A Case of Different Organ Responses to Immunosuppressive Therapy In a Microscopic Polyangiitis Patient with Interstitial Lung Disease

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Microscopic polyangiitis (MPA) is an idiopathic autoimmune disease characterized by systemic vasculitis associated with antineutrophil cytoplasmic autoantibodies. Interstitial lung disease is a less recognized manifestation of MPA and has a poor prognosis. A 61-year-old man presented with persistent cough, sputum and dyspnea. Laboratory examination revealed microscopic hematuria and renal insufficiency. Perinuclear anti-neutrophil cytoplasmic autoantibodies were positive according to serological testing. Computed tomography scans showed bibasilar reticulation and honeycombing in a peripheral distribution. Therefore, renal biopsy was performed, and MPA was diagnosed. After treating with corticosteroids and immunosuppressive agents, the patient had a complete renal response but progressive interstitial lung disease. We report a case of MPA presenting with interstitial lung disease in which the patient experienced different responses in each organ. (Korean J Med 2014;86:84-88)

Keywords: Microscopic polyangiitis; Interstitial lung disease; Anti-neutrophil cytoplasmic antibody-associated vasculitis

INTRODUCTION

Microscopic polyangiitis (MPA) is an idiopathic autoimmune disease. It is characterized by systemic vasculitis that affects the small blood vessels, such as arterioles, venules, and capillaries, and it is associated with anti-neutrophil cytoplasmic autoantibodies (ANCA). MPA is one of the major causes of pulmonary-renal syndrome, along with Goodpasture’s syndrome, systemic lupus erythematosus, and Wegener’s granulomatosis. Depending on the extent of systemic vascular involvement, clinical findings can be rather variable with cutaneous, musculoskeletal, neurological and gastrointestinal symptoms, as well as renal and pulmonary...
symptoms. Pulmonary involvement is seen in 25-55% of MPA patients, with hemoptysis being the predominant symptom as a result of alveolar hemorrhage. However, pulmonary involvement also consists of pulmonary infiltrates, pleural effusion, pulmonary edema, pleuritis, and interstitial fibrosis. Interstitial lung disease (ILD) is a less recognized pulmonary manifestation of MPA. It can present anywhere from years prior to years after MPA diagnosis. Glucocorticoid, cyclophosphamide and mycophenolic mofetil (MMF) are typically used for induction therapy, while azathioprine is used for maintenance therapy. We report a case of MPA associated with ILD in which the patient experienced different organ responses to immunosuppressive therapies.

CASE REPORT

A 61-year-old man presented with a 2-month history of persistent cough with productive sputum and a 10-day history of dyspnea. He was an ex-smoker who stopped smoking 18 years ago. He had a history of hypertension and worked as a carpenter. On admission, auscultation of the lungs revealed bibasilar crackles without wheezing. Additionally, there was no clubbing, evidence of congestive heart failure, or cor pulmonale. A plain chest radiograph demonstrated reticulation in both lower lung fields (Fig. 1A), and computed tomography (CT) scan showed bibasilar reticulation and honeycombing in a peripheral distribution (Fig. 1B). Pulmonary function tests revealed a normal pattern with a mildly decreased diffusing capacity of the lung for carbon monoxide (DLCO). Blood tests revealed a white cell count of 20,640/mm³ (93.8% neutrophils), hemoglobin level of 10.5 g/dL, hematocrit level of 30.3%, and platelet count of 314,000/mm³. The serum creatinine level was elevated to 6.42 mg/dL, and the estimated glomerular filtration rate was 9.44 mL/min. Biochemical tests revealed a urea nitrogen level of 77 mg/dL, glucose level of 281 mg/dL, total protein level of 7.5 g/dL, albumin level of 2.9 g/dL, aspartate transaminase (AST) level of 25 IU/L, alanine aminotransferase (ALT) level of 37 IU/L, alkaline phosphatase level of 183 IU/L, and total bilirubin level of 0.59 mg/dL. The C-reactive protein level was 21.68 mg/dL. The 24-hr urine protein was 478.2 mg/day, and microscopic urinalysis showed several RBCs per high-power field. Furthermore, serologic investigation revealed perinuclear ANCA (p-ANCA) with a titer of 1:640. Anti-glomerular basement membrane (anti-GBM) antibodies and antinuclear antibodies (ANA) were negative.

Renal biopsy revealed two global and four focal segmental sclerotic glomeruli out of 15 total glomeruli. Two cellular and fibrocellular crescent formations were present (Fig. 2A). Acute necrotizing arteritis was evident in the smaller vessels, showing mural and perivascular fibrinoid necrosis, with predominating neutrophils as well as occasional mononuclear leukocytes, including

![Figure 1](image1.png)

**Figure 1.** Chest X-ray and chest CT on the day of admission. (A) Reticulation in both lower lung fields. (B) Bibasilar reticulation and honeycombing in the peripheral lesion.