

## 복합 감염에 의해 발생한 급속진행성 반월상 사구체신염

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### Rapid Progressive, Pauci-immune Crescentic Glomerulonephritis by Multiple Infections

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**Background:** Crescentic glomerulonephritis are characterized by a crescent shaped cellular proliferation that may lead to glomerular destruction. Rapidly progressive glomerulonephritis (RPGN) results in a rapid decrease in the glomerular filtration rate of at least 50% over a short period. RPGN is classified pathologically into 3 categories: (1) anti-GBM antibody disease (3%), (2) immune complex disease (45%), and (3) pauci-immune disease (50%). Diagnosis is suspected in the context of extra-renal symptoms or immunological abnormalities, and confirmed by renal biopsy.

**Case:** A 13-year old girl was admitted because of high fever, nausea, and dyspnea. On admission, chest x-ray showed necrotizing pneumonia and effusion on both lung fields. Laboratory investigations showed WBC 77,800/mm<sup>3</sup>, Hemoglobin 4.9g/dL, Platelet 684k, BUN/Cr 187/15.8 mg/dL, Na/K/Cl 128/7.0/88 mol/L, protein/albumin 6.8/2.5 g/dL, ASO 477IU/mL, C3 below 16.5 mg/dL, C4 17.7 mg/dL, cold agglutinin 1:128, mycoplasma antibody 1:20,480, mycoplasma PCR negative, Streptococcus pneumoniae isolated on blood culture, and adenovirus IgM antibody was positive. We started peritoneal dialysis and medical treatment. Although pneumonic infiltration and laboratory findings were improved gradually with treatments, microscopic hematuria, proteinuria, and uremia state were persistent. Renal biopsy showed global sclerosis and crescent formation on LM, and paramesangial and mesangial electron dense deposit on EM but IF was negative. We started methylprednisolone pulse therapy but tapered due to steroid induced diabetes mellitus. We started low dose cyclophosphamide pulse therapy monthly with oral mizoribine. Proteinuria was reduced over several months and renal function was improved gradually. We performed follow-up biopsy after 11 months. On second biopsy, laboratory findings showed WBC 6,560 mm<sup>3</sup>, Hemoglobin 11.7 g/dL, Platelet 332k, BUN/Cr 22/1.0 mg/dL, Na/K/Cl 140/5.0/109 mol/L, protein/albumin 6.8/3.9 g/dL, ASO 70 IU/mL, C3 109 mg/dL, C4 22.5 mg/dL, mycoplasma antibody negative, and 24-hour urine protein was 140mg/day. Follow-up renal biopsy showed fibrocellular crescent formations in 8 of 10 glomeruli. The tubules and interstitium showed diffuse heavy infiltration of chronic inflammatory cells with pronounced tubular atrophy and fibrosis.

**Conclusion:** This patient was considered as c-ANCA associated pauci-immune glomerulonephritis by multiple infections. However, it is possible that she could progress to chronic renal failure although clinical manifestation was improved after administration of cyclophosphamide with oral mizoribine.

**Key Words:** 급속 진행성 사구체신염, 복합감염, 급성 신부전증  
PSGN, Multiple infections, ARF