

Diffuse Alveolar Hemorrhage in a 39-year-old Woman: Unusual Initial Presentation of Microscopic Polyangiitis

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Microscopic polyangiitis (MPA) is a necrotizing vasculitis involving the small vessels without granulomatous inflammation. Most MPA initially presents with renal involvement without pulmonary involvement. Isolated and initially presenting alveolar hemorrhage is very rare. The patient was a 39-year-old female with a progressive cough, dyspnea, and blood-tinged sputum for the previous 5 days. We determined that her condition was MPA though VATS lung biopsy and renal biopsy. After 2 months of steroid therapy, the chest lesions had improved. We report here a rare case of MPA with isolated and initial involvement of the lung with a review of the literature.

Key words: 1. Hemoptysis
2. Lung
3. Vascular disease

CASE REPORT

A 39-year-old woman visited the emergency room with a progressive cough, dyspnea, and blood-tinged sputum for the previous 5 days. The patient did not use tobacco or alcohol, nor did she have any specific medical history. At initial presentation, her blood pressure was 107/54 mmHg, pulse rate 74 beats per minute, respiration rate 22 breaths per minute, and body temperature 36.8°C. The patient was wide awake and well oriented. There were no other suggestive symptoms. Here CBC count was as follows: white blood cell count 8,420/mm³ (75.1% granulocytes, 17.7% lymphocytes, 5.6% monocytes and 0.4% eosinophils), hematocrit 25.2 mg/dL and platelets 374 K/mm³. Her serum chemistry and liver function were normal. There was no serologic evidence of hepatitis. A urine analysis was non-specific. A chest PA revealed an in-

creased opacity throughout both lower lung fields and a chest CT revealed extensive ground-glass opacities in both whole lung fields without any lymphadenopathy (Fig. 1). A sputum study was negative for acid-fast bacilli and blood culture findings were negative. Bronchoscopy findings were compatible with pulmonary alveolar hemorrhage at RML and RLL. At first we considered the case to be a simple alveolar hemorrhage associated with pneumonia, so we treated her conservatively and her symptoms, including hemoptysis, improved. On the 5th day from admission, her urine color changed abruptly to bloody and we performed further evaluation for simultaneous hematuria and hemoptysis. Serum protein electrophoresis revealed a decrease in total protein albumin but a normal immunotyping pattern. There was no serologic evidence of hepatitis. Serum antinuclear antibody, anti-glomerular basement membrane antibody (anti-GBM), mycoplasma

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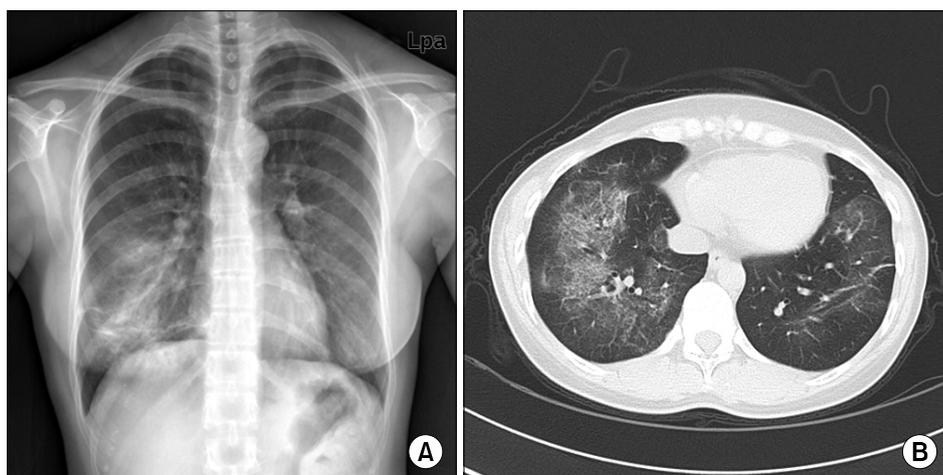


Fig. 1. Chest X-ray (A) and Chest CT (B) showed extensive ground-glass opacities in both whole lung fields without any lymphadenopathy.

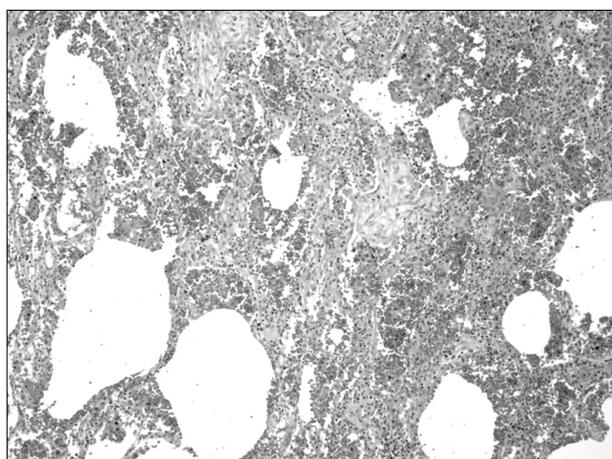


Fig. 2. Lung specimen showed intraalveolar hemorrhage with aggregation of pigment-laden macrophages and a few neutrophils suggesting capillaritis (hematoxylin-eosin stain, x100).

pneumonia, and mycoplasma tuberculosis titers were all negative, Complement levels (C3, C4, and CH50) were normal but anti MPO antibody (>100) and p-ANCA (1 : 160) were positive. For accurate diagnosis, we performed a VATS lung biopsy on the 9th hospital day and renal biopsy on the 14th hospital day. The pathology report showed the lung lesion was intralveolar hemorrhage suggesting capillaritis and the kidney lesion was focal segmental necrotizing glomerulonephritis without antibody deposition at the capillary basement membrane (Fig. 2, 3). Therefore, we diagnosed this condition as microscopic polyangiitis presenting with initial pulmonary involvement and we started her on steroid therapy. After 2

months of steroid therapy, her chest lesions had improved but hematuria remained (Fig. 4). More treatment will be needed.

DISCUSSION

There are many causes for hemoptysis or diffuse alveolar hemorrhage. One of them is vasculitis, which is rarely encountered in the thoracic surgery field [1,2]. Vasculitis is an inflammatory destructive process affecting blood vessels and surrounding tissue through diverse mechanisms. Microscopic polyangiitis (below MPA) is a necrotizing vasculitis involving the small vessels without granulomatous inflammation [3]. MPA is the most common antineutrophilic cytoplasmic antibody (ANCA)-associated small vessel vasculitis and is characterized by a lack of immune deposits in the involved vessels [1,4]. The incidence of MPA is about 1 : 100,000 with a slight predominance in men. The mean age of onset is about 40 or 50 years [4]. The kidney is the most frequently involved organ in 90% of patients. The lung is involved in 22%, and alveolar hemorrhage occurs in 11% [4,5]. Other organs and systems including the skin, GI tract, musculoskeletal system, heart, and nervous system can also be involved [3,6]. Most MPA initially presents as renal involvement without pulmonary involvement, while isolated and initially presenting alveolar hemorrhage is very rare [4]. The cause of MPA remains unknown, but some researchers have proposed genetic, environmental, and immunologic factors such as ANCA in the pathogenesis of MPA [1,4]. Chest radiographic findings

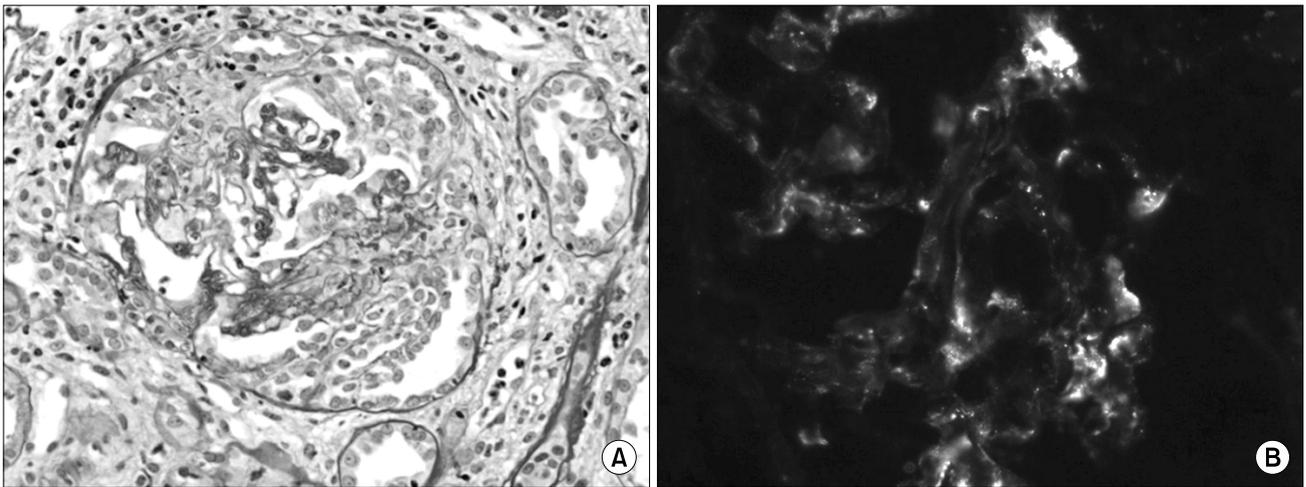


Fig. 3. Renal specimen shown was focal segmental necrotizing glomerulonephritis without antibody deposition at the capillary basement membrane (A: hematoxylin-eosin stain, $\times 400$, B: immunofluorescence stain, $\times 400$).

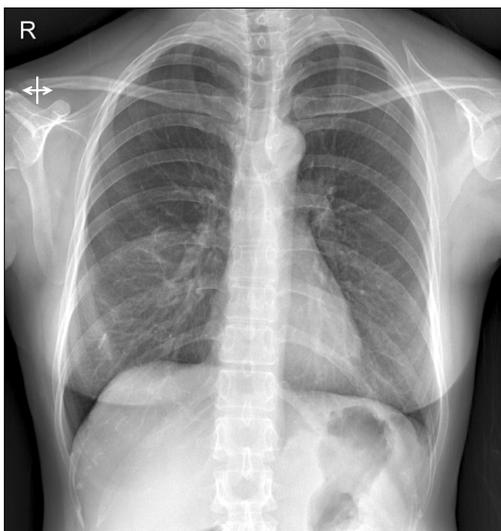


Fig. 4. Chest X-ray after steroid therapy for 2 months showed improved infiltration in both lung fields.

consist of ground-glass opacity, consolidation, thickened bronchovascular bundles and a honeycomb appearance caused by alveolar hemorrhage [2,4]. An ANCA test is used to aid in diagnosis and to monitor inflammatory activity in vasculitis including MPA, Wegener's granulomatosis, and Churg-Strauss syndrome [1,3]. Previous studies have reported that 50% to 80% of MPA patients test positive for ANCA [3,5].

Because lung biopsy can aggravate conditions of critically ill patients and pulmonary involvement usually follows after

renal involvement, lung biopsy is not usually recommended in the case of vasculitis-associated alveolar hemorrhage if the diagnosis can be obtained by biopsy of other organs and by a serum immunology study [1,4]. In our case, because of the unusual presentations, specifically, initial pulmonary involvement without renal involvement and hemoptysis improvement after conservative treatment, we needed a lung biopsy for accurate diagnosis. Lung specimens of MPA usually show only necrosis, nonspecific inflammation, and hemorrhage pulmonary capillaritis [1,4]. Renal specimens usually show focal segmental necrotizing glomerulonephritis without any immune deposition [1,4]. Our lung specimen showed intraalveolar hemorrhage with aggregation of pigment-laden macrophages and a few neutrophils, suggesting capillaritis and organizing pneumonia, and our renal specimen showed focal segmental necrotizing glomerulonephritis without immune deposits in the basement membrane. These lung and kidney specimen pathologies were consistent with pulmonary and renal involvement of MPA.

The differential diagnosis should include Goodpasture syndrome. Goodpasture syndrome is a rare but severe immunologic disease, which is characterized by rapidly progressing glomerulonephritis and diffuse intraalveolar hemorrhage [1,2]. However, in our case there was immune deposition at the capillary basement membrane [1], so we could rule out Goodpasture syndrome.

The mainstay of treatment of MPA is systemic corticosteroids and cyclophosphamide administration [3,6]. The treatment duration of at least 12 months of combined therapy has been recommended based on empirical research. Few papers have studied the course and prognosis of pulmonary involvement in MPA [4]. Alveolar hemorrhage usually improves within several days after the start of steroid therapy but can continue longer [2,4].

We describe a 39-year-old woman presenting with a progressive cough, dyspnea, and hemoptysis. We determined that her condition was MPA though VATS lung biopsy and renal biopsy. After 2 months of steroid therapy, her chest lesions had improved. We report here a rare case of MPA with isolated and initial involvement of the lung, along with a review of the literature.

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