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거대간낭종을 동반한 상염색체 우성 다낭신 환자에서 시행된 간 이식

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Liver Transplantation for Massive Polycystic Livers in Autosomal Dominant Polycystic Kidney Disease Patients

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Background: Polycystic liver is the most common extrarenal manifestation in autosomal dominant polycystic kidney disease (ADPKD). Patients with polycystic liver often suffer from abdominal discomfort, ascites, malnutrition or dyspnea. Liver transplantation (LT) might be the only curative treatment option for symptomatic polycystic liver. We investigated the outcomes of LT in ADPKD patients.

Methods: We retrospectively analyzed the medical records of ADPKD patients who underwent LT between 1988 and 2012 at Seoul National University Hospital.

Results: Five patients underwent LT. All were women, with a mean age of 51±10 years. Before LT, all patients had received transcatheter arterial embolization or partial hepatectomy for large hepatic cysts. The primary indications for LT were hepatomegaly-related symptoms and previous treatment-related hepatic failure. Two patients received transplants from living donors and 3, from deceased donors. The mean expected blood loss was 7.4±7.3 L, and mean duration of postoperative hospital stay was 27±16 days. The most common postoperative complications were ascites and pleural effusion. Although subclinical acute cellular rejection was found in 2 patients, all patients had good liver function at follow-up (median, 8 months; range, 4-45 months). Two patients were under hemodialysis when they underwent LT. Perioperative acute kidney injury occurred in 1 patient that underwent deceased donor LT. However, renal function showed no significant decline 3 months after LT. Interestingly, the total kidney volume did not increase after LT in any case.

Conclusion: Both living and deceased donor LT are effective and tolerable treatment options for symptomatic polycystic liver in ADPKD patients

Key Words: 상염색체우성다낭산, 간이식, 다낭간
ADPKD, Liver transplantation, Polycystic liver disease