According to one study prevalence of BCS is 1/100 000 in the general population[1]. Ascites, hepatomegaly, abdominal distention and pain are the typical symptoms of BCS. Acute thrombosis and stenosis of hepatic veins and IVC are usual underlying aetiologies of acute and chronic presentation respectively. The aim of treatment in cases of BCS is to relieve portal pressure and IVC hypertension [2]. Initially ascites and varices are managed conservatively and screening is done. Anti-coagulation is advised if the disease is secondary to thrombosis from hyper-coagulopathy. If symptoms persist despite medical treatment, Trans Juglar intrahepatic portosystemic shunt (TIPS) is the least invasive treatment modality. DIPS is performed in cases where TIPS is not technically possible due to hepatic vein thrombosis or occlusion[3]. IVC stenosis complicates the treatment both technically and clinically. Venoplasty is warranted in such cases. DIPS/TIPS are not readily available in developing nations such as Pakistan due to a shortage of interventional suites and competence. In this case, an 18-year-old female had DIPS as well as balloon dilatation of IVC stenosis.

Case Report

A female 18-year-old Afghan resident, complained of abdominal pain and distention for the past 6 months. She was initially treated as a case of abdominal tuberculosis and given anti-tuberculous drugs for 3 months and then referred to Liaquat National Hospital and Medical College, Karachi, Pakistan. Budd Chiari syndrome was diagnosed based on the patient's presentation and lab test results. The patient underwent paracentesis for persistent ascites multiple times. Spontaneous bacterial peritonitis (SBP) was ruled out and diuretics were started. An upper GI endoscopy revealed grade II oesophageal varices and portal hypertensive gastropathy, for which propranolol was prescribed as a primary prophylaxis. Hypercoagulable workup revealed protein C deficiency. Her ANA turned out to be positive along with Cardiolipin IgM and Beta 2Glycoprotien1 IgM. Despite receiving maximal diuretic therapy ascites persisted requiring multiple large-volume paracentesis. She was unable to walk and was having breathing difficulty due to markedly distended abdomen.

Doppler ultrasound of liver showed no flow in hepatic veins on Colour Doppler imaging (CDI) along with liver parenchymal changes secondary to congestion.

CT imaging showed large volume ascites with a typical flip flop pattern of hepatic enhancement, non-visualization of hepatic veins, stenosis of hepatic segment of IVC, caudate lobe enlargement, intrahepatic collateral vessels, and hypervascular nodules suggesting Budd-Chiari syndrome.

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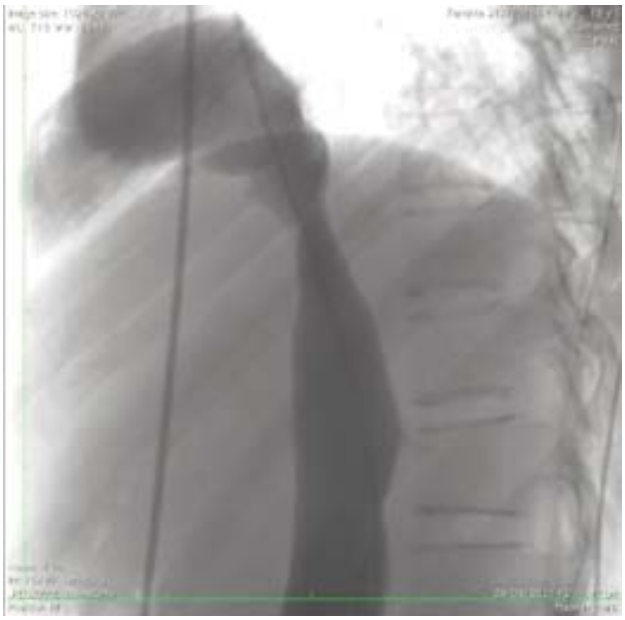

Figure-1: Venogram lateral view done through Femoral vein shows stenosis in hepatic segment of IVC

DIPS along with venoplasty of IVC was performed in Vascular and Interventional Radiology Department, Liaquat National Hospital and Medical College, Karachi, Pakistan on 31st March,2021. The venogram revealed a significant stenosis in intrahepatic IVC (fig.1) which was dilated with two balloons (12 mm each in diameter x 40 mm in length) keeping them side by side. (fig.2). This side by side double-balloon technique was used because of the non-availability of a single large diameter high-pressure balloon and also to save the cost, which is of great concern in the majority of patients in developing countries like Pakistan.

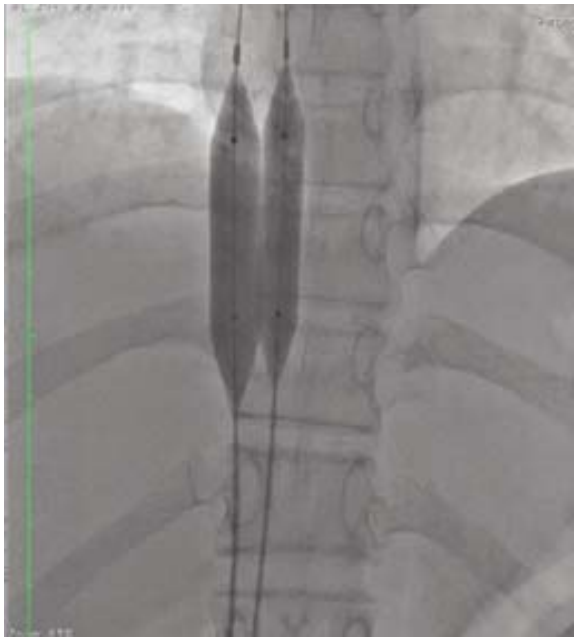

Figure-2: Shows Venoplasty of IVC being done by keeping Two balloons in side by side technique

The right branch of the portal vein was accessed via direct puncture from the intrahepatic part of IVC. The tract was dilated via angioplasty using a conquest (BARD) balloon 8mm x 40mm. Fluency stent graft (BARD) measuring 10mm x 80mm was placed across the intrahepatic tract with the proximal end in IVC and distal end just reaching the right branch of the Portal vein. Then the distal end was extended in the main Portal vein via overlapping bare metal self-expanding Rontis stent measuring 9mm x 60mm as shown in Fig.3 (postprocedure venogram). The double graft stent technique was used because of the nonavailability of the VIABAHN (GORE) stent graft, which is ideal for TIPS/DIPS.

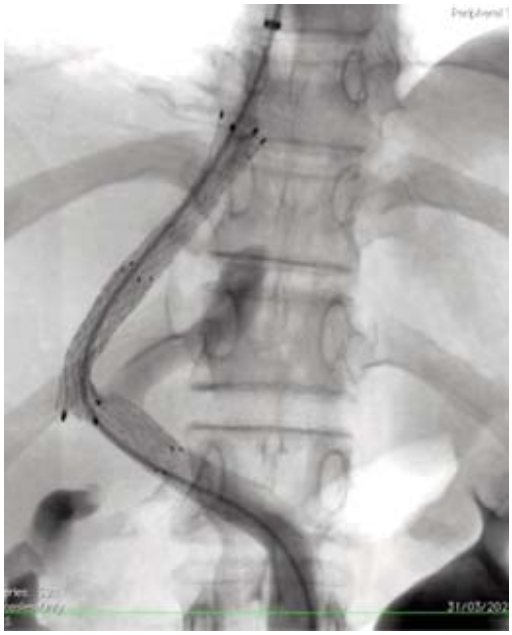

Figure-2: Post DIPS run shows shunt created between IVC and right branch Of portal vein using fluency graft and wall stent

The patient was reviewed after 3 days and an ultrasound result showed a significant resolution of ascites. The patient was discharged on dual antiplatelets along with low molecular weight heparin. The patient remained asymptomatic till three months follow-up. Before the procedure, she was unable to walk due to massive abdominal distention secondary to ascites. Now the patient is fully mobile and going to the university.

Discussion

A large percentage (60%) of BCS patients in Far East suffer from idiopathic membranous occlusion of IVC which in turn leads to IVC thrombosis without hepatic vein thrombosis. BCS develops secondary to thrombosis or structural constriction of the hepatic venous outflow, culminating in venous congestion, ischaemic necrosis, and, eventually, cirrhosis[4]. Clinical presentation can be fulminant, acute, or chronic but most of the time asymptomatic for a longer period and suddenly deteriorating with portal hypertension[5]. There is a severe morbidity associated with BCS if it is not assessed and addressed promptly, untreated BCS has mortality rates more than 70% and 89% at 1 and 3 years respectively[6,7]. The eventual aim of BCS is to reduce venous congestion to arrest disease development and hence reduce morbidity and death. The success rate for DIPS is reported in different articles across the world. One of the recent studies of 14 patients showing a success rate of 93%. While the resolution of ascites was seen in 64% of cases and post DIPS hepatic encephalopathy was observed in 14% of cases[8]. Patency rates for primary stents at 6 months, 1 year, and 2 years were 83%, 83%, and 58%, respectively. The patency rate of secondary stents was 100%. To date, transplant-free survival has been 100%[9]. Figures from India show a 94% transplant-free survival rate at one and five years[9].

Conclusion

If medical care fails and TIPS is not technically possible, patients with Budd Chiari syndrome can be treated using the DIPS procedure under a multidisciplinary team approach. DIPS can yield good clinical outcomes and promising results with proper patient selection and early referral to the centre of expertise.

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Conflict of Interest: None.

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