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한국 성인 환자에서 초점성 분절성 사구체 경화증의 병리학적 분류에 따른 임상적 특징과 예후

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Clinical Features and Outcomes of Focal Segmental Glomerulosclerosis (FSGS) Pathologic Variants in Korean Adult Patients

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Background: Focal segmental glomerulosclerosis (FSGS) is a common cause of nephrotic syndrome (NS) and end-stage renal disease (ESRD), particularly in African-Americans. Many studies have shown that clinical characteristics and outcomes differ depending on pathologic variants of FSGS. However, these are not well defined in Asian population.

Method: We conducted a retrospective cohort study to evaluate clinical features and outcomes according to pathologic variants of FSGS in 111 adult patients between January 2004 and February 2013. Primary outcome was a composite of doubling of the baseline serum creatinine concentrations (D-SCr) or the onset of ESRD. Secondary outcome included the rate of complete remission (CR) or partial remission (PR).

Results: Mean age at diagnosis was 47.2±16.2 years and 40% were male. There were 70 (63.1%), 20 (18.0%), 17 (15.3%), 3 (2.7%), and 1 (0.9%) patients with not otherwise specified (NOS), tip, perihilar, cellular, and collapsing variants, respectively. 42 (37.8%) patients presented NS and 50 (45.0%) patients were treated with immunosuppressants. NS occurred more commonly in patients with tip lesion than in those with other variants. During a mean follow-up of 39.1 months, 16 (14.4%) patients reached the composite of D-SCr or ESRD. CR and PR were achieved in 29 (26.1%) and 35 (31.5%) patients, respectively. The overall 5-year and 8-year renal survival rates were 76.8% and 56.3%. There was no difference in the development of primary outcome between patients with NOS, tip, and perihilar variants after adjustment of age, sex, eGFR, proteinuria, and immunosuppression. However, tip lesion was associated with a significantly increased probability of achieving CR. In addition, corticosteroid use resulted in higher rate of CR or PR.

Conclusion: Similar to other populations, Korean adult patients with FSGS have distinct clinical features except rare frequency of cellular and collapsing variants. Although pathologic variants were not associated with overall outcome, tip variant exhibited better outcome in terms of achieving CR. Further studies with a larger sample size are required to delineate long-term outcome and response to treatment of the pathologic variants.

Key Words: 초점성 분절성 사구체 경화증, 병리, 예후
FSGS, Pathology, Outcome