Splenectomy for Severe Intestinal Bleeding Caused by Portal Hypertensive Enteropathy After Pediatric Living-Donor Liver Transplantation: A Report of Three Cases

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ABSTRACT

Background. The incidence of portal vein thrombosis after pediatric living-donor liver transplantation (LDLT) is reported to be higher than that after deceased-donor or adult liver transplantation. Portal vein thrombosis can cause portal hypertension and related complications, including portal hypertensive gastropathy or portal hypertensive enteropathy (PHE). PHE, in particular, can lead to severe intestinal bleeding, which is extremely difficult to treat. However, the pathogenesis of and appropriate treatment for PHE are not clearly defined, especially after pediatric LDLT.

Methods. Herein, we report three cases of refractory intestinal bleeding caused by PHE after pediatric LDLT, which were treated with splenectomy.

Results. The time between LDLT and splenectomy was 43, 92, and 161 months, respectively. All 3 patients were discharged from the hospital without any peri-operative complications and were doing well, with no adverse effects at 174, 81, and 12 months after splenectomy, respectively. Although shunt surgeries, including the use of a meso-Rex shunt, are reported to be a useful option when the portal vein is completely occluded, adhesiotomy around the liver graft would be required, which could damage the hepatopetal collateral vessels that maintain portal vein flow to the graft. Therefore, shunt surgeries, which can lead to re-transplantation, are considered to be highly risky as a first-line treatment option, particularly considering the limited accessibility to deceased donor organs in our country.

Conclusions. Our data demonstrate that simple splenectomy, although considered a palliative treatment, can be a safe and effective method to control severe intestinal bleeding caused by PHE after pediatric LDLT.

PORTAL vein thrombosis (PVT) is a serious complication after liver transplantation. PVT after pediatric living donor liver transplantation (LDLT) is not rare, with a reported incidence of approximately 8% to 14% [1–3], which is higher than that after deceased donor or adult liver transplantation because of shorter portal veins and/or pre-transplant cholangitis. Although the hepatopetal collateral veins mostly preserve intra-hepatic portal flow and the function of the liver graft, PVT can cause portal hypertension and related complications, including portal hypertensive gastroenteropathy (PHGE). Portal hypertensive enteropathy (PHE), in particular, can lead to severe intestinal bleeding, which is extremely difficult to treat. Although severe intestinal bleeding caused by PVT-induced PHE can develop over the long term after LDLT, the pathogenesis and appropriate treatment are not clearly defined [4], especially after pediatric LDLT. In general, such intestinal bleeding is initially treated with portal pressure-reducing drugs, mainly nonselective β-blockers [5,6], endoscopic treatment [7,8], and/or a transjugular intra-hepatic porto-systemic shunt (TIPS) [9–11]. Nevertheless, intestinal bleeding caused by PVT-induced PHE is often persistent

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and difficult to control with drugs or TIPS alone. Interventional radiologic approaches and surgical small-bowel resection can be effective. However, in some cases, the point of bleeding cannot be detected through endoscopic or angiographic approaches. In such situations, it is extremely difficult to treat the intestinal bleeding and subsequent anemia, especially after LDLT. A meso-Rex shunt is reported to be a useful option when the portal vein is completely occluded [12,13]. However, adhesiotomy around the liver graft would be required, which could damage the hepatopetal collateral vessels that maintain portal vein flow to the graft. A distal spleno-renal shunt, which has been reported to be effective for PVT-induced variceal bleeding [14,15], is difficult to apply in transplant recipients because such patients undergo polysurgery, including the Kasai procedure and LDLT, and are susceptible to complicated procedures, such as a distal spleno-renal shunt.

In 2005, we reported the first case of severe intestinal bleeding caused by PVT-induced PHE treated with simple splenectomy (case 1). However, the follow-up period was insufficient to evaluate the efficacy of the procedure [16]. Here, we describe 3 cases, including the previously reported case, with much longer follow-up periods, who underwent splenectomy for treatment of persistent intestinal bleeding caused by PVT-induced PHE after pediatric LDLT.

Case 1

In 1998, a 10-month-old Japanese girl underwent LDLT after a failed Kasai procedure for biliary atresia [16]. The transplanted graft functioned well, but the hepatic artery was re-anastomosed because of hepatic arterial thrombosis on post-operative days (PODs) 4 and 11. Ten months after LDLT, the patient had tarry stools that gradually worsened and became more frequent. However, neither endoscopic nor angiographic examinations detected any obvious points of bleeding. Portography via the superior mesenteric artery revealed extra-hepatic portal vein occlusion with cavernous transformation. The patient required repeated blood transfusions. Therefore, in 2002, when the patient was 4 years old, simple splenectomy was performed to reduce blood flow into the portal venous system and to ameliorate thrombocytopenia. Intra-operative monitoring revealed a decrease in portal vein pressure from 21 to 14 mm Hg after splenectomy. The patient had an uneventful recovery and was discharged [16]. Thereafter, the patient was re-admitted 9 years after splenectomy because of tarry stools but was subsequently discharged without additional surgical treatment. Fourteen years after splenectomy, the patient was doing well, showed normal growth, and had no serious infection.

Case 2

In 2002, a 1-year-old girl underwent ABO-incompatible LDLT after a failed Kasai procedure for biliary atresia. LDLT was performed with end-to-end anastomosis of the hepatic vein, portal vein, and hepatic artery, and end-to-side Roux-en-Y hepaticojejunostomy, without complications. Tacrolimus, methylprednisolone, and azathioprine were administered for immunosuppression. On POD 3, titers of anti-ABO antibody increased sharply and liver function test results were elevated despite post-operative plasma exchange. Needle graft biopsy revealed antibody-mediated rejection, and treatment with rituximab, deoxyspergualin, plasma exchange, and steroid pulse therapy was initiated. Antibody titers and liver function test results markedly improved, and the patient was discharged on POD 74.

Three years after LDLT, follow-up ultrasonography detected feeble portal blood flow, and portography and computed tomography revealed extra-hepatic PVT along with the collateral vessels (Fig 1). Five years after LDLT, the patient had tarry stools. Seven years after LDLT, the patient was repeatedly hospitalized for tarry stools and consequent severe anemia. In 2010, when the patient was 9 years old, esophagogastroduodenoscopy and colonoscopy were performed under general anesthesia. However, no obvious points of bleeding were detected despite refractory tarry stools and severe anemia; her hemoglobin level was 5.7 g/dL. After vaccination against *Streptococcus pneumoniae*, simple splenectomy was performed to reduce blood flow into the mesenteric venous system and to ameliorate thrombocytopenia. Intra-operative monitoring revealed a decrease in portal vein pressure from 22 to 18 mm Hg after

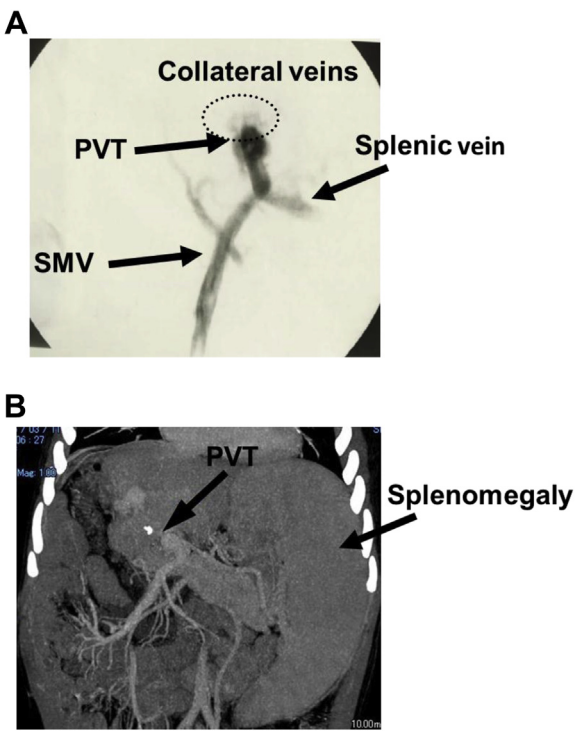

Fig 1. Portographic (**A**) and abdominal computed tomographic (**B**) images of case 2, taken 3 years after liver transplantation. Abbreviations: PVT, portal vein thrombosis; SMV, superior mesenteric vein.