

Distal Splenorenal Shunt for Portal Vein Thrombosis after Liver Transplantation

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A 17-yr-old female received a liver transplant for type I glycogen storage disease. A year later, when she experienced variceal gastrointestinal hemorrhage, an angiogram revealed thrombosis of the portal vein with hepatopetal collateral channels. A distal splenorenal shunt was performed because of failure of sclerotherapy to control subsequent bleeding episodes and the fact that the liver function was normal. This patient continues to have normal hepatic function with a patent splenorenal shunt 4 yr after the shunting procedure. This case illustrates the feasibility of a distal splenorenal shunt to alleviate portal hypertension in cases of thrombosis of the portal vein following hepatic transplantation if the liver function is normal.

INTRODUCTION

Survival rates and rehabilitation after orthotopic liver transplantation (OLT) have greatly improved during the so-called cyclosporine era (1). Nevertheless, technical problems are still significant (2) and reoperation is often required. Thrombosis of the hepatic artery and problems associated with biliary tract reconstruction represent the most important technical complications, with an incidence of 7% and 13.2%, respectively (3, 4).

Thrombosis of the portal vein after OLT (reported incidence, 1.8%) has been observed almost exclusively in children (5). The diagnosis usually results from the development of clinical signs of portal hypertension, such as variceal hemorrhage, splenomegaly, and hypersplenism. The treatment is aimed at relieving the presenting symptoms or correcting the underlying cause. In our experience, the therapy for posttransplantation portal vein thrombosis depends on the clinical presentation, residual portal flow, and liver function. Consequently, the therapeutic choices are sclerotherapy or retransplantation.

We report herein a very unusual case of late portal thrombosis following OLT in a child. As a sequel to

this, the patient developed portal-portal hepatopetal collaterals sufficient to maintain good liver perfusion but unable to decompress the portal hypertension. She was successfully treated with a splenorenal shunt.

CASE REPORT

A 17-yr-old white female with end-stage liver disease secondary to type 1 glycogen storage disease was referred in January 1982 to our institution for OLT evaluation. Nine years before the referral, she underwent end-to-side portacaval shunt to improve her growth and the metabolic abnormalities associated with glycogen storage disease (6). This treatment was effective in improving some aspects of her disease, such as lipid and uric acid metabolism, acidosis, and growth retardation. However, surgery did not relieve the frequency of hypoglycemia, and appeared to have worsened the clinical effect of the hypoglycemic episodes (7). Frequent daytime feeds and nighttime nasogastric feedings were necessary after the portacaval shunt to avoid symptomatic hypoglycemia.

During this period, the child was followed with serial ultrasound examinations of the liver which began to reveal intrahepatic masses. A biopsy of one of these lesions proved to be an adenoma.

In September 1981, the patient experienced the onset of severe abdominal pain and liver swelling which was thought to be related to a hemorrhage into the largest of the adenomas which led to biliary obstruction and hepatic failure. The child had an alkaline phosphatase of 3000 IU/L, ammonia of 315 μ g/L, and increased liver enzyme levels. Two months later, her condition deteriorated further, and she became jaundiced. The patient improved on total parenteral nutrition alone, and she returned to her base-line state of health. The decision was made to make the patient a liver transplant candidate, based on the fact that she had hepatic adenomas with the potential risk of developing carcinomatous changes or liver failure or both.

Liver transplantation was performed using the technique previously described (8, 9). Immunosuppression

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Received Mar. 1, 1988; accepted July 6, 1988.

consisted of cyclosporine and prednisone (1). Her initial postoperative course was complicated by a bile leak which was treated with drainage and sealed spontaneously. Nevertheless, liver function remained excellent and her subsequent clinical course was uneventful.

One year after OLT, the patient was evaluated for gastrointestinal tract bleeding. Endoscopy showed esophageal and gastric varices. Angiography demonstrated portal vein thrombosis and hepatopetal collateral formation (Fig. 1). Liver enzyme levels were normal. Treatment consisted of blood transfusions and endoscopic sclerosis of the varices. Despite this, she had several episodes of significant upper gastrointestinal tract bleeding over the next 3 months. Liver biopsy was normal, as were liver enzyme levels.

Because of repeated episodes of gastrointestinal tract bleeding and the subsequent need for multiple transfusions, a distal splenorenal shunt for control of portal hypertension was performed in June 1983. The anastomosis measured approximately 13 mm in diameter. The splenorenal shunt decompressed the varices and resolved the gastrointestinal tract bleeding.

Follow-up angiography performed in May 1984 (Fig. 2), ultrasound performed in December 1986, and, more recently, a nuclear magnetic resonance test demonstrated a widely patent splenorenal shunt. Over the past 4 yr, the patient has not had any significant gastrointestinal tract bleeding. The most recent liver enzyme levels, performed in March 1987, were normal.



FIG. 1. Hepatopetal venous collaterals after portal vein thrombosis following liver transplantation. Occlusion of the extrahepatic portal vein (*long straight arrow*) at the junction of the splenic and superior mesenteric veins demonstrated on subtraction venous phase film of selective superior mesenteric arteriogram. There is reconstitution of the intrahepatic portal vessels (*short straight arrow*) via multiple tortuous and dilated collateral vessels (*curved arrows*) to the right of the superior mesenteric vein and in the hilar region. There is portal hypertension as evidenced by retrograde flow in the splenic vein and marked gastroesophageal varices.



FIG. 2. A widely patent splenorenal shunt (*arrow*) is demonstrated on splenic venogram performed 1 yr after surgery. This study was performed by passing a catheter retrogradely through the shunt into the splenic vein via the left renal vein. (Courtesy of Philip Stanley, M.D., Children's Hospital of Los Angeles, CA).

DISCUSSION

Thrombosis of the portal vein after OLT has been reported by Lerut *et al.* (5) in seven of 393 hepatic grafts. The contributing factors in five of the seven patients were size mismatch, recipient portal vein thrombosis, and recipient portal vein hypoplasia. In the patient reported herein, the pre-existing end-to-side portacaval shunt may have predisposed the portal vein to thrombose.

Thrombosis of the portal anastomosis is usually followed by the development of collateral circulation, as first described by Hahn *et al.* (10). Such collaterals develop secondary to the high pressure in the extrahepatic portal venous system and, essentially, are of two types: portal-portal or hepatopetal, and portal-systemic or hepatofugal (11). Although some authors do not believe that hepatopetal collaterals that develop after portal vein thrombosis could be of any importance in maintaining good hepatic perfusion (12, 13), others have demonstrated that such collateral circulation is

important in reestablishing portal flow to the liver (11, 14).

In nontransplant patients with extrahepatic portal vein obstruction, the normal sinusoidal pressure is considered to be fundamental in favoring a periportal collateral pathway around the obstructed vein to the liver parenchyma (15). The development of a hepatopetal collateral circulation results from the presence of many small collateral veins around the extrahepatic portal vein and within the hepatoduodenal and hepaticocolic ligaments. These small vessels enlarge because of a high pressure gradient between the proximal thrombosed extrahepatic portal vein, and the intrahepatic portal vein and sinusoidal system.

The development of hepatopetal collaterals after portal vein thrombosis is clearly understandable in a native liver, but is more difficult to explain in a liver allograft. Thus, because all potential portal collaterals are divided during harvesting of the liver, the only connection between the splanchnic circulation and the allograft liver is via the portal anastomosis. Apparently, collaterals develop between the recipient extrahepatic portal segment and the allograft liver hilum, thus becoming the anatomical pathway for a possible hepatopetal collateral circulation. In our case, the blood flow through these new collaterals was sufficient to perfuse the allograft liver with the necessary hepatotrophic portal factors for a normal hepatic physiology, but not sufficient to maintain normal portal pressure. Consequently, portal hypertension developed and led to esophageal varices and severe gastrointestinal tract bleeding. In this setting, the therapeutic choices are sclerotherapy, retransplantation, or portal diversion surgery.

Sclerotherapy is a good method to use to control acute variceal bleeding, but it is not always effective in permanent control of gastroesophageal bleeding and in the treatment of gastric varices. In the latter condition, the persistent hypertension in the gastric veins represents a life-threatening situation, since bleeding from erosive gastritis is a leading cause of death in this condition (16). As demonstrated in our patient, repeated sclerotherapy was ineffective in achieving definitive control of bleeding.

Retransplantation is a therapeutic option practiced routinely at our institution, rescuing those patients whose primary grafts are failing for primary nonfunction, intractable rejection, or technical complications (17). In our case, retransplantation was not performed because graft function remained excellent and its histologic architecture was normal.

The final therapeutic option, portasystemic shunting, would hopefully eliminate the recurrent bleeding episodes. The use of a selective shunting procedure might allow control of bleeding while preserving the necessary high pressure in the intestinal venous system. To main-

tain collateral portal perfusion with hepatotrophic portal factors (16, 18), the distal splenorenal shunt was physiologically the most ideal shunt for the purpose just outlined. Further, the alternative mesocaval shunt was not selected because potential complications with this particular portosystemic diversion could lead to thrombosis of the superior mesenteric vein and subsequent loss of the hepatic allograft. In the distal splenorenal shunt, the surgical dissection is away from the hepatic hilum, which minimizes the risk of jeopardizing a future second transplant if one is needed (19). The long-term patency of the shunt without any recurrence of bleeding, and the excellent quality of liver function 5 yr posttransplantation and 4 years after portal shunting, clearly demonstrate the reliability of this therapeutic choice.

ACKNOWLEDGMENTS

Supported by research grants from the Veterans Administration and by Project Grant AM 29961 from the National Institutes of Health, Bethesda, Maryland.

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