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현미경적 다발성 동맥염(Microscopic Polyangiitis) 환자의 7년간의 장기 추적관찰 1례

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Seven Year Follow Up of Microscopic Polyangiitis Progressed to Chronic Sclerosing Glomerulonephritis: A Case Report

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Introduction : Microscopic polyangiitis (MPA) is a systemic necrotizing vasculitis that involves many organ systems including the skin, joint, kidneys, and lungs. The diagnosis of MPA is often difficult to make, and delayed because of the variability of the clinical presentation. Also in spite of early diagnosis and intensive care, 5year renal survival rate is 55%, and 5year patient survival rate is 65%.

Case : We experienced a case in 7- year- old girl of microscopic polyangiitis with positive serum perinuclear antineutrophil cytoplasmaic autoantibodies (P- ANCA), associated with pulmonary hemorrhage and gastrointestinal bleeding. The patient was managed under intensive care and has been following up since March 2001. The diagnosis of patient's first renal biopsy was MPA, P- ANCA associated crescentric glomerulonephritis. The patient's second renal biopsy was done 5years 6months later since first renal biopsy, and pathologic diagnosis was chronic sclerosing glomerulonephritis, advanced, due to MPA. We began methylprednisolone pulse therapy, combined with a low dose of cyclophosphamide and plasmapheresis therapy in 2001, and ACE inhibitor, Angiotensin II receptor blocker, cyclophosphamide were used until now and the patient's current age is 14 years old. The patient's laboratory findings showed BUN 117 mg/dL/Cr 2.3 mg/dL at initiation of disease, BUN 30.5 mg/dL /Cr 1.6 mg/dL in 2004, BUN 30.7 mg/dL/Cr 1.6 mg/dL in 2006. But renal function was progressed to chronic renal failure with latest laboratory data BUN 51.7 mg/dL and Cr 3.2 mg/dL.

Conclusion : We report a case of long- term follow- up and survival for 7 years in microscopic polyangiitis with chronic sclerosing glomerulonephritis. ACE inhibitor, Angiotensin II receptor blocker and small dose of immunosuppressant with close observation is the key to maintain the patient survival.

Key Words : 현미경적 다발성 동맥염, 추적관찰, 만성 신부전
Microscopic Polyangiitis, Follow up, Chronic renal failure