

거대 간낭종을 동반한 상염색체 우성 다낭신 환자에서 거대 간낭종을 동반한 상염색체 우성 다낭신 환자에서 시행된 간 이식과 부분간절제술, 간동맥색전술의 비교연구

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Outcomes of Liver Transplantation, Partial Hepatectomy, and Transcatheter Arterial Embolization for Massive Polycystic Liver in Autosomal Dominant Polycystic Kidney Disease Patients

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Background: Polycystic liver disease (PLD) is the most common extra-renal manifestation in autosomal dominant polycystic kidney disease (ADPKD). PLD patients suffer from hepatomegaly leading to pain, dysphagia, and caval obstruction. Interventions for symptomatic PLD include transcatheter arterial embolization (TAE), partial hepatic resection and liver transplantation (LT). We compared outcomes of each treatment modality.

Methods: We retrospectively analyzed 29 patients who had undergone TAE, partial hepatectomy, or LT in ADPKD patients in a single center.

Results: Six and seven patients underwent LT and partial hepatectomy, respectively, while sixteen patients underwent TAE. Living donor LT was performed for intractable hepatomegaly-related symptoms despite TAE in 2 patients, and for uncontrolled ascites in a patient. Deceased donor (DD) LT was performed in 2 patients for hepatic failure or uncontrolled ascites after TAE. Another patient underwent DDLT for hepatic failure after partial hepatectomy. The most common complications were ascites and pleural effusion. Two patients were under HD at LT, and acute kidney injury resulted in kidney failure in 1 DDLT patient. The estimated blood loss and transfusion amount were larger in LT group. All patients had maintained good liver function at a median follow-up of 15.5 months, although subclinical acute cellular rejection was found in 2 patients. Causes of partial hepatectomy were recurrent cyst infection and hepatomegaly-related symptoms. One patient underwent partial hepatectomy, because multiple TAE had failed to control symptoms. Preoperative CTP score was lower in the partial hepatectomy group than in the LT group. A few serious infectious complications occurred, and antibiotic treatment cured them without mortality. Eleven patients received single TAE, and five patients underwent TAE twice. Five patients (31.3%) in the TAE group needed LT or partial hepatectomy to relieve symptoms or rescue hepatic failure. Two patients died of obstructive jaundice and cyst infection during follow up. Except one LT patient resulted in kidney failure, there were no evidence of worsening in kidney function during 1year follow up in all groups.

Conclusion: Liver transplantation is more effective and tolerable treatment option for symptomatic PLD in ADPKD patients compared to partial hepatectomy and TAE especially patients with decreased liver function.

Key Words: 다낭성신질환, 간이식

Autosomal polycystic kidney disease, Liver transplantation