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Clinical Features and Long-Term Outcomes of Nephrotic Syndrome in Patients with IgA Nephropathy

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Nephrotic syndrome (NS) is a rare manifestation of IgA nephropathy. We conducted a multicenter observational study of 1,076 patients with IgA nephropathy to delineate clinical characteristics and long-term outcomes of this rare condition. The primary endpoint was a doubling of the baseline serum creatinine concentration. Of these patients, 100 (10.2%) presented with NS; complete remission (CR), partial remission (PR), and no response (NR) occurred in 48 (48.0%), 32 (32.0%), and 20 (20.0%) patients, respectively. During the median follow-up of 44.0 months, 24 patients (24.0%) in the NS group reached the primary endpoint compared to 63 (7.2%) in the non-NS group ($p<0.001$). The risk of primary endpoint was significantly higher in the PR (HR, 14.49; 95% CI, 1.14 to 183.7; $p=0.039$) and the NR group (HR, 215.97; 95% CI, 15.63 to 2983.6; $p<0.001$) than in the CR group. Among patients with NS, 24 (24.0%) underwent spontaneous remission (SR). SR occurred more frequently in female patients, patients with serum creatinine level $\leq$1.2 mg/dL, and a >50% decrease in proteinuria within 3 month after the onset of NS. None of the patients with SR reached the primary endpoint, with fewer relapses during follow-up. In conclusion, this study showed that the prognosis of NS in IgA nephropathy was not favorable unless PR or CR was achieved. In addition, SR was more common than expected, particularly in patients with preserved kidney function and a prompt decrease in proteinuria after the onset of NS, and these patients had excellent outcomes with a lower incidence of relapse.

Key Words: IgA nephropathy, Nephrotic syndrome, Spontaneous remission