Antineutrophil Cytoplasmic Antibody Associated Microscopic Polyangiitis and Sinobronchial Syndrome

**Key words:** MPO-ANCA, chronic sinobronchial syndrome, lung hemorrhage, chronic infection, small vessel vasculitis

Antineutrophil cytoplasmic antibodies (ANCA) are well known to be associated with small sized vessel vasculitic diseases such as microscopic polyangiitis (MPA), allergic granulomatous angiitis (AGA), and Wegener’s granulomatosis (WG) (1). One subtype is an antibody against myeloperoxidase (MPO), which stains in a perinuclear pattern (P-ANCA) by indirect immunofluorescence (IIF) using a neutrophil substrate, and the other subtype is an antibody against proteinase-3 (PR-3), which stains in a diffuse granular cytoplasmic pattern (C-ANCA) by IIF. PR-3 ANCA is more specific in WG than the other primary vasculitides. However, MPO-ANCA can be found in MPA and AGA more frequently than WG. The association of MPA with MPO-ANCA is reported to be in the range of 40-90%, and MPA with MPO-ANCA is reported to be frequently associated with necrotizing glomerulonephritis and/or pulmonary capillaritis (2). Pulmonary manifestations were observed in 40%, and renal manifestations were observed in 90% of the patients as the initial manifestations, showing that the organs mainly involved in MPA were the lungs and the kidneys. Pulmonary lesions are more frequently present in anti-PR-3 positive patients, whereas alveolar capillaritis with lung hemorrhage resulting from radiographically hemorrhagic capillaritis is found more frequently in anti-MPO-positive patients (2).

In this issue, an analysis is reported by Nagata et al (3) of a very rare MPA patient with a long history of chronic bronchitis and sinusitis (i.e. sinobronchial syndrome) who later evidenced long-standing consolidation in both lungs accompanied with areas of active inflammation.

See also p 544.

Two weeks after admission, MPO-ANCA was highly positive and pulmonary hemorrhage and hematuria occurred. Steroid therapy was administered; however, despite an initial positive response, the patient finally died of pneumonia. The systemic vasculitis was revealed through autopsy. Vasculitis may be caused by chronic inflammation in the lungs, associated with sinobronchial syndrome. Some reports have suggested the association of chronic airway infection with vasculitis, in which developed MPO-ANCA expression was associated with vasculitis (4). Recently, some reports have indicated a possible association of vasculitis via ANCA with some diseases which cause chronic airway infection; gram-negative bacilli are likely pathogen agents. In gram-negative bacilli infection, bacterial/permeability-increasing (BPI) protein existed in the azurophilic granules in neutrophils (5). ANCA against BPI was identified by Zhao et al (6); they suggested an association between BPI-ANCA and vasculitis. In the case of microscopic polyangiitis associated with sinobronchial syndrome, BPI ANCA was not measured; however it might have played some role in the pathogenesis (7). Further study is necessary on the characteristics of ANCA, closely related to the pathogenecity of chronic infection (e.g. sinobronchial syndrome) in the production of MPO-ANCA in MPA, as well as the pathophysiology of ANCA-associated vasculitis.

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References