

다발성 골수종 환자에서 공존한 원섬유 사구체 신염 1예

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Coexistence of Fibrillary Glomerulonephritis in a Patient With Multiple Myeloma: A Case Report

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Fibrillary glomerulonephritis is a rare glomerulopathy, it diagnosed by randomly deposit of amyloid-like fibrills on mesangium and microvascular wall on electron microscopy. These fibrills were negative for Congo red staining. It usually presents with nephrotic range proteinuria, microhematuria, hypertension and renal insufficiency, and it has poor prognosis. We present a case of the pathological characteristic fibrillary glomerulonephritis in a patient with multiple myeloma.

A 71-year-old man who had a no specific past history was admitted for both leg edema for 2 weeks. The blood pressure was 160/80 mmHg and other findings were not notable. Blood urea nitrogen was 29 mg/dL and serum creatinine was 2.9 mg/dL. 24 hour urinary protein was 6348 mg/day. Urinary and serum immunoelectrophoresis showed positive for the Kappa light chain and a bone marrow aspiration test presented 18.3% of plasma cell infiltration. These finding confirmed his diagnosis of multiple myeloma. ANA was positive and it's titer was 1:80. ANCA and serum cryoglobulin were negative. Serum total protein and albumin were 4.4 and 2.7 g/dL. His renal biopsy specimen consists of 1 piece of cortex and medulla containing 21 glomeruli, 8 of which are globally sclerosed. The glomeruli demonstrated nodular expansion of mesangial areas. There are occasional proteinaceous casts, without crystals. There is 15–20% early interstitial fibrosis and edema, proportional to tubular atrophy. The interstitium shows patch infiltration of lymphocytes and plasma cells. The arterioles show intimal hyalinosis. Immunohistochemistry for kappa light chain demonstrates positive (1+) staining of the mesangium and capillary loop walls. Lambda light chain shows weakly, segmental staining on the capillary loop walls. Proteinaceous casts are positive for kappa and lambda light chain. On electron microscopy, fibrillary deposits in mesangial matrix and subendothelium were increased by randomly arranged 12 nm nonbranching fibrills. We report an unusual case of coexistence of fibrillary glomerulonephritis in a patient with multiple myeloma.

Key Words: 원섬유 사구체신염, 경쇄, 다발성 골수종

Fibrillary glomerulonephritis, Light chain, Multipel myeloma

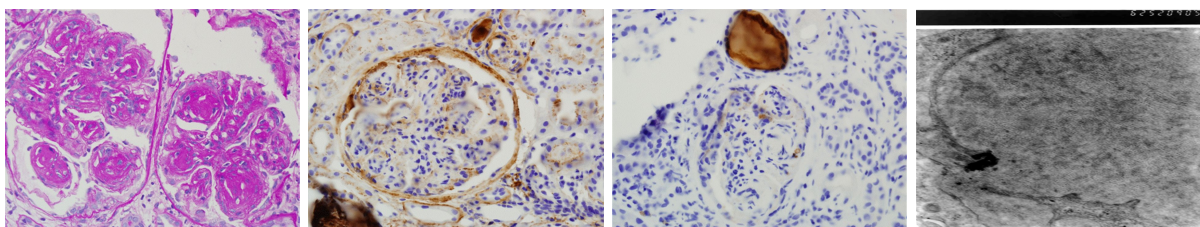


Fig. 1. Light photomicrograph revealed mild periodic acid-Schiff positive mesangial expansion (×400) Fig. 2. Immunohistochemistry for lambda light chain demonstrated segmental staining of capillary loop wall (×400) Fig. 3. Immunohistochemistry for kappa light chain demonstrated positive staining of the mesangium and capillary loop wall (×400) Fig. 4. Electron microscopy showed depositions of randomly arranged fibrils in the mesangium (×25,000)