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Case Report

Budd-Chiari syndrome treated with direct intrahepatic portocaval shunt: A case report ☆

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ABSTRACT

Budd-Chiari syndrome is an uncommon disorder characterized by occlusion of hepatic veins. It can lead to portal hypertension. Most common causes of this syndrome are hypercoagulability states. Transjugular intrahepatic portosystemic shunt is often not possible given the portal venous thrombosis. In these cases, direct intrahepatic portocaval shunt, involving the creation of an access between the portal vein and the systemic circulation via the intrahepatic inferior vena cava has proven to be a feasible alternative, and can improve portal hypertension in these patients. Herein, we present a case of a 37-year-old woman diagnosed with Budd-Chiari syndrome that was successfully treated with percutaneous ultrasound (US)-guided direct intrahepatic portocaval shunt.

Introduction

Budd-Chiari syndrome (BCS) is a relatively uncommon congestive hepatopathy featuring hepatic venous outflow obstruction [1]. Primary BCS is the result of obstruction to outflow due to a venous process, such as phlebitis, thrombosis, or its fibrous sequelae. In Secondary BCS, the obstruction of outflow is caused by neighboring structures, such as tumor invasion or extrinsic compression [2]. Most BCS cases are related to hypercoagulability. This is demonstrated in primary myeloproliferative diseases, which are the leading cause of hepatic vein thrombosis. The annual incidence of Budd-Chiari Syndrome is 0.1 to 10 per million [3]. BCS can cause a variety of symptoms with varying severities. While patients may be asymptomatic, most patients develop symptoms that result from liver damage. Common symptoms are abdominal pain, ascites, leg swelling, jaundice, and hepatomegaly [4]. Severe centrilobular congestion, hepatocellular necrosis, and atrophy result from BCS. Over the course of the disease, nodular re-

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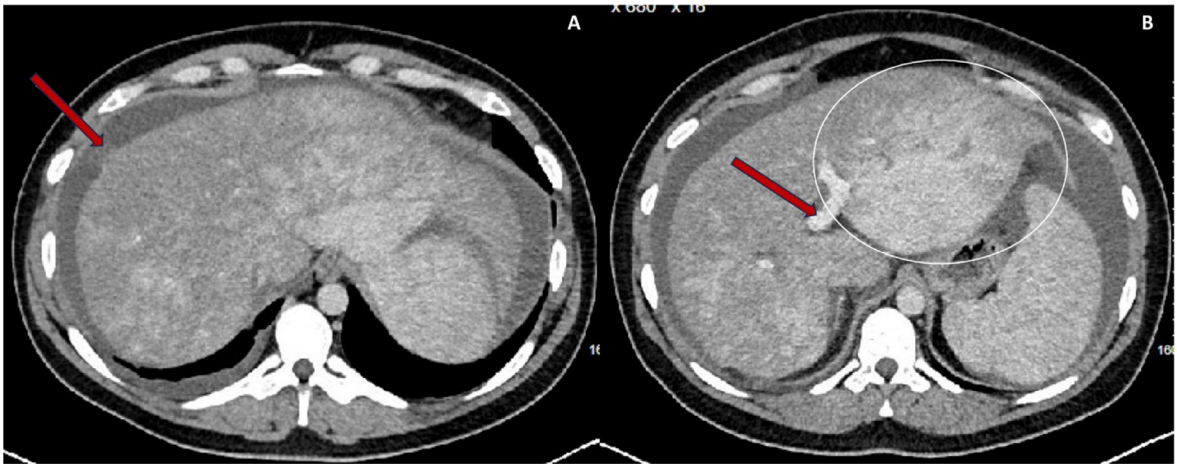

Fig. 1 – (A) Axial portal venous phase CT of the upper abdomen demonstrating the "nutmeg appearance" of the liver parenchyma, with no discernible hepatic venous flow. Perihepatic ascites is noted (Red arrow). (B) Axial portal venous phase CT of the upper abdomen demonstrating that the portal vein is patent (Red arrow). There is hypertrophy of the left lobe (white circle).

generative hyperplasia, progressive fibrosis, and cirrhosis may also develop.

Case report

A 37-year-old woman with no significant past medical history presented to the emergency department with progressive abdominal distention over a 1-month period. Prior to current presentation, she was seen by her gynecologist, and sonographic evaluation was significant for ascites. No suspicious adnexal masses were noted. Contrast-enhanced abdominopelvic computed tomography (CT) was ordered (Figs. 1A and B), which was, in addition to the ascites, significant for hepatosplenomegaly associated with heterogeneous hepatic parenchymal enhancement.

While the portal veins were patent, the hepatic veins were not visible on CT, raising suspicion for BCS. No gynecologic etiology for the ascites was noted on CT.

Paracentesis was performed and laboratory evaluation was consistent with transudative etiology. Considering these findings, the patient was started on systemic anticoagulation, with the presumed diagnosis of ascites due to portal hypertension induced by hepatic venous occlusion (BCS). A hypercoagulability state workup was performed, and the patient was diagnosed with protein S deficiency.

Given the findings of BCS, and the need of frequent paracenteses with the drainage of at least 5 L of ascitic fluid every other week, and because the patient's ascites could not be controlled with conservative management, evaluation for the creation of a portosystemic shunt was performed. The patient was noted to have a normal renal panel and liver function test, despite an INR (international normalized rate) of 1.6 and an albumin blood level of 3.0 g/dL. Her MELD (model for end-stage liver disease) score was noted to be 13. The patient has also undergone esophagogastroduodenoscopy (EGD) showing no esophageal or gastric varices, and an echocardiogram showing no evidence of heart failure.

A right internal jugular (IJ) venous sheath access was obtained at the beginning of the procedure, and a paracentesis was performed. Given the occlusion of the hepatic veins, the creation of a transjugular intrahepatic portosystemic shunt (TIPS) was not possible, with intravascular sonographic evaluation showing no discernible patent hepatic veins. As such, a direct intrahepatic portosystemic shunt (DIPS) creation was considered. Intraoperatively, the patient was found to have a portosystemic gradient of 14 mmHg (portal venous pressure of 26 mmHg and a systemic venous pressure of 12 mmHg) (Fig. 2A).

The gun sight technique for DIPS stent placement was considered, following obtaining direct ultrasound-guided percutaneous portal venous access and vascular sheath placement. Two 1.5 cm loop snares were placed, one in the right portal vein and another in the intrahepatic inferior vena cava (IVC). A separate 20-gauge percutaneous needle (Fig. 2B) access was obtained targeting both snares. Once wire access of the IVC was obtained, the IVC snare (Fig. 2C) was used to capture and pull the wire to the right IJ sheath (with the other side of the wire snared into the portal venous sheath. A 5-French Glide catheter was advanced over the guidewire (which was a 0.018 inch wire). Once it was in the expected location of the portal vein, a buddy 0.035 inch Glide wire microwire was advanced into the superior mesenteric vein under fluoroscopic guidance. Under fluoroscopic guidance, the Glide catheter was advanced over the Glide wire into the superior mesenteric vein and the portosystemic access was secured. Following the measurement of the length of the portosystemic tract, a Viatorr stent with 5 cm covered and a 2 cm uncovered portions) was placed, and the tract was post dilated to a diameter of 8 mm (Fig. 2D). Portal venogram was performed showing robust flow of portal blood through the newly placed stent. The portosystemic gradient dropped to 6 mmHg (Fig. 2E).

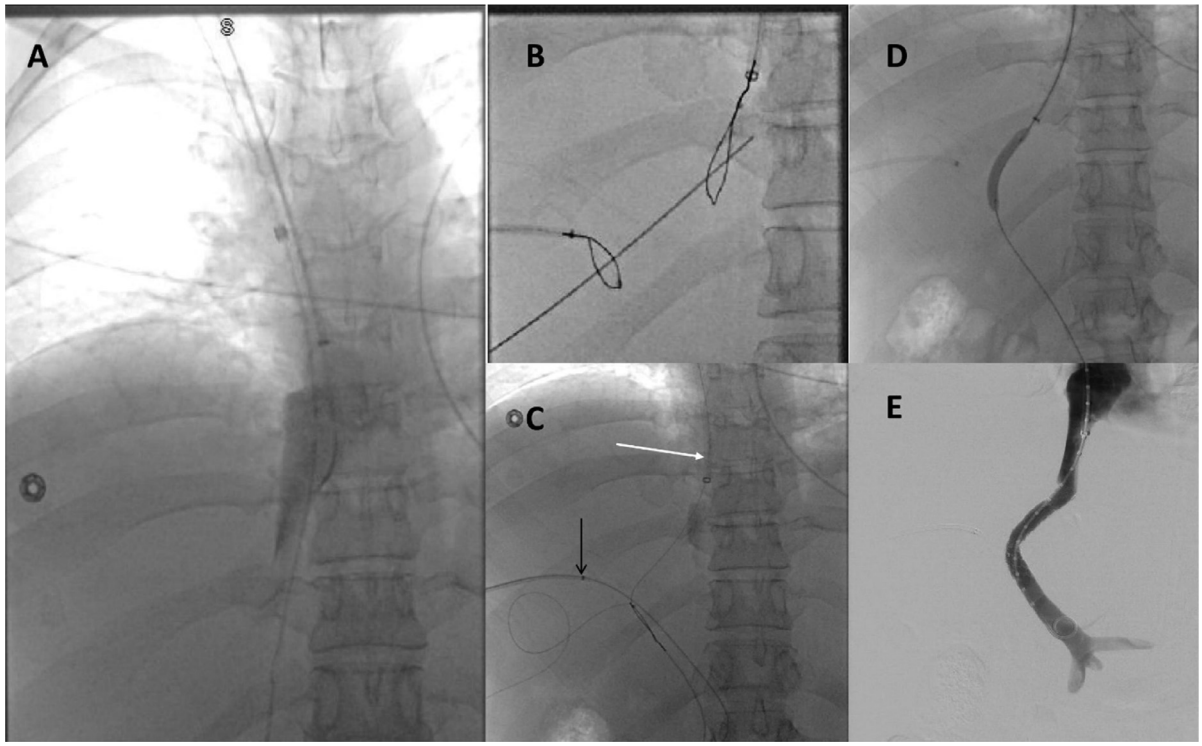

Fig. 2 – (A) Inferior venacavogram was performed showing no evidence of hepatic venous inflow. (B) 20-gauge spinal needle access through 2 snares (1 placed in the right portal vein via a percutaneous vascular sheath, and another in the intrahepatic IVC). (C) Following snaring of the wire, it was pulled into the internal jugular sheath (white arrow), creating a portosystemic access for subsequent stent placement. The percutaneous portal venous sheath is demonstrated (black arrow). (D) Predilation of the portosystemic tract was performed using a 6 mm balloon. (E) Portal venogram following the placement of a Viatorr stent (with 5 cm covered portion and a 2 cm uncovered portion) demonstrating robust flow from the portal venous system to the systemic circulation and right atrium.

After the procedure, the patient had significant clinical improvement. The patient's ascites subsequently resolved. Follow up Doppler ultrasound both 1 week after the procedure and 3 months after the procedure showed a patent stent and no ascites. She continues to be on anticoagulation to this date.

Discussion

BCS occurs in approximately 1/100,000 of the general population. Patients can present with nonspecific signs and symptoms such as abdominal pain, hepatomegaly, and ascites [2]. BCS is more frequently diagnosed in females than in males [5]. Furthermore, ascites is the most common symptom in a retrospective analysis of 44 patients [5]. BCS is a group of disorders that is characterized by the occlusion of hepatic venous outflow that is caused by either structural obstruction or thrombosis leading to complications such as ischemic necrosis and possibly cirrhosis [5]. BCS is often asymptomatic for a long period of time before it progresses to cirrhosis and portal hypertension [6]. If the diagnosis and treatment are not promptly started, there will be significant morbidity associated with BCS with a 3-year estimated mortality rate of up to 89% [7,8]. The main goal of management of BCS is to reduce the risk of mortality and morbidity by decreasing the hepatic congestion [9]. Since BCS can be caused by several etiologies, management of this syndrome depends on many factors such as clinical symptoms and anatomy.

In a patient with high suspicion of BCS, Doppler ultrasonography assists in confirming the diagnosis and is the initial test of choice [10]. Images of hepatic veins with absent flow signal, appearance of spider web/nutmeg pattern, hepatic venous collaterals, and turbulent flow are often indicative of BCS. Moreover, hepatic veins that are not visualized and tortuous are non-specific findings of BCS [2]. Nevertheless, intrahepatic venous collaterals are considered sensitive findings that can be present in up to 80% of BCS cases [2]. MRI is considered a second line imaging modality. This modality can show hepatic vein thrombosis and can be used for evaluation of IVC, but it is more expensive than computed tomography (CT) scanning [2]. 3D-contrast enhanced MR angiography has a comparable sensitivity to hepatic venography [11]. MRI is not effective in demonstrating intrahepatic collateral vessels and showing flow direction [2]. CT is effective for the evaluation of hepatic vascular anatomy when TIPS/DIPS is considered. Non visualization of hepatic veins is considered as an indicator of the disease on CT. However, the false-positive rate can happen in up to 50% of the BCS cases [12]. Hepatic venography is considered the main procedure for evaluation

of hepatic veins, thrombosis extent, and caval pressures [2]. However, the disadvantages of hepatic venography are the use of iodine containing contrast agents. Spider-web pattern is considered the major sign on hepatic venography [2]. Liver biopsy is required to differentiate BCS from Veno-occlusive disease, which occurs following administration of toxic agents and after bone marrow transplantation [13].

Anticoagulants are the cornerstone of treatment for BCS [14]. Some patients with BCS, such as those with acute forms, fail to respond to treatment. In these refractory forms of BCS, a transjugular intrahepatic portosystemic shunt (TIPS) may be used in treatment [15]. This shunt is formed by placing a stent between the portal vein and hepatic vein and is thus an effective method by which to decompress the portal system [16]. However, TIPS is not successful in all patients due to hepatic vein thrombosis and the inability to catheterize the hepatic veins such as in our case. In these patients, direct intrahepatic portocaval shunt (DIPS), a modified TIPS procedure where a stent is directly inserted from the inferior vena cava to the portal vein, is a viable alternative that may alleviate portal hypertension [8]. There are only a few reported cases of BCS patients successfully treated with DIPS procedure to this date [9,17,18].

Techniques for DIPS include the Gun-sight technique such as the one reported in this case report, where portosystemic access is obtained through placing a snare in the IVC (at the level of the caudate lobe) and another in the portal system (using either percutaneous transhepatic or trans-splenic access). The 2 snares are lined up together in the same fluoroscopic plane and a long needle is advanced percutaneously such that access to both snares is established. The vena cava snare is used to pull a wire advanced through the needle into the internal jugular access (with the other side of the wire snared into the percutaneous transhepatic or trans-splenic access). The relatively more expensive intravascular ultrasound can be used to also establish portosystemic access. Additionally, some cases of BCS were managed with percutaneous ultrasound-guided DIPS, which can be achieved if both the portal vein and IVC are visualized sonographically [19].

Angioplasty and stenting seem to be feasible options. Nevertheless, they generally have low applicability in the treatment of patients with BCS as they only prevent the progression to need of TIPS and transplant in one third of BCS patients [20]. Among pediatric patients diagnosed with BCS, the success rates of angioplasty, hepatic vein stenting, and TIPS were 43%, 66%, and 72% respectively [21]. However, these success rates cannot be applied to our patient as there are no available randomized controlled trials for the adult population to this date [9].

Conclusion

Doppler US has proven to be successful in confirming the diagnosis of BCS such as in our patient. DIPS, a modified TIPS procedure, can be used to alleviate portal hypertension in BCS patients with hepatic vein thrombosis where other procedures such as TIPS are contraindicated.

Patient consent

Written informed consent for the publication of this case report was obtained from the patient.

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