The use of intravascular ultrasound (IVUS) for direct portosystemic shunt (DIPS) evaluation: Case report

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Abstract: A 32-year-old male with Budd–Chiari syndrome secondary to paroxysmal nocturnal haemoglobinuria (PNH) underwent direct portosystemic shunt (DIPS). The procedure was difficult, and after initial attempts via the jugular vein without success, a stent was placed with ultrasound (US) guidance via femoral approach. Unfortunately, following these two prolonged DIPS procedures, two weeks apart, the patient developed contrast-induced nephropathy, precluding further use of conventional contrast media and necessitating an alternative imaging modality to assess stent function. Transabdominal US was unable to clearly visualize the stent, so intravascular ultrasound (IVUS) was utilized. Good quality images were obtained and the stent’s patency was accurately evaluated, identifying the presence of thrombus and leading to balloon angioplasty and improved flows. IVUS may be useful for DIPS stent evaluation.

Keywords: Budd–Chiari syndrome, paroxysmal nocturnal haemoglobinuria, direct portosystemic shunt, intravascular ultrasound

Introduction

Paroxysmal nocturnal haemoglobinuria (PNH) is a rare condition which leads to intravascular haemolysis and hepatic vein thrombosis [1]. In patients with PNH, acute and chronic renal failure have been described [2, 3], and these patients may therefore be more vulnerable to contrast-induced nephropathy.

Budd–Chiari syndrome (or hepatic outflow syndrome) is caused by thrombosis of the hepatic veins and leads to portal hypertension, intractable ascites and variceal bleeding [4–6]. The conventional transjugular intrahepatic portosystemic shunt (TIPS) procedure for decompression of the portal circulation may not be feasible to perform because of the occlusion of the hepatic veins, and therefore, the direct portosystemic shunt (DIPS) procedure has been successfully developed [7–10].

Case Report

A 32-year-old male patient suffering from chronic Budd–Chiari syndrome due to PNH was referred to our unit for a TIPS procedure. The patient was checked for thrombophilic factors (antiphospholipid syndrome and factor V Leiden thrombophilia) but the result was negative. The patient was also checked for antiphospholipid syndrome and the result was negative. The patient did not have any symptoms of encephalopathy or previous variceal bleeding. Laboratory values revealed slight liver function impairment but normal renal function and coagulation status. In particular, aspartate aminotransferase (AST) was 199 U/L, alanine aminotransferase (ALT) 55 U/L, gamma-glutamyl transferase (GGT) 178 U/L, total bilirubin 1.41 mg/dL, creatinine 0.9 mg/dL and INR value 1.15. Abdominal Doppler ultrasound (US) examination demonstrated partially occluded hepatic veins but a patent portal vein and inferior vena cava (IVC). A contrast-enhanced computed tomography (CT) scan revealed patchy enhancement of the liver parenchyma with areas of thrombosis within the hepatic veins and the presence of ascites (Fig. 1).

Having drained the ascites the day prior to the procedure and administered adequate local anaesthesia and sedation, a 10-Fr introducer sheath was advanced, via the right internal jugular vein, into the IVC and the middle hepatic vein was catheterized. A 10-Fr angled catheter with a 14G stiffening cannula (Rösch-Uchida Transjugular Liver Access Set, Cook, Denmark) was used to gain access to the portal system. Several attempts were made but without success, because of the extensive hepatic vein thrombosis.

A direct approach from the IVC was then decided, using transabdominal US guidance. The distal cannula was...
modified manually in order to obtain a more acute angle and to be more visible in the US picture. A 3–6-MHz convex US probe was used (Esaote, Exaote, Florence, Italy) and positioned in the right 8th intercostal space in the mid-axillary line. Following several attempts, the portal system was successfully catheterized. Nevertheless, the tract formed was considered as not suitable for stent deployment due to the acute angulation between the IVC and the segment of the portal vein that was punctured. The procedure was abandoned at that point, and a second attempt was scheduled for 2 weeks later.

Two weeks later, percutaneous drainage of ascites was again performed prior to the procedure. Access via the right femoral vein was obtained and the 10-Fr angled catheter/14G stiff cannula set (Rösch-Uchida Transjugular Liver Access Set, Cook, Denmark) was advanced in the IVC. Under transabdominal US guidance, the right branch of the portal vein was punctured (Fig. 2). The portal vein was successfully catheterized and a guide wire was advanced securing the portosystemic communication. Two 8 mm × 80 mm self-expandable nitinol metallic stents (Luminexx, C. R. Bard Inc, Murray Hill, NJ, USA) were deployed in the tract (Fig. 3). There was still an acute angulation between the portal vein and IVC but stent deployment was feasible this time. Anticoagulation with heparin infusion followed (1000–2000 units/h by intravenous infusion, Heparin Sodium, Wockhardt UK) for 72 h post-procedure.

The patient had received 400 mL of iso-osmotic iodine contrast media during the two procedures, which compounded by the inability to adequately fluid hydrate him given the risk of pulmonary oedema, and despite N-acetyl cysteine administration, he developed acute renal failure necessitating haemofiltration. An evaluation of the stent position and patency was still important, in order to exclude stent thrombosis because of the patient’s pro-thrombotic state and also because of the caudal alignment of the stents with respect to the IVC.

Transabdominal Doppler US was suggestive of stent dysfunction since very low in-stent flow rates were detected, but the exact cause of this could not be estab-
lished. In order to define the aetiology of the stent dysfunction, and bearing in mind that no conventional contrast agents could be administered, intravascular ultrasound (IVUS) was employed for stent’s assessment. Through a 6-Fr introducer sheath, via transfemoral approach, a 0.018” guide wire was advanced into the stent’s lumen. Over the guide wire, an IVUS transducer was advanced (Visions® PV, Volcano Corporation, USA) connected with a mechanical sector scanner (In-Vision Gold, Volcano Corporation, USA). The IVUS detected the presence of limited in-stent thrombosis (Fig. 4), which was treated with balloon angioplasty. IVUS confirmed improved patency within the stents post-angioplasty. The patient’s liver function and renal function returned to normal 3 weeks later. The ascites improved and the patient is now on the waiting list for liver transplantation.

Discussion

Budd–Chiari syndrome is the clinical expression of a heterogeneous group of disorders that lead to thrombosis of the hepatic veins, the IVC and even the right atrium, resulting in portal hypertension, intractable ascites and variceal bleeding [4–6]. Interventional radiology offers a valid treatment for portal hypertension in patients affected with Budd–Chiari syndrome following the development of the conventional TIPS procedure and lately with the DIPS procedure [7–10].

Petersen described the possibility of performing DIPS with the use of IVUS located in the IVC in order to guide the needle directly to the portal vein [7]. Other authors have performed DIPS or modified DIPS with the guidance of transabdominal US. Boyat et al. in 2006 reported a case of simultaneous transhepatic puncture of the portal vein and vena cava in a case of Budd–Chiari syndrome, guided by a 2–5-MHz external US with a curved-array transducer through a subcostal window [9]. Quateen et al. reported a series of six patients, in whom, under external US guidance, a transhepatic puncture of the portal vein from the IVC was performed [8]. Hoppe et al. [10] described the use of a covered device in 19 cases where DIPS was performed and revealed that patency after DIPS creation with a nitinol PTFE-covered stent-graft was superior to that after TIPS with the same device.

Our case demonstrates this same technique performed from the femoral vein as opposed to the jugular vein. Despite the potentially unfavourable direction of the stent placement using the femoral approach, the shunt successfully drained from the portal vein to IVC. We recognize depending on stent alignment and relative pressures between the portal vein and IVC that there is a risk of procedural failure using the femoral technique.

The aetiology of Budd–Chiari in our case was PNH. This is a rare condition in which an acquired defective clone of the haemopoietic stem cell is manifested by an increased sensitivity of the red blood cells, granulocytes and platelets to complement lysis [1]. The major clinical features of PNH are chronic intravascular haemolysis with episodes of haemoglobinuria and diffuse venous thrombosis which in our case led to hepatic vein thrombosis.

Acute and chronic renal failure have been described in patients affected by PNH in several reports [2, 3], although there are no reports of contrast-induced nephropathy. Chronic renal damage has been attributed to tubular atrophy and interstitial fibrosis from haemosiderin deposits in the renal tissue due to the intravascular haemolysis, whereas acute renal failure usually occurs because of acute tubular necrosis [2]. In our case, the renal function of the patient was normal before the procedures and returned to normal several weeks later.

Owing to the increased thrombogenicity of the patient and the ongoing production of ascites, assessment of the implanted endoprosthesis was necessary in the early post-procedural period. We used IVUS for in-stent assessment of patency, which is an established method of in-stent thrombosis assessment, particularly in the coronary arteries [11, 12]. IVUS was used by Petersen [7] and Hoppe et al. [10] for DIPS creation. The authors used a low-frequency probe (variable 5–10 MHz) which was not available in our department, and puncture guidance was performed with transabdominal US. Nevertheless, the high-frequency IVUS device that was used was successful in in-stent thrombosis assessment. Alternatively, the use of CO₂ as a contrast medium could be considered.

In conclusion, in patients undergoing DIPS procedure, the use of US (transabdominal or IVUS) is, in our opinion, critical for needle guidance. In addition, access via the femoral approach may be considered in cases where the jugular approach is occluded or has previously failed. If conventional contrast media are contraindicated, high-frequency IVUS can be considered as a valid imaging modality for the assessment of in-stent pathology.
The manuscript is prepared according to the regulations described in the IMAS ethical statement (13).

References