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미만성 폐출혈과 막증식성사구체신염을 동반한 항인지질증후군 치료경험 1례

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Antiphospholipid Syndrome with Membranoproliferative Glomerulonephritis Accompanied by Diffuse Pulmonary Hemorrhage

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Background: Pulmonary renal syndrome, a glomerular disease associated with pulmonary hemorrhage and glomerulonephritis, is rare but can be fatal. It is usually caused by Wegener's granulomatosis, microangiopathic polyarteritis, and Goodpasture syndrome. However, a rare case of antiphospholipid antibody syndrome (APS), which is representative of a pulmonary renal syndrome, was recently identified.

Case presentation: The 43-year-old man was admitted with chest pain and edema. The patient had renal failure and pulmonary hemorrhage. Anticardiolipin antibody and lupus anticoagulant were detected, and kidney biopsy results showed membranoproliferative glomerulonephritis (MPGN). The patient was diagnosed with hemorrhagic antiphospholipid syndrome and treated with high-dose steroids. Although the patient responded well to the treatment even in the second relapse, the response was not maintained. So several sessions of plasmapheresis were performed, and immunosuppressants, including rituximab and bortezomib were used additionally. The patient's clinical symptoms improved through several months after treatment. However the patient's renal function declined again and he has been undergoing hemodialysis.

Conclusion: APS concomitant with MPGN accompanied by diffuse pulmonary hemorrhage is a rare disease and various treatment strategies including plasmaphresis, rituximab and bortezomib can be applied

Key Words: 항인지질증후군, 막증식성사구체신염, 미만성 출혈
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