

막성사구체신염에서 발생한 폐포단백증

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Membranous Nephropathy and Pulmonary Alveolar Proteinosis: A Case Report

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Membranous nephropathy is one of the most common causes of the nephrotic syndrome in the adults. In a majority of idiopathic membranous nephropathy, circulating autoantibodies against M-type phospholipase A2 receptor (mainly IgG4) are deposited on the subepithelial side of the glomerular basement membrane. Pulmonary alveolar proteinosis (PAP) is a rare lung disease in which abnormal accumulation of surfactant occurs within the alveoli. Autoimmune PAP accounts for the majority of cases and is caused by autoantibodies to granulocyte-macrophage colony-stimulating factor, which results in impaired catabolism and clearance of surfactant lipids and proteins. Up to date, only 2 cases of the concurrence of membranous nephropathy and PAP have been described and this case report is the first case in Korea. Here, we report a 61-year-old man with membranous nephropathy and PAP, simultaneously.

He was referred to our hospital because of nephritic syndrome. He did not have any respiratory symptoms at time of admission. He had been diagnosed with diabetes for 20 years. Biochemistry showed serum protein of 4.4 g/dl, albumin of 1.6 mg/dl, total cholesterol of 534 mg/dl, blood urea nitrogen of 19.3 mg/dl, and creatinine of 0.94 mg/dl. Urinary excretion of protein was 8.3 g/day. A routine chest x-ray showed ground glass opacity (GGO) and consolidation in the both lung and chest CT scan revealed diffuse GGO with reticulation, so called crazy paving appearance in the both lung. Renal biopsy was performed and the result was consistent with membranous nephropathy. Broncho-alveolar lavage (BAL) was performed and displayed increased cell numbers with a dominant lymphocyte and Periodic acid-Schiff positive proteinous components are found. Based on these findings, we diagnosed with concurrence of membranous nephropathy and PAP. He has taking mycophenolate mofetil and his proteinuria was reduced to 0.8 g/day, however, PAP was worsening and required whole lung lavage.

Key Words: 막성사구체신염, 폐포단백증, 자가면역질환

Membranous nephropathy, Alveolar proteinosis, Autoimmune