CASE REPORT

Extrahepatic portosystemic shunt after liver transplantation. Percutaneous embolization for hepatic encephalopathy

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Introduction

Portal-systemic encephalopathy (PSE) is caused by substances originating in the intestine that directly enter the general circulation without passing through the liver, as in extrahepatic portosystemic shunt (EHPS). This shunt is usually a sequel of cirrhosis and severe portal hypertension.1 It is seldom observed in neurological complications without portal hypertension and chronic liver disease.2–4

The aetiology of portosystemic shunt may involve other rare causes, such as congenital shunts,2,3,5 iatrogenic shunts (after abdominal operation or liver biopsy),5,6 trauma and idiopathic shunts.2,7 The literature includes only a few cases of portosystemic shunt not associated with portal hypertension,2,7,8 mostly in Western countries.

The occlusion of shunts is sometimes indicated for PSE.6–9 Recently, good results have been reported for the percutaneous approach using interventional radiology in cases of EHPS occlusion, via the liver or the inferior vena cava.5

We report a case of a 54-year-old woman presenting with one episode of PSE 27 months after undergoing liver transplantation, without recurrence of cirrhosis or portal hypertension. The portosystemic shunt was diagnosed by abdominal colour-Doppler and magnetic resonance image angiography (MRIA). Spectroscopic cranial MR confirmed PSE. This shunt was successfully treated with percutaneous vascular embolization.

In summary, PSE may occur many years after liver transplantation, even in the absence of portal hypertension and liver fibrosis, and may be due to old shunts not eliminated during transplant surgery. Percutaneous embolization can achieve good results. We report a case where the portosystemic shunt occurred between the superior mesenteric vein and the left ovarian vein. To the best of our knowledge, there is only one previous report of percutaneous embolization of portosystemic shunts after liver transplantation.10

Case report

A 54-year-old woman underwent liver transplantation at our unit for decompensated cirrhosis due to chronic alcoholism. The early postoperative course was complicated by intra-abdominal bleeding at day 5, prompted by abdominal drain removal and requiring further surgery.

Pretransplantation abdominal imaging included colour Doppler sonography and MRIA, and showed a collateral vessel draining from the superior mesenteric vein into the right ovarian vein (Fig. 1). During the transplantation no attempt was made to interrupt this large vessel. Routine intraoperative colour Doppler evaluation was made after the graft reperfusion, and indicated a 24 cm/s mean rate velocity of portal vein hepatopetal flow.

The first postoperative months were uneventful, except for an episode of cytomegalovirus (CMV) pneumonitis treated with 21 days of intravenous ganciclovir (DHPG), with resolution. The woman
had a regular follow-up with blood samples and colour Doppler sonography. Serum chemistry was repeatedly normal, sonography showed no abnormal vessels, and a 56 cm/s mean rate flow in the main portal vein was noted.

At 24 months, colour Doppler sonography showed an abnormal vessel (2.0 cm in diameter) running to the superior mesenteric vein, on the right side of the abdomen, raising the suspicion of a portal vein stenosis at the level of the anastomosis and extrahepatic portal hypertension. MRIA clearly demonstrated that this vessel originated from the superior mesenteric vein and communicated with the right ovarian vein, as in the pretransplantation image. Histology of the liver demonstrated a normal portal triad, without fibrosis. Upper tract endoscopy showed no varices.

Percutaneous transhepatic portography was carried out in an attempt to perform a balloon dilatation of suspected portal vein stenosis. The portography showed no stenosis and 12 mmHg portal vein pressure, both below and above the level of the anastomosis (without gradient pressure), and balloon dilatation was not attempted.

The woman continued close follow-up on an outpatient basis. Three months later she was admitted to the hospital complaining of memory lapses and sleeping difficulties for 1-month. The neurological symptoms included confusion and abnormal behaviour, progressing to coma. Laboratory investigations demonstrated normal chemistry; brain CT and lumbar puncture showed no anomalies. Spectroscopic cranial MR confirmed PSE with high signal in globus pallidus T1-weighted images (Fig. 2). The woman received oral lactulose and neomycin, and improved after 48 h.

As colour Doppler sonography demonstrated a large varix on the superior mesenteric vein to the right side of the abdomen, percutaneous vascular embolization was considered suitable. The shunt was performed percutaneously, via the right femoral vein (Fig. 3), and two stainless coils were successfully delivered through the right ovarian vein into the large collateral vein (Fig. 4).

The postoperative course was uneventful, and colour Doppler sonography revealed disappearance of the shunt flow. The woman was discharged at day 4, and 14 months postoperatively repeated colour Doppler sonography showed complete obstruction of the shunt. There were no symptoms of encephalopathy and no medical therapy was required.

Figure 1 Magnetic resonance imaging angiography, showing a collateral vessel (open arrow) draining from the superior mesenteric vein (short arrow) into the right ovarian vein (white arrow).

Figure 2 Spectroscopic cranial magnetic resonance confirmed portosystemic encephalopathy with high signal in globus pallidus T1-weighted images.
Discussion

One of the complications of end-stage liver disease is hepatic encephalopathy. This neurological condition is almost always due to portosystemic venous shunts, caused by portal hypertension and impaired liver function secondary to progressive hepatic fibrosis in cirrhotic individuals. There have also been reports of a few cases in which “hepatic” encephalopathy developed in non-cirrhotic patients without portal hypertension; here the term “hepatic” might not be appropriate. Watanabe,2 in an extensive review of these cases of encephalopathy in non-cirrhotic patients, used the distinctive term “portal-systemic encephalopathy” rather than “hepatic encephalopathy”. Some of these cases of encephalopathy without liver cirrhosis have been misdiagnosed as psychoneurological disorders.

Portosystemic shunts may be located inside or outside the liver. The intrahepatic type occurs between the portal and hepatic veins. This is sometimes congenital, accompanied by anomalies of the portal system such as Rendu-Osler-Weber disease, or a patent venous duct between the left branch of the portal vein and the left hepatic vein.

Intrahepatic acquired shunts may be due to trauma, biopsy or surgery. The extrahepatic portosystemic shunts may also be congenital in the rare cases of portal vein absence (when the superior mesenteric vein runs to the inferior vena cava). Many extrahepatic shunts occur between the splenic, the left gastric or superior mesenteric vein and the left renal vein. In exceptional cases, the shunt passes through ovarian or testicular veins.

In liver transplantation large extrahepatic portosystemic shunts are of major significance and are found at preoperative angiographic assessment in up to 60% of cirrhotic patients.11 These shunts can steal portal flow after the graft implantation, and may account for graft hypoperfusion, poor graft function and portal vein thrombosis. However, sometimes the “steal” phenomenon may be present solely after an acute rejection episode.11,12 These
collateral veins should be divided during the liver transplantation procedure.

In 1995 Sezai et al. described the first case of percutaneous embolization in a cirrhotic patient. Sekido et al. also described liver transplantation in which graft hypofunction was determined by large mesenteric varices; the shunt was, however, ligated by conventional surgery.

We report a case where the portosystemic shunt occurred between the superior mesenteric vein and the left ovarian vein. To the best of our knowledge, there is only one previous report of percutaneous embolization of portosystemic shunts after liver transplantation.

In summary, portosystemic encephalopathy due to portosystemic shunt may occur many years after liver transplantation, even in the absence of portal hypertension and liver fibrosis. This can be due to old shunts not eliminated during the transplant surgery. The trigger of this complication so long after the liver transplantation remains controversial. Percutaneous embolization treatment can achieve good results and should be considered in these patients.

References