Asymptomatic Intrahepatic Portosystemic Venous Shunt: To Treat or Not To Treat?

Arkadiy Palvanov, MD, MS¹ Ruth Leah Marder² David Siegel, MD, FSIR¹

¹Radiology Department, North Shore-LIJ Health System, New Hyde Park, New York

²University of Michigan, Ann Arbor, Michigan

Address for correspondence Arkadiy Palvanov, MD, MS, Radiology Department, North Shore-LIJ Health System, 270-05 76th Ave, New Hyde Park, NY 11040 (e-mail: apalvano@nshs.edu).

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Abstract

Keywords

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- ▶ acquired portal vein malformations

Intrahepatic portosystemic venous shunts (IPSVs) are rare vascular malformations. They can be asymptomatic or present with various symptoms including encephalopathy. We present two cases of IPSVS, one involving a patient presenting with altered mental status and the other discovered incidentally. While there is no question that patients presenting with symptomatic IPSVS should undergo definitive treatment, there is no consensus regarding elective therapy for asymptomatic lesions.

Intrahepatic portosystemic venous shunts (IPSVs) are rare hepatic vascular malformations that may be congenital or acquired.^{1,2} The term IPSVS is used specifically to describe a lesion with abnormal communication(s) between the portal and hepatic veins, and four morphological types have been described.³ Type 1, the most common, involves a single large vessel of constant diameter connecting the right portal vein to the inferior vena cava (IVC). In type 2 IPSVS, one or multiple connections are present between peripheral branches of portal and hepatic veins in a single hepatic segment. The shunt is classified as type 3 if there is an aneurysmal connection between portal and hepatic veins. Finally, multiple communications between portal and hepatic veins involving both lobes of the liver is found in type 4 IPSVS. These lesions are usually asymptomatic and are discovered incidentally when ultrasound (US), computed tomography (CT) scan, or magnetic resonance are performed for other reasons. When symptomatic, patients may present with encephalopathy, pulmonary hypertension, or heart failure,

among other manifestations.^{1,4} Patients may be misdiagnosed with psychiatric or neurologic disorders if the IPSVS is not identified as a cause for encephalopathy.⁵

In this manuscript, we describe two cases of IPSVS with different presentations. Our first case involves a patient who presented with encephalopathy and an extremely elevated ammonia level. Our second case, on the other hand, involves a patient who was asymptomatic and in whom IPSVS was an incidental finding on a CT performed for other indications. It has previously been reported and accepted that patients with IPSVS that are symptomatic should be treated. However, the approach toward asymptomatic patients with IPSVS has yet to be widely discussed or agreed upon.

Patient 1

An 80-year-old woman with altered mental status, confusion, weakness, and general lethargy presented to an outside institution, where an abdominal CT was performed

as part of the work-up. An IPSVS was identified on that study (►Fig. 1) and an ammonia level then obtained was elevated at 70 mmol/L (normal range 11–32 mmol/L). She was started on lactulose and transferred to our institution for further work-up of possible hepatic encephalopathy. The patient was seen by the Interventional Radiology Service, and embolotherapy was offered as a treatment option. The patient was taken to the procedure suite after informed consent was obtained from the patient's next of kin. Venographically, multiple channels communicating between portal vein and hepatic vein, consistent with a type 2 IPSVS, were discovered. Successful embolization was achieved using a 16-mm Amplatzer vascular plug deployed at the confluence of all the abnormal communicating branches. Follow-up CT in 2 days demonstrated occlusion of venous-venous malformation with thrombosis of the posterior segment branch of the right portal vein (►Fig. 2). Clinical and laboratory follow-up indicated a normal ammonia level and clinical resolution of the encephalopathy.

Patient 2

A 72-year-old woman who presented for evaluation of bladder pain and recurrent urinary tract infections was found to have a hepatic venous vascular malformation. CT urogram, which was initially performed, revealed a densely enhancing 2-cm lesion in the lateral segment of the left hepatic lobe (►Fig. 3). Subsequent abdominal US clearly demonstrated the morphology of the lesion (►Fig. 4): a fistula between the left hepatic and left portal veins. The patient was not encephalopathic and had no symptoms referable to the shunt. A slightly elevated ammonia level of 37 mmol/L (normal range 11–32 mmol/L) was noted.

Fig. 1 Axial contrast-enhanced CT images demonstrate an IPSVS in an 80-year-old woman who presented with altered mental status, weakness, lethargy, and elevated ammonia level.

Fig. 2 Axial contrast-enhanced CT images status postembolization using a 16-mm Amplatzer vascular plug demonstrate occlusion of venous-venous malformation with thrombosis of the posterior branch of the right portal vein.

The risks, benefits, and alternatives of transvenous embolization were discussed. The option of not undergoing shunt closure and continued clinical and laboratory monitoring for encephalopathy was offered to the patient. The patient decided to proceed with embolization which was performed utilizing a 16-mm Amplatzer vascular plug. Postembolization venography demonstrated successful occlusion of the left portohepatic venous fistula, which was confirmed with follow-up CT (►Fig. 5). Additionally, postoperative laboratory

Fig. 3 Axial contrast-enhanced CT images demonstrate incidentally found densely enhancing 2-cm lesion in the lateral segment of the left hepatic lobe of an asymptomatic 72-year-old woman.

Fig. 4 Grayscale (a) and color Doppler (b) sonographic evaluation of the lesion confirmed a fistulous communication between the left hepatic and left portal veins.

Fig. 5 Axial contrast-enhanced CT images status postembolization using a 16-mm Amplatzer vascular plug demonstrate occlusion of the left portosystemic venous fistula.

tests demonstrated a decrease in ammonia levels to the normal range.

Materials and Methods

In the first case described, the patient was transferred to our institution for embolization of her intrahepatic portohepatic venous shunt due to hepatic encephalopathy. The risks, benefits, and alternatives were explained to the patient's next of kin and consent was obtained. Deep sedation and monitoring were provided by an attending anesthesiologist. Routine right common femoral access was used after standard prep and local anesthesia using 1% lidocaine. Micropuncture technique with US guidance was used. An unremarkable IVC gram was followed by placement of a 6 French curved angiographic guiding sheath (RSR02; Terumo Interventional Systems, Somerset, NJ) into the IVC. A 5 French hydrophilic Cobra catheter (G11994; Cook, Inc., Bloomington, IN) was then advanced through the sheath and the right hepatic vein catheterized over a 0.035-long taper guidewire (GR3507; Terumo Interventional Systems). Venogram showed a large lobulated structure (►Fig. 6), which correlated with the lesion visible on CT. Numerous catheter, guidewire, and microcatheter combinations were used in an attempt to enter the portal venous system, which was finally successful with a 4 French hydrophilic Cobra catheter (G31021; Cook, Inc.). Venography then clearly demonstrated multiple venous channels between the portal vein and hepatic vein (►Fig. 7). While attempted several times, the sheath could not be advanced through the shunt and into the portal vein due to the caliber of the communication branches, but a satisfactory position for vascular plug deployment was achieved at the common point where the several portal venous communicating branches drained. Thus, after removal

Fig. 6 Venogram showing a large lobulated structure in the right hepatic lobe, which correlates to the lesion demonstrated on CT (►Fig. 1).

Fig. 7 Venography demonstrating multiple venous channels between the portal vein (long blue arrow) and hepatic vein (short red arrow).

Fig. 9 Postembolization hepatic venogram demonstrating no abnormal communication between the portal and hepatic veins.

of the inner sheath dilator and guidewire, two 16-mm Amplatzer II plugs (9-AVP2-016; St Jude Medical, Inc., St. Paul, MN) were sequentially advanced through the sheath and deployed (►Fig. 8). Postembolization hepatic venogram was performed (►Fig. 9) and the catheter was removed. Approximately 100 mL of nonionic contrast (Isovue-370; Bracco Diagnostics Inc., Monroe Township, NJ) was used.

In the second case, the patient was electively seen in the Interventional Radiology clinic and decided to proceed with embolization after discussing the risks, benefits, and alternatives of the procedure. As in the first case, intravenous sedation was provided by an anesthesiologist. After obtaining

Fig. 8 Fluoroscopic images demonstrating two 16-mm Amplatzer plugs deployed at the portal-hepatic venous communication.

informed consent, the right femoral vein was accessed and an unremarkable IVC study performed, using the same technique as in the first case. A 5 French Cobra catheter (G11994; Cook, Inc.) was then advanced into the left and subsequently the middle hepatic vein and their major identified branches, but the lesion could not be catheterized or identified venographically. It was felt that the fistula was probably quite proximal, and while attempted with numerous catheters, in conjunction with the curved sheath, a stable position at the insertion of the left hepatic vein into the IVC could not be maintained. Therefore, a 0.035-inch Rosen guidewire was advanced distally into a left hepatic vein branch to stabilize and maintain the position of the 6 French sheath tip. Venography was performed in anteroposterior and lateral projection, through the sheath, which revealed flash filling of the fistula (►Fig. 10) draining into the left hepatic vein just proximal to its insertion into the IVC. A microcatheter/microguidewire combination (MC-PE28131YB Terumo Interventional Systems), which was preloaded into the 5 French Cobra catheter, was advanced through the sheath while the Rosen “buddy” wire stabilized the sheath tip position. The microcatheter was then advanced into the fistula, injection performed (►Fig. 11), and then advanced into the left portal vein. The Rosen wire was then removed to allow room within the sheath for the Cobra catheter, which was then advanced into the left portal vein over the microcatheter. Venography was performed after removal of the microcatheter, clearly demonstrating the portohepatic venous fistula (►Fig. 12). The sheath was advanced into the right portal vein over a Rosen wire which had been placed through the cobra catheter and venography performed through the sheath, followed by embolization of the portohepatic venous fistula utilizing a 16-mm Amplatzer vascular plug (9-Plug-016; St Jude Medical, Inc.). Postembolization venography (►Fig. 13) showed no flow through fistula, indicating a successful embolization, and

Fig. 10 Venogram demonstrating a flash filling of the fistula (long yellow arrow) draining into the left hepatic vein (short red arrow) just proximal to its insertion into the IVC (blue arrowhead).

the sheath was removed. Approximately 140 mL of nonionic contrast (Isovue-370; Bracco Diagnostics Inc.) was used.

Discussion

IPSVS are rare hepatic vascular malformations which are usually congenital, but they may also be acquired due to portal hypertension or iatrogenic causes. Congenital IPSVS may present in early infancy with neonatal hyperammonemia, cholestasis, and/or hypergalactosemia. In some

Fig. 12 Venogram after removal of the microcatheter clearly demonstrates the portohepatic venous fistula (red arrow).

cases, the portosystemic shunts may close spontaneously within the first 2 years of life, or it may remain asymptomatic. If the shunt persists to adulthood, patients can present with encephalopathy, pulmonary arterial hypertension, and/or heart failure.^{1,2} It has been suggested that portosystemic shunting should be considered in the differential diagnosis of any patient with hyperammonemia.⁶ When diagnosed, periodic laboratory evaluations are important because of the ongoing potential risk of hepatic encephalopathy due to exposure to high ammonia levels.⁷ While most asymptomatic IPSVS are discovered incidentally as in

Fig. 11 Fluoroscopic image demonstrating advancement of microcatheter into the fistula, followed by advancement into the left portal vein.

Fig. 13 Postembolization venogram demonstrating no flow through portohepatic venous fistula indicating a successful embolization.

our case, if suspected, color Doppler sonography, in conjunction with pulsed Doppler, is highly sensitive and specific.⁸

Successful treatment of symptomatic shunts associated with hyperammonemia has been widely reported.^{5,9,10} Several case involving children and adults, including our case, demonstrate resolution of hyperammonemia and encephalopathy with successful closure of the IPSVS.^{4,9–11} It is clear that endovascular closure has become the standard of care for the treatment of symptomatic IPSVS. One caveat is in cases with a preprocedure portal pressure measurement above 32 mm Hg, due to the risk of developing significant acute portal hypertension. In these cases, gradual closure in two stages is recommended; and surgical banding of the shunt followed by endovascular embolization was suggested in one report.⁴

The more difficult situation is that of the asymptomatic patient. Although transfemoral transcatheter embolization with coils or other mechanical occluders is a relatively safe procedure, complications do occur including infection, hemorrhage, portal vein occlusion, and portal hypertension, among others.¹² Risk/benefit analysis is difficult, due to the unknown natural history of an IPSVS that has remained asymptomatic into adulthood. Several reports have shown that IPSVS can be a cause of hepatic encephalopathy and the incidence of hepatic encephalopathy has been found to increase with age.^{5,11} Unlike most other congenital diseases, IPSVS may not be recognized for several decades due to the time it takes to develop hepatic encephalopathy, which is believed to develop after middle age, and most patients do tend to be asymptomatic while young.¹³ But the question remains, should these patients be offered a purely prophylactic procedure? This question is really the impetus for the publication of this case report. As we have mentioned earlier, the entity of IPSVS and their endovascular treatment have been described previously; however, the approach to the asymptomatic patient and the question of whether a prophylactic procedure should be offered have never been discussed in the literature.

As in many situations without clear guidelines or evidence-based literature, we believe the patient should be involved in this decision. The basics of the procedure, its potential risks and benefits, and, as in our case, an honest discussion that includes the strong possibility that the patient could remain asymptomatic for life needs to take place. Lee et al suggest that calculating shunt ratio can be valuable. This is accomplished using Doppler US, by dividing the blood flow volume at the shunt orifice by the total portal blood flow volume. The risk of encephalopathy increases with increasing shunt ratios. If this ratio exceeds 60%, they suggest treatment is needed, even without hepatic encephalopathy.^{9,14} In our asymptomatic case, serial Doppler sonography was declined by the patient who opted for definitive management. The authors feel this is reasonable based on the relatively low procedure-related morbidity. Additionally, it is our feeling that if the procedure could not be completed using standard technique, no additional procedural risk should be taken. For instance, if for some reason, shunt closure could not be achieved from the femoral approach, a transhepatic portal approach, with its added inherent risks, would not be warranted in an asymptomatic adult patient.

Conclusion

IPSVS are rare hepatic vascular malformations. Although usually asymptomatic and discovered incidentally on imaging, symptomatic IPSVS may present with hyperammonemia and encephalopathy, among others symptoms. We described two cases of IPSVS, one involving an encephalopathic 80-year-old patient and the other an asymptomatic 72-year-old, whose shunt was incidentally discovered. While widely accepted that patients with symptomatic IPSVS should be treated, there is no consensus on how to approach asymptomatic patients with IPSVS. In light of the observation that encephalopathy increases with increasing age, we believe that treatment of asymptomatic IPSVS, if straightforward, should be offered to otherwise healthy patients, regardless of age, as an alternative to clinical and Doppler ultrasound surveillance.

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