Auxiliary Partial Orthotopic Living Donor Liver Transplantation in a Patient With Wilson’s Disease: A Case Report

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ABSTRACT

A patient with end-stage liver cirrhosis and neurological disorder due to Wilson’s disease (WD) underwent auxiliary partial orthotopic liver transplantation (APOLT) using a living donor. He first visited our institute complaining of hand tremor, which was diagnosed as WD. Despite medical therapy, hepatic impairment progressed toward portal hypertensive complications. He was considered a suitable candidate for living donor-related liver transplantation. However, because of the impossibility of mobilization of the lateral section due to severe splenomegaly at the time of the recipient operation, we performed an APOLT using a right lobe graft. After transplantation, he suffered hepatic vein stenosis and biliary stenosis, receiving interventional therapy. The remnant native liver volume decreased, and the volume of the graft increased serially after transplantation. At the time of reporting, the patient had a normal working life with normal serum ceruloplasmin level and without neurologic problems at 26 months posttransplantation. APOLT may be a therapeutic option for patients with WD.

CASE REPORT

The patient was a 36-year-old man whose medical history indicated no WD-related complications until the age of 22 years when he developed an intention tremor of right hand. WD was diagnosed with a low serum ceruloplasmin level of 2 mg/dL. Since then, he was followed at the department of neurology with chelation treatment. At age 35, he suffered gastrointestinal bleeding due to advanced esophageal varices and ascites. Therefore, he was considered to be a suitable candidate for liver transplantation and was transferred to our department. His preoperative liver condition was grade B on the Child-Turcotte-Pugh (CTP) classification (score 7) and his Model for End-Stage Liver Disease (MELD) score was 17. There was no evidence of malignancy, of another space-occupying lesion mimicking a malignant tumor, or of a focal thrombus in the portal vein.

The donor was the 31-year-old brother of the patient whose liver function tests were normal. The graft-to-recipient weight ratio (GRWR) was 0.93% using a right lobe graft. Genetic testing of the donor showed no mutations within the ATP7B gene.

At the time of the recipient operation, mobilization of the lateral section was impossible due to severe splenomegaly. Because of this problem, we decided to perform APOLT using the donor’s right lobe.

At first, right trisectionectomy was performed, and then the donor right lobe was implanted in the right side of the recipient. Middle hepatic vein (MHV) reconstruction of the right lobe graft was performed at the bench using the cryopreserved iliac vein. The right hepatic vein of the graft was anastomosed in end-to-end fashion to the recipient right hepatic vein, followed by anastomosis of the reconstructed MHV of the graft to the recipient MHV. The portal vein, hepatic artery, and bile duct of the graft were anastomosed to the corresponding recipient structures in end-to-end fashion. The immunosuppressive protocol consisted of basiliximab for induction therapy, and cyclosporine-steroid for maintenance therapy. MHV stenosis and
biliary strictures observed on postoperative days (POD) 7 and 15, respectively, were treated by vascular intervention and percutaneous transhepatic biliary drainage methods. Serum ceruloplasmin level increased to the normal range on POD 12 (15.1 mg/dL). The patient was discharged with normal liver function after 30 days. Computed tomography (CT) performed at 18 months after transplantation showed a well-regenerated graft (153% of the original volume) and an atrophied native liver (21% of the original volume) without tumor development (Fig 1). Twenty-six months after transplantation, the recipient had a normal working life with a normal serum ceruloplasmin level.

DISCUSSION

Wilson’s disease associated with severe liver involvement is effectively cured by OLT. Even progressive neurological deterioration with no hepatic insufficiency is considered a suitable indication for OLT. We successfully performed APOLT in a patient with end-stage liver cirrhosis and neurologic disorder due to WD, observing normal ceruloplasmin level and liver functions as well as cure of neurologic symptoms. Therefore, we suggest that APOLT may also be considered for WD patients with combined hepatic and neurological involvement. However, OLT in the case of severe neurological impairment is still open to debate.

One shortcoming of APOLT for cirrhotic liver disease is the potential risk for carcinogenicity of the remnant native liver. This problem remains to be resolved. However, a report from the Kyoto group with more than 5 years follow-up showed no tumor development in the remnant native liver. Therefore, delayed native hepatectomy after complete graft regeneration might not be necessary.

Because we performed a right trisectionectomy of the recipient liver, it was easy to isolate the MHV and perform an anastomosis between the reconstructed MHV of the graft to the recipient MHV. The right portal vein and hepatic duct were isolated sufficiently for a successful end-to-end anastomosis to the corresponding graft structures. A portal flow diversion technique might not be mandatory in cases of cirrhosis. We performed APOLT without this technique, because of the presumption that the grafted liver would be supplied by the most portal flow in the struggle between the graft and the native liver. We were able to prove this presumption correct, because of the markedly regenerated graft and atrophy of the remnant native liver, in agreement with other authors.

REFERENCES