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BRIEF REPORT

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Living related liver transplantation for polycystic liver disease

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Abstract Polycystic liver disease (PCLD) is a rare inherited disorder, often associated with polycystic disease of the kidney. Although liver failure is unusual, some patients suffer from hepatic enlargement associated with severe complications such as abdominal distention, cachexia and dyspnea. Until recently, many surgical attempts had been made to reduce hepatic size, however, results have been unsatisfactory [3, 9, 10]. Today, liver transplantation is recommended as a therapeutic option, and excellent outcome has been demonstrated [1, 2, 4, 5, 6, 8, 11]. In this paper, we present the first case study of total hepatectomy and partial liver transplantation for PCLD, from a living, related donor. The patient is a 38-year-old man with PCLD who underwent living related liver transplantation (LRLT). He is alive and well 21 months after the operation, with complete resolution of the symptoms. He has returned to his previous job, with a marked improvement in his quality of life. Our experience demonstrates that LRLT can be an option for treatment of PCLD.

Keywords Liver transplantation · Polycystic liver disease

Introduction

A 38-year-old male patient was admitted to Tokyo Women's Medical University Hospital in December 1999 for living related liver transplantation (LRLT). He had first presented 6 years previously with abdominal distention and had been diagnosed as having polycystic liver disease. Since then, while under close medical observation, he complained of increasing abdominal distention, anorexia and dyspnea. His father, who had also been diagnosed with polycystic disease, had died of renal failure. On examination, our patient was found to have diffuse bi-lobar multiple cysts in the liver, and also in the kidney (Fig. 1). Hepatomegaly increased progressively until he had gained 15 kg in 5 years. Resective and fenestration procedures were not undertaken because of the significantly associated morbidity and mortality, as well as a lack of consistent long-term palliation. His symptoms kept him from pursuing his daily full-time employment and limited his routine activities of daily living, although liver function was normal. Hematological study showed malnutrition, with a serum prealbumin level of 13.5 mg/dl and retinal-binding protein level of 2.8 mg/dl. Serum creatinine was 1.0 mg/dl, and creatinine clearance was 40.9 ml/min.

Case report

He underwent LRLT from his 62-year-old mother in December 1999. On entering the peritoneal cavity we noted a huge polycystic liver and 2,000 ml of ascitic fluid. Before the hepatectomy, we

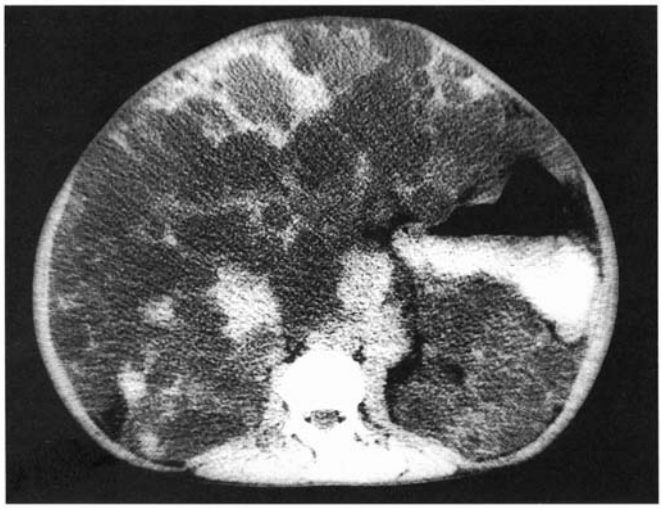

Fig. 1 Computed tomographic scan in our patient showing diffuse PCLD without areas of significant parenchymal sparing. Note the presence of polycystic disease in the bilateral kidney

performed extensive cyst punctures and decompression to facilitate dissection. The weight of the excised liver was 20,000 g, including 5,000 ml of aspirated intracystic fluid. We transplanted the left lobe, which weighed 350 g, employing the standard operating technique in an orthotopic position [7]. The intraoperative transfusion requirement was 26 units of packed red blood cells. For postoperative maintenance immunosuppression, tacrolimus and prednisolone were administered. The patient, who had an uneventful course after the operation, experienced complete relief from his previous symptoms and is now enjoying excellent quality of life since his discharge. Pulmonary function tests showed improvements as follows: increase in vital capacity from 65% to 88%, and forced expiratory volume from 86% to 94%. The blood urea nitrogen level was 23 mg/dl and the serum creatinine 1.4 mg/dl at the time of discharge. To date he has survived for 18 months after the operation, with normal liver function.

Discussion

Polycystic liver disease (PCLD) is a rare disorder of the liver parenchyma, often associated with polycystic disease of the kidney. It is characterized by multiple, diffuse, cystic lesions within the liver, which may cause debilitating symptoms, occasionally requiring surgical treatment. The treatment of PCLD is controversial, and the optimal surgical approach still needs to be clearly defined. Several methods of treatment have been described for PCLD, such as aspiration and sclerosis, and fenestration with and without hepatic resection [3, 9, 10]. Only patients with one dominant, large cyst may be suitable candidates for these procedures, since they are associated with high complication and recurrence rates. Starzl et al. [5] were the first to report on the series of liver transplantations for PCLD. They indicated that lethal exhaustion should be a major indication for this procedure. Washburn et al. [11] suggested the following: patients with massive hepatomegaly and bi-lobar, predominantly small multicystic disease, without areas of parenchymal sparing, should be offered liver transplantation as a primary treatment. Some other recent reports have also recommended liver transplantation as a therapeutic modality and have demonstrated an excellent outcome of the procedure in those patients [1, 2, 4, 6, 7, 8].

In our case, the patient had been working regularly until 2 years previously, but had since become housebound and unable to perform the activities of daily living. Although liver transplantation is a radical procedure, especially from a living donor, we performed the transplantation after obtaining fully informed consent. Since most PCLD patients have preserved hepatocellular function, other fatal diseases should be given priority for liver transplantation, considering the shortage of donor organs. Living transplantation is now legal, and we may take living transplantation into consideration for this disease. After the operation, the patient was relieved from the symptoms and returned to his previous job, with a marked improvement in his quality of life. The donor, his mother, is also quite well, with normal liver function after the operation. Transplantation of the left lobe, which was smaller in size (graft volume, standard liver volume ratio: 29.6%), was sufficient in this case, because preoperative liver function was normal, and no infections were found.

Most patients with PCLD have preserved liver function until the very end and are difficult to keep on a waiting list for cadaveric transplantation. Yet these patients can be severely disabled, due to symptoms such as cachexia, malnutrition, and cyst infection. LRLT can be a suitable treatment for PCLD.

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