Budd–Chiari Syndrome With Spontaneous Intrahepatic Portosystemic Shunts: A Case Series

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Intra-hepatic portal-venous collaterals are characteristic of Budd–Chiari syndrome (BCS) and are usually of small caliber and seen on Doppler. Creation of large portal-systemic shunt, either radiologically (Transjugular intrahepatic porto-systemic shunt) or surgically results in excellent long term outcomes in BCS. Here, we report a series of three rare cases of asymptomatic BCS, who had spontaneous large intra-hepatic portal-systemic shunts. (J CLIN EXP HEPATOL 2019;9:412–415)

Budd–Chiari syndrome (BCS) is defined as an obstruction of hepatic venous drainage due to various causes, leading to progressive liver damage and portal hypertension (PHT). BCS is a rare disease with a potentially dismal outcome, if not treated optimally. It is also important to recognize that BCS is not always a severe disease requiring aggressive treatment.¹ Advent of radiological intervention such as transjugular intrahepatic portosystemic venous shunt (TIPS) results in improved long-term response with fewer complications.² Small intra-hepatic shunts (1 or 2 mm in diameter) between the portal and hepatic veins (HVs) were described in 8% of cirrhotic liver in the injection postmortem study.³ The large intrahepatic venous shunt occurs rarely, whereas extrahepatic shunts such as spleno-gastro-renal shunts are common. A spontaneous portosystemic shunt can lead to recurrent hepatic encephalopathy (HE) due to excessive shunting of toxins such as ammonia into systemic circulation. Spontaneous intrahepatic portosystemic venous shunt (SIPSS) may develop in patients with BCS secondary to PHT. Intrahepatic veno-venous collaterals are common in patients with BCS (up to 80%).⁴ Intrahepatic veno-venous collaterals in patient with BCS drain into systemic circulation

via the subcapsular vessels or drain blood from the occluded segments to the nonoccluded segments of the HV.

In this report, we present three cases of an asymptomatic BCS with intrahepatic portosystemic venous shunt that was diagnosed on computed tomography (CT) or magnetic resonance (MR) abdomen angiography and managed with oral anticoagulation alone.

Case-1

A 20-year-old woman was presented with the right upper abdominal quadrant discomfort for a month, which subsided spontaneously. Clinical examination showed a palpable firm liver, one centimeter below the mid-costal line. Liver function test (LFT) was normal; total bilirubin was 0.8 mg/dL; aspartate aminotransferase (AST), 25 IU/mL; alanine aminotransferase (ALT), 28 IU/mL; serum albumin, 3.6 gm/dL; and international normalised ratio (INR), 1.1. Hepatoportal Doppler showed thrombosis of all three HVs with multiple intrahepatic veno-venous collaterals and patent retrohepatic inferior vena cava (IVC). The liver was irregular and had coarse echotexture and portal vein was patent. CT scan of the abdomen showed an intrahepatic shunt in the form of a single large vessel of constant diameter (size: 20 mm) connecting the right portal vein and IVC (type 1 intrahepatic portosystemic venous shunt, Figure 1A) with nodular configuration of the liver surface traversing through segments 7 and 1 and nonopacification of HVs. Gastroscopy showed small esophageal varices. She had no history of previous liver biopsy, liver trauma, and surgery. She did not have a history of encephalopathy resulting from the large portosystemic shunt. Her thrombophilia workup was negative for any congenital or acquired prothrombotic state. She was started on oral anticoagulation and is being followed up regularly on an outpatient basis and continues to fulfill the response

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Abbreviations: BCS: Budd–Chiari Syndrome; HE: Hepatic Encephalopathy; HV: Hepatic Vein; IVC: Inferior Vena Cava; LFT: Liver Function Test; MTHFR: Methylene tetrahydrofolate Reductase; PHT: Portal Hypertension; SIPSS: Spontaneous Intrahepatic Portosystemic venous shunt; TIPSS: Transjugular intrahepatic portosystemic shunt

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Figure 1 A) A large shunt connecting the right portal vein to the inferior vena cava–type 1 intrahepatic portosystemic venous shunt. (B) A large shunt connecting the right portal vein to the inferior vena cava–type 1 intrahepatic portosystemic venous shunt. (C) An aneurysmal communicating vessel between the right portal vein and the right hepatic vein–type 3 intrahepatic portosystemic venous shunt.

criteria for medical management for BCS (36-month follow-up).⁵ A repeat hepatoportal Doppler after 1 year showed no changes as compared with baseline Doppler findings.

Case-2

A 28-year-old woman presented with fatigue for 6 months and was diagnosed with chronic liver disease and PHT dur-

ing evaluation. She did not have jaundice, ascites, gastrointestinal bleeding, or HE. She had history of splenectomy performed at the age of 5 years for splenomegaly. She had mild pallor. There were no stigmata of liver parenchymal cell failure. Biochemistry reports showed that total bilirubin was 0.9 mg/dL; AST, 37 IU/mL; ALT, 17 IU/mL; serum albumin, 3.5 gm/dL; and INR, 1.28. Ultrasound abdomen showed coarse nodular liver with multiple

veno-venous and portosystemic collaterals, dilated portal vein, and thrombosed HVs. MR venography was performed which suggested HV thrombosis with multiple intrahepatic veno-venous collaterals, nodular liver, portal vein 13 mm, and absence of spleen. A collateral (size: 12 mm) was seen arising from the left branch of intrahepatic portal vein draining into IVC (type 1 intrahepatic portosystemic venous shunt, Figure 1B). Gastroscopy suggested large esophageal varices. Prothrombotic workup performed suggested methylenetetrahydrofolate reductase C667T mutation positive. She was treated with anticoagulation. She followed up every 3 months without any episode of overt encephalopathy (12-month follow-up). There were no interval changes in findings of hepatoportal Doppler after 12 months.

Case-3

A 70-year-old man underwent preoperative evaluation for laparoscopic cholecystectomy for symptomatic gall stones on CT scan in view of recurrent right upper quadrant pain since 4 months. He was found to have spontaneous intrahepatic portosystemic venous shunt during this evaluation. His physical examination and LFTs were normal. His total bilirubin was 0.6 mg/dL; AST, 32 IU/mL; ALT, 29 IU/mL; serum albumin, 4.1 gm/dL; and INR, 1.22. CT scan showed atrophy of the right inferior lobe of the liver and thrombosis of the middle and left HVs. The main portal vein was 14 mm. There was a communicating vessel (size, 12 mm) between the right portal vein and the right HV (type 3 intrahepatic portosystemic venous shunt, Figure 1C). His thrombophilia workup showed factor V leiden mutation. His gastroscopy was normal and did not show varices. He was started on anticoagulation and fulfilled the criteria for medical response (9-month follow-up). Repeat Doppler after 6 months had same findings.

DISCUSSION

Spontaneous portal-systemic shunts within the liver are caused either by a congenital venous malformation or by a collateral pathway due to PHT. In the congenital origin theory, intrahepatic shunt developed because of persistent communication between cranial and caudal hepatic sinusoids.⁶ Hepatic venous outflow tract obstruction leads to the development of PHT. Intrahepatic venous collaterals are a specific diagnostic criterion for BCS.⁴

BCS is considered asymptomatic when there are no signs of abdominal pain, ascites, hepatomegaly, edema, encephalopathy, gastrointestinal bleeding, or a history of any of them.⁷ Asymptomatic BCS accounts for 15–20% of cases. The absence of symptoms is strongly associated with large HV collaterals.⁷ Here, we report three patients of spontaneous portal-systemic shunts with BCS at

different ages. These patients were predominantly asymptomatic, had good hepatic function, and managed with medical therapy alone. Two of these three patients had thrombophilic disorder. The portosystemic shunts were probably decompressing the portal system and resulting in the lack of symptoms and preserved liver function.

Park et al have classified portosystemic shunts into four different morphologic types.⁸ Type 1 is a single large tube of constant diameter that connects the right portal vein to the IVC. Type 2 is a localized peripheral shunt in which single or multiple communications are found between peripheral branches of portal and HVs in one hepatic segment. Type 3 is an aneurysmal communication between the peripheral portal and the HVs. Type 4 shows multiple communications between the peripheral portal and the HVs diffusely in both lobes.

HE occurs in patients with age more than 50 years and patients with cirrhosis and type II shunt.⁹ HE and portal shunt ratio higher than 60%, even without encephalopathy, is an indication for therapeutic intervention in non-cirrhotic patients.¹⁰ Asymptomatic patients with BCS should receive treatment for the potential underlying prothrombotic disease when present like myeloproliferative syndrome, with additional anticoagulation. There is only one case report describing SIPSS in patients with BCS.¹¹ Obliteration of portal-systemic shunts surgically or by interventional radiological techniques is fairly effective in reversing intractable portal-systemic encephalopathy but can be associated with increase in portal pressure and development of ascites and or esophageal varices.^{11–13}

In our three cases of BCS with intrahepatic portosystemic shunt with normal liver biochemistry, we found good short-term outcomes and no changes in the shunts, albeit over a short period of follow-up. It would have been interesting to evaluate the proportion of the size of these spontaneous shunts to the portal circulation. This should be considered in future cases with similar clinical scenarios. Interestingly, none of our patients developed overt HE. We have not evaluated these patients for covert HE. This absence of overt HE, despite spontaneous portosystemic shunts, may be due to mild disease or compensation due to chronicity of the disease process. It is also possible that we have diagnosed these patients early because of better imaging techniques available now, thereby leading to diagnosis of such rare shunts.

To conclude, all asymptomatic patients with BCS should be carefully evaluated for the presence of spontaneous shunts. These patients have good outcomes with medical management alone.

CONFLICTS OF INTEREST

The authors have none to declare.

AUTHOR CONTRIBUTIONS

Pratin Bhatt, Akash Shukla, and Deepak Gupta helped in manuscript writing. Hemant Deshmukh, Krantikumar Rathod, Pratin Bhatt, and Dhiraj Agrawal helped in radiology image retrieval. Pratin Bhatt, Akash Shukla, Deepak Kumar Gupta, Dhiraj Agrawal, Hemant Deshmukh, and Krantikumar Rathod helped in manuscript drafting. All the authors involved in manuscript revision and approval.

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