Case Report

Transhepatic embolization of a congenital intrahepatic portosystemic shunt for the treatment of hepatic encephalopathy in a noncirrhotic patient using Amplatzer vascular plug device

Rachel Ann Brader BSa, Kyung Rae Kim MDb, *

a University of North Carolina at Chapel Hill School of Medicine, Chapel Hill, NC, USA
b Vascular & Interventional Radiology, University of North Carolina at Chapel Hill School of Medicine, 2016B Old Clinic, Campus Box 7510 Chapel Hill, NC 27599, USA

ABSTRACT

A 73-year-old male with no history of liver disease was hospitalized for weakness, confusion, ataxia, and new onset hepatic encephalopathy with hyperammonemia. After management with lactulose and rifaximin, his symptoms persisted, and he underwent transjugular liver biopsy. Biopsy showed normal liver, but a portosystemic shunt was incidentally identified on postbiopsy venogram. The patient underwent occlusion of the shunt with two Amplatzer vascular plugs and four Nester coils. Following embolization, the patient’s symptoms resolved completely. Our case reports one of the oldest adults to present with symptoms from a congenital portosystemic shunt. Congenital portosystemic shunts can be considered in patients with new onset hepatic encephalopathy in the absence of underlying liver disease. Prognosis after embolization of congenital portosystemic shunt is great, and embolization may result in full reversal of symptoms.

© 2016 the Authors. Published by Elsevier Inc. under copyright license from the University of Washington. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

Case Report

A 73-year-old man with no history of liver disease presented to our hospital in July 2015 with weakness, confusion, asterixis, and ataxia. His additional medical history was significant for congestive heart failure, hypertension, and atrial fibrillation. He had been recently seen by an outpatient physician for similar symptoms and had a workup that included a normal vitamin B12, folate, antinuclear antibody, human immunodeficiency virus, rheumatoid factor, cortisol, and thyroid-stimulating hormone. Laboratory studies during his admission showed an elevated serum ammonia (117 umol/L), hyperbilirubinemia (2.4mg/dL), and elevated INR (3.7). He was hospitalized and seen by gastroenterology and neurology. He had no history of trauma in the liver. After an extensive workup, his encephalopathy was poorly explained and thought to be due to hepatic congestion caused by congestive heart failure and cirrhosis after a remote history of alcohol
abuse 40 years prior. He was started on lactulose and rifaximin and noted some improvement of his symptoms. The patient continued to have increasing difficulty ambulating, slurred speech and generalized weakness and was followed by a hepatologist following discharge from the hospital. His laboratories and physical examination findings were not consistent with cirrhosis, so it was suggested that he undergoes a transjugular liver biopsy with hepatic venous pressure measurement to further characterize his liver disease.

A transjugular liver biopsy was done in November 2015, and during the procedure, corrected wedge pressures were found to be 5–6 mm Hg, biopsy samples were taken, and a venous abnormality was noted on postbiopsy hepatic venogram (Fig. 1). The lesion was characterized as a dilated area of hepatic vein in the right lobe and communicating with the right portal vein, and further imaging was recommended.

Pathology report of the biopsy showed normal liver with minimal nonspecific portal chronic inflammation and no evidence for congestive hepatopathy. Follow-up CT of the abdomen showed hepatic segment V/VI portosystemic shunt with a likely vascular malformation measuring 2.6 cm (Fig. 2).

Given the patient’s history of normal biopsy and hepatic vein pressures, lack of trauma, and absence of malignancy, the malformation was thought to be congenital. At the age of 73 years, our case reports one of the oldest patients to present with symptoms from a congenital intrahepatic portosystemic shunt.

The case was evaluated by the interventional radiology department and was scheduled for a percutaneous transhepatic embolization of the intrahepatic portosystemic shunt. Ultrasound guidance was used to access the right portal vein, and a 6-Fr MAK-NV introducer system (Merit Medical, South Jordan, UT) was used to place a 6-Fr × 25 cm sheath (Terumo, Elkton, MD). During the procedure, contrast injection portal venogram using a 5-Fr Omni-flush catheter (Angiodynamics, Queensbury, NY) demonstrated direct fistula from the distal right posterior portal vein branch to the right hepatic vein (Fig. 3).

Given the risk of coil migration due to high-flow fistula from the portal vein to the hepatic vein, an Amplatzer vascular plug was used. An 8-mm Amplatzer 4 vascular plug (AGA Medical Corporation, Plymouth, MN) was advanced through the 5-Fr Simmons-1 catheter (Angiodynamics, Queensbury, NY) into the one of the two feeding portal vein branches.

---

Fig. 1 – Transjugular hepatic venogram demonstrates a dilated area (arrowhead) of right hepatic vein (white arrow) communicating with the right portal vein (black arrow.)

Fig. 2 – (A) Venous phase axial CT scan demonstrates an aneurysmal connection (arrow) between the right portal and right hepatic veins in the segment VI. (B) Superior slice of the CT scan depicts the right posterior portal vein (white arrow) and the right hepatic vein (black arrow) converging into the aneurysmal connection.
branches (more lateral branch), and the plug was detached. Repeat portal venogram demonstrated decreased, but persistent flow to the fistula and another 8 mm Amplatzer vascular plug was detached in the portal vein immediately proximal to the first plug. Repeat portal venogram demonstrated decreased flow through the more lateral feeding portal vein branch and significant flow to the more medial feeding portal vein branch (Fig. 4).

Given the plugs covered the orifices of the venous fistula and no risk of coil migration, coils were placed subsequently. A total of four, 0.035 inch, 8 mm × 14 cm Nester coils (Cook, Bloomington, IN) were placed proximal to the plugs. There was a small fistula from the right anterior portal vein to the right hepatic vein; however, given too small size of the feeding vessel, it was decided not to embolize the right anterior portal vein branch.

Final portal venogram demonstrated complete blockage of the fistula and no evidence of fistula flow from the right posterior portal vein to the right hepatic vein. The right posterior portal vein proximal to the plugs and coils was patent with no evidence of thrombosis. The right anterior portal vein and the left portal vein were also patent with no evidence of thrombosis (Fig. 5).

Following the procedure, the patient experienced complete resolution of his encephalopathy, correction of his laboratory abnormalities, and was able to discontinue his use of lactulose and rifaximin.

**Discussion**

The cause of hepatic encephalopathy is believed to be due to neurotoxic substances, like ammonia, in the presence of inflammatory mediators though the exact mechanism is unknown. While ammonia contributes to hepatic encephalopathy, the severity of symptoms often does not correlate with the elevation of ammonia. Alterations in mental status and gross motor disturbances are the hallmark symptoms of hepatic encephalopathy.

Hepatic encephalopathy can be due to intrinsic hepatic failure or spontaneous portosystemic shunts [1-3]. Cirrhosis is the most common cause of spontaneous portosystemic...
shunts via portal hypertension and development of portosystemic collaterals or through sinusoidal fibrosis leading to intrahepatic portosystemic shunting [4]. A very small proportion of portosystemic shunts are not related to underlying cirrhosis and can be the result of congenital venous malformations or spontaneous venous malformations.

Congenital portal venous malformations can be extrahepatic or intrahepatic, with intrahepatic shunts being much less common. Intrahepatic shunts have been categorized into four types: type 1—connection between the right portal vein to the inferior vena cava; type 2—shunt between peripheral branches of portal and hepatic veins in one hepatic segment; type 3—aneurysmal connection between portal and hepatic veins; and type 4—multiple connections between peripheral branches of portal and hepatic veins in both hepatic lobes [5-7]. The origin of these shunts is not well elucidated, but proposed mechanisms include persistent embryonic venous anastomoses and rupture of portal vein aneurysms into the hepatic vein [5,8]. Embryologically, derivatives of the vitelline veins to differentiate into the inferior vena cava, hepatic veins, and portal vein, while derivatives of the umbilical veins terminate in the sinus venosus and largely degenerate when they come in contact with hepatic sinusoids. When a segment of the liver fails to generate sinusoid formation, there may be persistent communication between the vitelline veins of the omphalomesenteric system and the sinus venosus [5,6].

Though congenital shunts are present from birth, hepatic encephalopathy remains relatively rare and is often not diagnosed until adulthood. One proposed mechanism for the delay in the development of hepatic encephalopathy in congenital shunts is that the adult central nervous system is more sensitive to hyperammonemia. Shunt ratio also seems to play a role in the development of hepatic encephalopathy in patients with congenital shunts, with ratios greater than 60% showing an increased risk for hepatic encephalopathy [5]. In patients with undiagnosed shunts, hepatic encephalopathy may be the presenting symptom. The prevalence of portal-systemic encephalopathy in the absence of liver disease is rare, and no studies have been published in the United States examining the prevalence. One study in Japan reported 47 cases of hepatic encephalopathy in the absence of underlying liver disease in the year 2000 [9]. Another study found that an additional 24 cases have been reported worldwide through 2015 [10].

Treatment of hepatic encephalopathy associated with portosystemic bypass and no intrinsic liver disease can be medical or procedural. Medical management is similar to management of hepatic encephalopathy seen in cirrhotic patients and includes lactulose or nonabsorbable oral antibiotics [9]. Surgical occlusion of shunts is also successful but is very invasive. Percutaneous obliteration of shunts causing refractory hepatic encephalopathy is often performed in a minimally invasive manner. Embolization with ethanol, coils, balloon retrograde embolization, and Amplatzer vascular plugs have all been reported as effective means of shunt closure.

Currently, the use of Amplatzer vascular plugs and coils is the most common methods of embolization. The use of an Amplatzer vascular plug offers several benefits such as the ability to deploy and check positioning and reposition as needed before releasing the plug into its final position [11]. Using an Amplatzer device prior to deploying coils is effective because there is reduced a risk of migration of the coils and more effective shunt closure than use of Amplatzer device alone. Time to occlusion is also very short with use of Amplatzer device as compared to coils alone allowing for thorough evaluation of the shunt occlusion during the procedure. It is also superior to embolization with ethanol because you do not risk hemolysis and pulmonary edema [10].
One literature review found that embolization of porto-systemic shunts may be more effective in patients without underlying cirrhosis and noted that of the patients undergoing embolization for shunts without underlying liver disease, all remained symptom free at 12 months \[10\]. A European study reported 41% recurrence rate of hepatic encephalopathy after embolization of portosystemic shunts in patients with underlying liver disease \[12\], and a more recent US study showed a recurrence rate of 8% at 1 year \[11\]. Our patient has remained symptoms free after intervention but has not reached 1 year since his procedure.

Hepatic encephalopathy due to portal-systemic bypass in patients without underlying liver disease is a well described and potentially reversible cause of encephalopathy. It is important that physicians recognize hepatic encephalopathy in non-cirrhotic patients and attempt to accurately diagnose the cause of their hepatic encephalopathy in order to best treat their patients. Prognosis following interventional radiology occlusion of shunts with Amplatzer vascular plugs has been reported to be very good. In several cases such as ours, patients experienced resolution of hepatic encephalopathy without return of symptoms.

REFERENCES

[1] Dharel N, Bajaj JS. Definition and nomenclature of hepatic encephalopathy. J clin Exp Hepatol 2015;5:S37–41.
[2] Eroglu Y, Byrne W. Hepatic encephalopathy. Emerg Med Clin North Am 2009;27:401–14.
[3] Saad W. Portosystemic shunt syndrome and endovascular management of hepatic encephalopathy. Semin Intervent Radiol 2014;31:262–5.
[4] Tapper E, Gordon Jiang Z, Patwardha V. Refining the ammonia hypothesis: a physiology-driven approach to the treatment of hepatic encephalopathy. Mayo Clinic Proc 2015;90:646–58.
[5] Gallego C, Miralles M, Marin C, Muyor P, Gonzalez G, Garcia-Hildago E. Congenital hepatic shunts. Radiographics 2004;24:755–72.
[6] Stringer MD. The clinical anatomy of congenital portosystemic venous shunts. Clin Anat 2008;21:147–57.
[7] Park JH, Cha SH, Han JK, Han MC. Intrahepatic portosystemic venous shunt. AJR 1990;155:527–8.
[8] Remer EM, Motta-Ramirez GA, Henderson JM. Imaging findings in incidental intrahepatic portal venous shunts. AJR 2007;188:162–7.
[9] Watanabe A. Portal-systemic encephalopathy in non-cirrhotic patients: classification of clinical types, diagnosis and treatment. J Gastroenterol Hepatol 2000;15:969–79.
[10] Asakura T, Nobutake I, Takahiro S, Nobuaki M. Portosystemic encephalopathy without liver cirrhosis masquerading as depression. Intern Med 2015;54:1619–22.
[11] Lynn A, Singh S, Congly S, Khemani D, Johnson D, Wiesner R, et al. Embolization of portosystemic shunts for treatment of medically refractory hepatic encephalopathy. Liver Transpl 2016;22:723–31.
[12] Laleman W, Simon-Talero M, Maleux G, Perez M, Ameloot K, Soriano G, et al., for EASL-CLIF- Consortium. Embolization of large spontaneous portosystemic shunts for refractory hepatic encephalopathy: a multicenter survey on safety and efficacy. Hepatology 2013;57:2448–57.