Pneumomediastinum in a Patient with Microscopic Polyangiitis Preceded by Interstitial Pneumonia

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Abstract

A 73-year-old woman was diagnosed with interstitial pneumonia in 2006; however, the disease was not progressive. Four years later, purpura, peripheral neuropathy, and increased levels of myeloperoxidase anti-neutrophil cytoplasmic antibodies (583 EU/mL) and C-reactive protein (2.27 mg/dL) were observed, and a diagnosis of microscopic polyangiitis was made. Treatment with prednisolone and azathioprine was initiated. However, on the 35th hospital day, chest computed tomography showed pneumomediastinum and subcutaneous emphysema without aggravation of the interstitial pneumonia. To our knowledge, this is the first report of pneumomediastinum as a complication of microscopic polyangiitis associated with interstitial pneumonia.

Key words: ANCA-associated vasculitis, anti-neutrophil cytoplasmic antibodies (ANCA), interstitial pneumonia, microscopic polyangiitis (MPA), pneumomediastinum


Introduction

Microscopic polyangiitis (MPA) is a form of systemic necrotizing vasculitis characterized by positive myeloperoxidase anti-neutrophil cytoplasmic antibodies (MPO-ANCA) that affects small blood vessels without immune complex deposition and involves systemic organs, such as the skin, kidneys, heart and lungs (1). Pulmonary involvement is a common feature; however, to our knowledge, there are no reports of MPA associated with pneumomediastinum. We herein report the case of a patient with pneumomediastinum that developed during treatment of MPA secondary to interstitial pneumonia (IP).

Case Report

A 73-year-old woman was diagnosed with IP in 2006 based on X-ray and chest computed tomography (CT) findings. She was positive for anti-nuclear antibodies; however, no other disease-specific autoantibodies, including MPO-ANCA, were detected. The IP was not progressive, and the patient was followed by a general practice physician. In October 2010, she noticed diminished mobility, numbness and a sensory disturbance in the lower limbs. In November, edema and purpura of the lower legs appeared, and the movement disorder of the lower limbs was aggravated. The patient was therefore referred and admitted to our hospital. A neurological examination and nerve conduction study showed peripheral neuropathy predominantly involving the motor nerves in the lower limbs. A blood examination revealed increased levels of C-reactive protein (2.27 mg/dL) and MPO-ANCA (583 EU/mL); however, tests for proteinase 3-ANCA were negative. A diagnosis of MPA was made based on these findings. Treatment with prednisolone (50 mg/day) and azathioprine (50 mg/day) was initiated. The levels of C-reactive protein and MPO-ANCA decreased to 0.02 mg/dL and 52 EU/mL, respectively, concomitant with an improvement in the peripheral neuropathy, lower leg purpura and IP (Figure). However, on the 35th hospital day, chest CT (Figure) showed pneumomediastinum and subcutaneous emphysema without aggravation of the IP, although the patient had no related symptoms, such as dyspnea or neck pain. Because the MPA was not active, the treatment strategy was not altered, and oxygen therapy and rest were prescribed. The complete disappearance of the pneumomediastinum and subcutaneous emphysema was confirmed on a chest X-ray and chest CT scan approximately three months later.
considered it to be associated with the patient’s MPA. Therefore, the IP observed in this case cannot be associated with MPA, including purpura and peripheral neuropathy. In contrast to idiopathic IP. In this case, the patient’s IP improved following the initiation of treatment with prednisolone and azathioprine in parallel with improvements in the symptoms associated with MPA, including purpura and peripheral neuropathy. Therefore, the IP observed in this case cannot be attributed to a disease process other than MPA, and we considered it to be associated with the patient’s MPA.

Pneumomediastinum as a complication predictive of a poor prognosis is primarily observed in patients with dermatomyositis and only rarely develops in those with other connective tissue diseases (CTDs) (4). A few cases have been reported in which pneumomediastinum developed in patients with systemic sclerosis, systemic lupus erythematosus or rheumatoid arthritis (5-7). Pneumomediastinum occurs in 5-15% of patients with idiopathic pulmonary fibrosis, although it is not a fatal complication (8, 9). To our knowledge, this is the first reported case of MPA complicated by both IP and pneumomediastinum. In the present patient, the severity and prognosis of MPA were not influenced by the pneumomediastinum. Generally, rupture of alveoli or honeycombed cysts leading to pneumomediastinum occurs due to the vulnerability of alveolar connective tissue induced by interstitial inflammation, alveolar vasculitis and the administration of high-dose corticosteroids. Kono et al. suggested that the presence of mucosal necrosis in the airway due to vasculitis at the level of the bronchial arterioles causes pneumomediastinum in patients with dermatomyositis (10). Vasculitis has been confirmed in the bronchial and/or pulmonary arterioles in IP patients as a complication of

**Discussion**

The major complications of MPA in the lungs include alveolar hemorrhage, IP, bronchitis and organizing pneumonia (1). Alveolar hemorrhage and IP occur in 11% and 51% of patients, respectively, and are associated with a poor prognosis (1, 2). IP can present as an initial manifestation of MPA and may predate the development of symptoms associated with vasculitis by several years (2, 3). MPA-associated IP responds to steroids and immunosuppressants, such as methotrexate, cyclophosphamide and azathioprine, in contrast to idiopathic IP. In this case, the patient’s IP improved following the initiation of treatment with prednisolone and azathioprine in parallel with improvements in the symptoms associated with MPA, including purpura and peripheral neuropathy. Therefore, the IP observed in this case cannot be attributed to a disease process other than MPA, and we considered it to be associated with the patient’s MPA.
MPO-ANCA-associated vasculitis (2). We considered that the present patient’s pneumomediastinum possibly occurred following the rupture of vulnerable alveolar connective tissue induced by mucosal necrosis associated with vasculitis of MPA and the administration of high-dose corticosteroids.

To our knowledge, this is the first reported case of MPA complicated by IP and pneumomediastinum. The exact cause of pneumomediastinum as a complication of IP associated with CTD remains unclear. This case may help to clarify this pathological condition.

The authors state that they have no Conflict of Interest (COI).

References


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