

헤노흐쎈라인자색반 신장염 성인 환자에서 급속 진행성 사구체신염의 재발

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Relapse of Rapidly Progressive Glomerulonephritis in an Adult Patient with Henoch-Schönlein Nephritis

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Introduction : Henoch-Schönlein nephritis (HSN) is an infrequent condition in adults. HSN Patients with extensive glomerular crescent formation and acute renal failure carry a grave renal outcome. Therefore, remission and relapse in crescentic HSN are unusual. We herein describe a rare case that developed a rapidly progressive glomerulonephritis twice in a male adult patient with HSN during a course of follow-up.

Case : A 33-year-old previously healthy male patient was referred to the hospital for further evaluation of multiple red purpuric rashes and unspecified renal failure. His serum creatinine at that time was 1.65 mg/dL with 24 hr urine protein loss of 2.54 g/day. Skin biopsy showed the findings consistent with Henoch-Schönlein purpura (HSP) and renal biopsy revealed that 23.1% of total glomeruli had cellular crescents and the deposition of IgA and complements was mild staining in the study of immunofluorescence, which findings were consistent with crescentic glomerulonephritis associated with HSN. Treatment with oral prednisolone continued at our clinic. Renal function improved with a serum creatinine decrease to below 1.2 mg/dL. The patient experienced no symptoms or signs associated with vasculitis during a follow-up period of 57 months. 57 months later, he was readmitted with a suddenly developed generalized edema. 24 hr urine protein loss was 6.7 g/day and serum creatinine increased to 4.14 mg/dL. Repeated renal biopsy showed that 54% and 8% of total glomeruli had fibrocellular and fibrous crescents, respectively. He was treated with high-dose steroids and plasmapheresis. Although there was a slight response initially, he eventually developed irreversible renal failure and required hemodialysis.

Conclusion : This unusual case shows that crescentic glomerulonephritis associated HSN relapsed after approximately 5 years' remission. Through unknown mechanisms, non-specific or unidentified insult might be able to trigger relapse.

Key Words : 헤노흐쎈라인자색반 신장염, 급속 진행성 사구체신염, 신부전
Henoch-Schönlein nephritis, Crescent, Renal failure