

Case 2

A case of crescentic glomerulonephritis by severe infection

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[Background]

Crescentic glomerulonephritis are characterized by a crescent shaped cellular proliferation that may lead to glomerular destruction. Patients present rapidly progressive glomerulonephritis (RPGN) with renal failure, proteinuria and hematuria. Diagnosis is suspected in the context of extra-renal symptoms or immunological abnormalities, and confirmed by renal biopsy.

[Case presentation]

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 77,800mm³, Hemoglobin 4.9g/dL, Platelet 684k, BUN/Cr 187/15.8 mg/dL, Na/K/Cl 128/7.0/88 mol/L, protein/albumin 6.8/2.5g/dL, ASO 477IU/mL, C3 below 16.5mg/dL, C4 17.7mg/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 hyperglycemia. 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 6,560mm³, Hemoglobin 11.7g/dL, Platelet 332k, BUN/Cr 22/1.0 mg/dL, Na/K/Cl 140/5.0/109mol/L, protein/albumin 6.8/3.9g/dL, ASO 70IU/mL, C3 109mg/dL, C4 22.5mg/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. Granular depositions of IgA and C3 were observed on IF.

[Points of Discussion]

1. Was she infected with Mycoplasma, Adenovirus, and Streptococcus pneumoniae altogether?
2. Isn't there any evidence of immune deficiency for her?
3. Is it possible that she had already had IgA nephropathy and progress to crescentic glomerulonephritis by severe infection?
4. Follow-up biopsy showed fibrocellular crescent formations in 80%, tubular atrophy and fibrosis.

How to treat this patient?