

A successful case of combined liver and kidney transplantation for autosomal dominant polycystic liver and kidney disease *

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With advances in transplantation, multiorgan transplantation has become a treatment of choice for end-stage organ failure which can not be reversed with other modalities. In 1984, the first case of combined liver and kidney transplantation was introduced by Witts at Innsbruck University Hospital in a 30-year-old man with HBsAg positive cirrhosis. He survived more than 9 years^[1]. Since then, an increasing number of such combined transplantation has been performed. But in Asia, this technique has not been put into clinical practice yet. We report such a case below.

CASE REPORT

Physical examination

A 52-year-old woman was admitted to our hospital with fatigue, jaundice and severe abdominal pain. She suffered from congenital polycystic liver and kidney disease at 40 years of age. Two weeks before her admission, she underwent laparotomy in a local hospital for unbearable abdominal pain. She was found to have anemia and jaundice in physical examination. She had an enormous liver extending to pelvic cavity with an enlarged spleen. Biochemical tests showed hemoglobin 103 g/L, WBC 3.2×10^9 /L, platelet 7.4×10^9 /L, serum total bilirubin 42.7 $\mu\text{mol/L}$, albumin 30 g/L, creatine 62 $\mu\text{mol/L}$, BUN-8.5 $\mu\text{mol/L}$ and prothrombin time 18 seconds (control 12 seconds). The renogram indicated that her renal function had been slightly damaged. Two weeks after her admission, she re-

ceived combined liver and kidney transplantation.

Procurement of donor organs

The donated liver and kidney were harvested using the rapid multiple organs harvesting technique^[2]. Both organs were flushed *in situ* and then cold preserved in the Winsconson University solution. Lymphocyte cross-matching was negative and the panel reactive antibody (PRA) was 8%.

Operative and postoperative course

The liver was transplanted orthotopically using venovenous bypass during anhepatic phase. Following implantation of the liver, the kidney was placed intraperitoneally into the right iliac fossa after 12.5h of cold ischemia. Bile was produced promptly, indicating immediate graft function. The kidney assumed normal color and consistency after revascularization and produced copious amounts of urine. The total operative blood loss was 3500mL, and 2800mL blood products were infused. Immunosuppressive regimen including cyclosporine A, 2mg/kg per day intravenously, and methylpredisolone (starting with 1 g per day and tapering to 10 mg daily) was given.

Results

The early postoperative course was uneventful. The patient was discharged two months after combined liver and kidney transplantation. Six months after transplantation, however, she developed jaundice though her general condition was good. Following the jaundice period, liver function began to show signs of deterioration. The biliary sludge was diagnosed. She was reoperated due to biliary obstruction. The biliary sludge which was 2.0 cm \times 0.5 cm in size located in the middle portion of common bile duct. The patient was soon recovered from the second operation and discharged on the 20th day with nearly normal liverfunction. She is doing well and is in good health.

DISCUSSION

The high success rate achieved with single organ transplantation has stimulated the assumption of double organ transplantation for patients with complex multiorgan failure. The development of new immunosuppressive agents and improved surgical ap-

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proaches as well as sophisticated postoperative management make assumption become clinical practice.

The main concern in combined liver and kidney transplantation is the selection of candidates. According to Margreiter^[3], the indications for combined liver-kidney transplantation are divided into three categories: ① Disease affecting both organs, such as autosomal dominant polycystic disease. ② Renal disorders with liver involvement or liver disorders with renal involvement, including primary type I hyperoxaluria, type I glycogen storage disease, cholesterol acyltransferase deficiency, etc. ③ Separate diseases of both organs. This category consisting of patients on hemodialysis with liver disease and patients with end-stage liver disease and renal function impairment other than hepatorenal syndrome is the main indication for combined liver and kidney transplantation.

Autosomal dominant polycystic kidney disease is often present in newborns, but does not become clinically evident until adulthood. It is often associated with liver cysts which may reach enormous size (in our case, it weighs 5.0kg) and can therefore be extremely disabling considering both the rigors of renal ischemia during transplantation and nephrotoxic effects of cyclosporine A, and some necessitating antibiotics may worsen the previously impaired renal function and lead to postoperative renal failure. We treated the patient with combined liver-kidney transplantation.

The surgical technique of combined liver-kidney transplantation was exactly the same as for transplantation of the liver or the kidney alone. For the reason that liver is more subject to cold ischemia damage than kidney, the liver is generally implanted prior to the kidney. In case a venovenous bypass

should be needed, the axilla and thigh, preferentially on the left side, must be prepared. Because of the poor coagulation status of the recipient, heparin may not be required for prevention of clotting.

A protective role of the liver allografts in the survival of other solid organ transplants has been noted, but the exact mechanism of this phenomenon has not been elucidated. The liver has the potential to protect the other organs immunologically. The efficacy of simultaneous liver-kidney transplantation in the prevention of hyperacute renal rejection in patients with reformed anti-HLA lymphocytotoxic antibodies is also demonstrated in clinical practice. However, these observations have been challenged by reports documenting hyperacute rejection of kidney or/and liver allografts when the roles of ABO matching and donor/recipient crossmatching have been violated^[4-5]. Although our patient did not experience any rejection and showed some protection of the transplanted kidney by the liver graft, we believe that careful selection of suitable donor with HLA typing crossmatching and PRA test are essential to success in combined liver-kidney transplantation.

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