A Case of Crescentic Glomerulonephritis Associated with Polymyositis

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Dear Sir,

Polymyositis is an autoimmune collagen disease which preferably involves proximal striated muscles. Though other collagen diseases, such as systemic lupus erythematosus (SLE) and progressive systemic sclerosis (PSS), involve various organs including the kidney, polymyositis is rarely complicated with renal disease. We hereby describe an adult patient with polymyositis which was followed by crescentic glomerulonephritis. Clinical course and laboratory examination suggest an association of two immune-mediated diseases: crescentic glomerulonephritis and polymyositis in this case.

The patient, a 56-year-old man, was first seen at another hospital in 1985 because of progressive muscle weakness of the trunk and lower extremities. After several examinations, including muscle biopsy of the deltoid muscle, a diagnosis of primary idiopathic polymyositis was established in 1985, according to the criteria offered by Bohan and Peter [1]. Prednisolone treatment (30 mg/day) was started, with subsequent reversal of muscle weakness and improvement in laboratory data. In July 1990, however, laboratory data showed exacerbation of polymyositis. Serum creatine phosphokinase (CK) gradually increased to 400-529 IU (normal range 30-160) and aldolase was 9.1-10.9 BU (normal range 2-8). Cyclophosphamide (100 mg/day) was subsequently added to steroid therapy. In December 1990, macroscopic hematuria was noted. Serum creatinine was 1.3 mg/l. The patient was admitted to Osaka City University Hospital in May 1991. On physical examination, the patient was 167 cm tall and weighed 58 kg. His temperature was 36.8°C and blood pressure 166/68 mm Hg. His chest was normal except for occasional irregular heartbeats. There was no edema in the extremities. Neurological examination revealed symmetric weakness and atrophy of the muscles of the trunk and extremities.

Laboratory findings on admission were as follows: urine gave a 1 + test for protein (1.52 g/day) and 3 + for occult blood, urinary sediment contained 120-550 red cells, 3-7 white cells, a few granular casts and hyaline casts per high-power field. Erythrocyte sedimentation rate was 50 mm

in 1 h. RBC was $297 \times 10^6/\text{mm}^3$, Hb 9.5 g/dl, WBC $4,700/\text{mm}^3$ and platelet count $11.6 \times 10^9/\text{mm}^3$. Serum total protein was 7.5 g/dl, with low value of serum albumin (3.6 g/dl) and elevated γ -globulin (2.3 g/dl). Serum transaminase was normal. CK was 165 IU and aldolase 5.0 BU. Blood urea nitrogen was 25 mg/dl and creatinine 1.8 mg/dl. Serum and urinary myoglobin was elevated, being 140 (normal range: < 50) and 15 ng/ml (normal range: < 5), respectively. Rheumatoid factors, antinuclear antibody and anti-DNA antibody were negative. Serum complements were normal. Circulating immune complex measured by C1q binding assay was $47.1 \mu\text{g/ml}$ (normal range 0-30). Renal biopsy was performed on June 18, 1991. Two of 27 glomeruli showed global sclerosis

on light microscopy. The remaining glomeruli of them were mildly to moderately collapsed with little hypercellularity. Focal and segmental expansions of mesangial matrix were seen. Cellular or fibrocellular crescents were seen in approximately 50% of glomeruli (fig. 1). Most tubules were atrophic, and extensive interstitial infiltration of mononuclear cells was seen. On electron microscopy, small dense deposits were seen in glomerular basement membranes. Unfortunately, the specimen for immunofluorescence did not contain glomeruli. Histopathological diagnosis of 'sclerotic glomerulonephritis with fibrocellular crescent' was made.

After renal biopsy, a combined therapy of plasma exchanges (PE) 3 times a week and oral administration of prednisolone (20 mg/day) was started. The concentration of circulating immune complex markedly decreased in the first week of the treatment and renal function improved in 1 month; serum creatinine decreased to 1.5 from 1.8 mg/dl. Ten months after treatment, serum creatinine was still 1.1-1.2 mg/dl.

In the present case, crescentic glomerulonephritis was found 5 years after the onset of polymyositis. Though serum CK and aldolase were not so highly elevated on admission in 1991, ESR was at a high level and the systemic weakness of limb-girdle muscles had been progressing since late in 1990, when skeletal muscle enzymes were at one of the highest levels in the clinical course (CK was

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400-529 IU and aldolase 9.1-10.9 BU). Activity of polymyositis was very high in 1990, when the onset of hematuria caused by glomerulonephritis was seen. This clinical course could suggest an association of polymyositis with glomerulonephritis in this case. Dense deposits seen on electron-microscopic renal biopsy specimens and demonstration of circulating immune complex suggest that the crescentic glomerulonephritis of this case was caused by an immune complex, i.e., type II

rapidly progressive glomerulonephritis with an antigen-antibody complex according to the classification of Lewis [2]. The clinical course shows that, after disappearance of circulating immune complex by use of plasma exchange in the first week, renal function gradually improved. The clinical course after therapy also supports the theory that glomerulonephritis of the case was caused by an immune complex. Causer et al. [3] reported immune-mediated crescentic glomerulone-

phritis in collagen vascular disease. Dyck et al. [4] reported the possibility of immune-mediated renal diseases induced by immune complex consisting of myoglobin-anti-myoglobin antibodies in polymyositis. In this case, although no anti-myoglobin antibody was examined, serum myoglobin and circulating immune complexes were at a high level on admission. These

laboratory findings may also suggest a positive association of polymyositis and crescentic glomerulonephritis in the present case. Polymyositis is sometimes overlapped with other collagen diseases, such as SLE, rheumatoid arthritis or PSS. In these overlapped syndromes, glomerulonephritis can be seen. In the present case, however, no clinical or laboratory features indicated other collagen diseases overlapping with polymyositis. In patients with polymyositis, there have been few reports concerning the coexistence of glomerular disease, including crescentic glomerulonephritis. Kamata et al. [5] reported a rare case of childhood type polymyositis with crescentic glomerulonephritis. We consider that such association of polymyositis and crescentic glomerulonephritis is very rare in adults. The therapeutical role of plasma exchange with low-dose steroids remains still under discussion in the therapy might be one of the possible treatments in this case with elevated circulating immune complex.

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