Abstract Type : Oral
Abstract Submission No. : 1682

Factors associated with the development and severity of polycystic liver in patients with autosomal-dominant polycystic kidney disease

Yaerim Kim[1], Hayne Cho Park[2], Yong-Chul Kim[3], Curie Ahn[4], Kyu-Beck Lee[5], Yeong Hoon Kim[6], Seungyeup Han[1], Eun Hui Bae[7], Kook-Hwan Oh[3], Yun Kyu Oh[8]
[1]Department of Internal Medicine-Nephrology, Keimyung University School of Medicine, Korea, Republic of
[2]Department of Internal Medicine-Nephrology, Hallym University Sacred Heart Hospital, Korea, Republic of
[3]Department of Internal Medicine-Nephrology, Seoul National University Hospital, Korea, Republic of
[4]Department of Internal Medicine-Nephrology, National Medical Center, Korea, Republic of
[5]Department of Internal Medicine-Nephrology, Kangbuk Samsung Hospital, Korea, Republic of
[6]Department of Internal Medicine-Nephrology, Inje University Busan Paik Hospital, Korea, Republic of
[7]Department of Internal Medicine-Nephrology, Chonnam National University Hospital, Korea, Republic of
[8]Department of Internal Medicine-Nephrology, SMG-SNU Boramae Medical Center, Korea, Republic of

Objectives: Factors related to the development and severity of polycystic liver disease (PLD) have not been well established. We aimed to evaluate the genetic and epidemiologic risk factors of PLD in patients with autosomal dominant polycystic kidney disease (ADPKD).

Methods: Adult patients with inherited cystic kidney disease were enrolled from May 2019 to May 2021. Demographic, clinical, and laboratory data were collected at the initial study visit. The severity of PLD was graded based on the height-adjusted total liver volume (HtTLV): <1000 mL/m (Gr1), 1000-1800 mL/m (Gr2), and >1800 mL/m (Gr3). Targeted exome sequencing was done by a gene panel including 89 ciliopathy-related genes. We searched out the relative factors to the presence and the severity of PLD using logistic regression analysis.

Results: Of 602 patients with typical ADPKD, 461 (76.6%) patients had liver cysts. The patients with liver cysts showed female predominance and a higher frequency of other ADPKD-related complications. An older age, hypertension, and genetic variants with truncating mutation of PKD1 (PKD1-PT) or PKD2 commonly affected the development and severity of PLD. Higher kidney volume with Mayo classification 1C-1E was significantly associated with the development of liver cysts, but not with the severity of PLD. On the other hand, higher body mass index, lower hemoglobin, and higher alkaline phosphokinase (ALP) were the significant risk factors of severe PLD (≥Gr 2).

Conclusions: Hepatic involvement in ADPKD could be related to kidney manifestations and genetic variants including PKD1-PT or PKD2. Monitoring hemoglobin and ALP and evaluating the genetic variants might help predict severe PLD.