Hepatic encephalopathy due to intrahepatic portosystemic venous shunt successfully treated by balloon occluded retrograde transvenous embolization with GDCs

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Abstract

We report a 65-year-old man with hepatic encephalopathy due to an intrahepatic portosystemic venous shunt that was successfully occluded by balloon occluded retrograde transvenous embolization with Guglielmi and interlocking detachable coils as performed percutaneously.

Keywords: Hepatic veins, interventional procedures, portal vein, therapeutic blockade, shunts, portosystemic

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Although reports have been rare since the first report by Raskin et al. in 1964 (1), intrahepatic portosystemic venous shunt (IPSVS) has become encountered more frequently in parallel with the development of various imaging modalities such as ultrasonography and computed tomography (CT) (2–4). However, even now, IPSVS is not widely known and is sometimes overlooked. When an IPSVS causes hepatic encephalopathy, treatment is required (3), and transcatheter embolization has been used to treat symptomatic IPSVS (5–8). We present such a symptomatic case and its treatment by balloon occluded retrograde transvenous embolization with Guglielmi and interlocking detachable coils (GDCs; Boston Scientific, Watertown, MA, USA).

Case report

A 65-year-old man was referred with a history of repeated episodes of unconsciousness. The patient had no history of trauma, liver biopsy, or liver disease. Gastrectomy had been performed for gastric cancer 3 years previously, and at the time of admission the patient was free from tumor. A review of the entire family tree revealed no inherited disorder. Laboratory studies showed no liver dysfunction with the exception of a mild increase in the serum ammonia level (147 μg/dl; normal range, <79 μg/dl). Contrast-enhanced multidetector-row CT revealed a large abnormal vessel in communication with a peripheral right portal branch and the right hepatic vein, which suggested the existence of an IPSVS. The shunt was 15 mm in diameter. These findings confirmed that a hepatic encephalopathy caused by an IPSVS was responsible for the repeated episodes of unconsciousness. Therefore, we attempted embolization of the IPSVS.

After the patient provided written informed consent, the following procedures were performed with the patient under local anesthesia. First, a 5-F cobra-shaped catheter (Terumo Clinical Supply, Gifu, Japan) was inserted from the right femoral artery. Then, superior mesenteric arterial portography was performed, which confirmed the presence of an IPSVS forming a communication between the right portal vein and the right hepatic vein. Next, a 6-F sheath introducer (Medikit, Tokyo, Japan) was inserted from the right femoral vein after which a 6-F hook-shaped guiding catheter (Terumo Clinical Supply, Gifu, Japan) was inserted from the right femoral artery. Then, superior mesenteric arterial portography was performed, which confirmed the presence of an IPSVS forming a communication between the right portal vein and the right hepatic vein. Next, a 6-F sheath introducer (Medikit, Tokyo, Japan) was inserted from the right femoral vein after which a 6-F hook-shaped guiding catheter (Terumo Clinical Supply) was advanced to the right hepatic vein. The shunt was 15 mm in diameter. These findings confirmed that a hepatic encephalopathy caused by an IPSVS was responsible for the repeated episodes of unconsciousness. Therefore, we attempted embolization of the IPSVS.

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catheter was inflated to decrease hepatofugal blood flow in the intrahepatic portosystemic venous shunt and to avoid migration of the coils to the systemic venous site. Injection of the contrast medium via the micro-catheter advanced into the portal vein (arrows) shows one connection between one of the right portal branches and the right hepatic vein (large arrow). (b) Superior mesenteric arterial portography after the procedure showed no IPSVS. Note that the IPSVS is sufficiently occupied by the GDCs (arrows) and that the micro-balloon catheter positioned in the right hepatic vein has been deflated (arrowhead).

Fig. 1  (a) Injection of the contrast medium via the micro-catheter advanced into the portal vein (arrows) shows one connection between one of the right portal branches and the right hepatic vein (large arrow). (b) Superior mesenteric arterial portography after the procedure showed no IPSVS. Note that the IPSVS is sufficiently occupied by the GDCs (arrows) and that the micro-balloon catheter positioned in the right hepatic vein has been deflated (arrowhead).

Currently, 14 months after this interventional procedure, the patient has no symptoms of hepatic encephalopathy and his clinical condition is good.

Discussion

From a review of the literature on IPSVS, Park et al. (3) categorized IPSVSs into four different morphologic types: (i) single large tube of constant diameter that connects the right portal vein to the inferior vena cava; (ii) localized peripheral shunt in which single or multiple communications are found between peripheral branches of portal and hepatic veins in one hepatic segment; (iii) connection between peripheral portal and hepatic veins through an aneurysm; and (iv) multiple communications between peripheral portal and hepatic veins diffusely in both lobes. The present case had features of Park’s type II.

The cause of IPSVS has been controversial. Two theories, the congenital origin theory and the acquired theory, have been reported (1, 3). The former suggests a persistent embryonic venous anastomosis, such as the ductus venosus or a vitelline vein (1) while the latter suggests that the shunt results from portal hypertension, trauma, or rupture of a portal vein aneurysm (4).

Treatment is required for patients with symptoms of hepatic encephalopathy after increased blood flow through the shunt (3). Traditionally, surgical removal of the shunt was used to alleviate symptoms (1). In recent years, however, less invasive treatments using interventional techniques such as transcatheter embolization have been widely accepted in parallel with the rapid development of various interventional instruments such as micro-catheters, imaging modalities, and embolic agents (5–8). For multiple IPSVSs, the mesenteric venous route through a small abdominal incision often has been selected (5, 6), whereas for a single IPSVS or a few IPSVSs the less invasive percutaneously trans-hepatic (5, 7) or trans-caval approach (5, 8) has been chosen. When sufficient embolization of IPSVS by a single approach is difficult, the use of two routes was reported to be effective (5).

Stainless steel coils or micro-coils have been widely used for embolism (5, 7, 8). However, when the shunt is large and blood flow in the shunt is very rapid it has been considered technically difficult to achieve shunt occlusion with coils because of the limitations of conventional coils. Once pushed from the cartridge into the introducer catheter, none of the coils can be withdrawn; therefore, the only way to abort the embolization is to withdraw the entire catheter. Another limitation is that once a coil emerges from the catheter tip, a stage of irreversibility is soon reached as catheter withdrawal means coil deployment (9). On the other hand, the GDC, which is an electrical detachable coil and was used in the present case, has the advantage of permitting accurate coil positioning (9). The coil can be freely advanced or withdrawn before its final release. If a coil is positioned incorrectly, it can be withdrawn and reinserted. In addition, GDCs are available in a wide range of lengths and diameters, which may support and extend their application to lesions with large or high-flow shunts.
In the present case, we succeeded in occlusion of the IPSVS safely through the trans-caval approach using GDCs after decreasing blood flow in the shunt by inflating the balloon catheter positioned in the hepatic vein, which communicated with a portal venous branch through the IPSVS. To our knowledge, the successful embolization of an IPSVS with GDCs performed with flow in the shunt carefully controlled using a micro-balloon catheter has not been reported in the medical literature in the English language. Indeed, that technique might seem somewhat complicated, especially in relation to the newly developed occlusion devices like the Amplatzer vascular plug (10). However, in countries in which such devices have not become commercially available, such as in Japan, our method might be considered.

In conclusion, balloon occluded retrograde transvenous embolization with GDCs would be among the choices for endovascular treatment of IPSVS.

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