

만성 염증성 탈수초 신경병증을 동반한 면역보체음성 섬유원성 신염

대전을지대학병원 내과학교실 신장내과

정진욱, 방기태, 신종호

Immun Complex Negative Fibrillary Glomerulonephritis Combined with Chronic Inflammatory Demyelinating Polyneuropathy

Jinuk Jeong, Kitae Bang, Jongho Shin

Department of Nephrology, Eulji University School of Medicine

Fibrillary glomerulonephritis (FGN) is an idiopathic glomerular disease characterized by randomly arranged Congo red-negative fibrils of 16-24 nm in diameter. The glomerular deposits are predominantly composed of polyclonal IgG and C3. A 58-yr-old man was admitted to our hospital because of leg edema progressive both leg weakness, which had appeared more than two months before admission. He had paresthesia of both extremities also. He has history of diabetes mellitus and hypertension for 10 years. On laboratory investigation, urinalysis revealed 3+ protein and rare red and white blood cells. A 24-h urine collection revealed 4.7 g of protein. The serum chemistry disclosed the following: blood urea nitrogen 38 mg/dL, creatinine 1.73 mg/dL, total protein 6.8 g/dL, albumin 4.2 g/dL, total cholesterol 91 mg/dL, respectively. Liver function tests were within normal. Serum immunofixation disclosed a monoclonal gammopathy (lamda type). Urine immunofixation disclosed a monoclonal immunoglobulin light chain (lamda type). On the renal biopsy, the glomeruli show variable mesangial widening in the absence of significant hypercellularity and focal capillary wall thickening, probably due to the accumulation of amorphous extracellular material in the mesangium and capillary walls. The degree of PAS staining displays a slightly uneven texture of coarseness and irregularity. On electron microscopy reveals fibrillary glomerular deposits of randomly arranged, straight, nonbranching fibrils, ranging in diameter from 12 to 20 nm, disrupting the glomerular basement membrane and on the mesangium. The foot process effacement on the podocytes is present. The capillary walls shows focally wrinkling and irregular thickening. A Congo red stain for amyloid was negative. We report on a case exhibiting the pathological characteristics of FGN and CIDP with monoclonal gammopathy of undetermined significance (MGUS).

Key Words: 섬유원성 신염, 탈수초성 신경병증, 단클론감마병증
CIDP, Fibrillary glomerulonephritis, MGUS