

MGUS와 동반된 Fibrillary glomerulonephritis

부산대학교 의학전문대학원 내과학교실¹, 부산대학교 의학전문대학원 병리학교실²

박종만¹, 신지연¹, 신나리², 송상헌¹, 이하린¹, 정우진¹, 박수민¹, 강진숙¹
황나경¹, 김일영¹, 이동원¹, 이수봉¹, 성은영¹, 곽임수¹

Fibrillary Glomerulonephritis Associated with Monoclonal Gammopathy of Undetermined Significance

Jong Man Park¹, Ji Yeon Shin¹, Nari Shin², Sang Heon Song¹, Harin Rhee¹, Woo Jin Jung¹
Su Min Park¹, Jin Suk Kang¹, Nakyoung Hwang¹, Il Young Kim¹, Dong Won Lee¹
Soo Bong Lee¹, Eun Young Seong¹, Ihm Soo Kwak¹

Department of Internal Medicine¹, Pusan National University School of Medicine, Busan, Korea
Department of Pathology², Pusan National University School of Medicine, Busan, Korea

Fibrillary glomerulonephritis (FGN) is a rare disease diagnosed by deposition of fibrillary material in glomerulus. Generally FGN was associated with conditions of immune system dysfunction or chronic immune system activation. We report the case of 70-yr-old woman with a diagnosis of FGN. She presented to the outpatient clinic for general edema and the initial blood lab showed hypoalbuminemia, nephrotic level of proteinuria and dyslipidemia. She admitted to evaluate nephrotic syndrome and to manage general edema and hypertension. A renal biopsy was performed to evaluate the cause of nephrotic syndrome. Light microscopy study showed lobular accentuation pattern with severe endocapillary, mild mesangial hypercellularity and extracapillary hypercellularity. On immunofluorescence study, IgG, IgA, Kappa and Lambda were positive on glomerular capillary wall and mesangium. On electron microscopy study, variable fibrillary deposits (12-17nm) were identified on the mesangium, subendothelium, and subepithelium. Congo red stain was negative. Fibrillary Glomerulonephritis was diagnosed. Electroporesis and immunofixation study showed monoclonal band in lambda region. The aspirate smear of the bone marrow showed infiltration of increased plasma cells (8% among total nucleated cells). Monoclonal gammopathy of undetermined significance (MGUS) was confirmed. For FGN combined with MGUS, treatment was started with intravenous methylprednisolone and followed by oral prednisolone. She was followed in the outpatient clinic. To our knowledge, there has not been reported FGN combined with MGUS in Korea and only several cases were reported in the English literature. This report showed a rare case of FGN with MGUS.

Key Words: 원삼유성 사구체신염, MGUS
FGN, MGUS