中文題目：嗜中性白血球細胞質抗體血管炎合併急進性腎小球腎炎與尋常性間質性肺炎:案例報告

英文題目：Anti-neutrophil cytoplasmic autoantibody associated vasculitis associating rapidly progressive glomerulonephritis and usual interstitial pneumonia: A case report

作　　者：趙祐麟[1] 蔡哲嘉[1,2]

服務單位：高雄醫學大學附設中和紀念醫院[1]內科部 [2]腎臟內科

Introduction

Anti-neutrophil cytoplasmic autoantibody (ANCA)-associated vasculitis (AAV) is common with renal and respiratory involvement. We presented a case of anti-MPO antibody AAV with rapidly progressive glomerulonephritis and usual interstitial pneumonia (UIP).

Case report

This 50-year-old man presented with generalized weakness, progressive poor appetites, body weight loss (more than 10 percent of body weight in recent 2 month), and exertional dyspnea in recent one month. The results of a physical examination revealed bilateral fine crackle breathing sound and lower leg pitting edema. Laboratory tests were performed, and it revealed a serum creatinine level of 4.3 mg per deciliter; the creatinine level had been 1 mg per deciliter 1 months earlier. Urinalysis revealed 3+ occult blood and 2+ protein by dipstick. Because rapidly progressive glomerulonephritis was highly suspected, the laboratory tests survey and renal biopsy was arranged. The laboratory tests revealed anti-myeloperoxidase (anti-MPO) antibody more than 134 IU per milliliter. Histopathologic examination of biopsy specimen showed more than 50 percent of glomerulus show cellular crescents formation and the electron microscopy finding revealed no electron-dense depositions was seen in subepithelial or subendothelial region, consistent with pauci-immune cresentic glomerulonephritis. In addition, chest CT show honeycombing and traction bronchiectasis in the both lungs with peripheral and lower lobe predominance, suspected UIP. Under impression of ANCA-associated vasculitis with RPGN and UIP, the pulse therapy with methylprednisolone followed by daily oral prednisone and azathioprine for further treatment.

Discussion

ANCA-associated vasculitis (AAV) initially presents with constitutional symptoms lasting or weeks to months. The following evidences of specific involvement may appear. In this case, the patient presented of RPGN and positive result of anti-MPO antibody. According to the lung involvement, the most common clinical manifestation is diffuse alveolar hemorrhage, occurring in up to 29–36 percent of patients. However, several studies have

described the association of ILD and ANCA vasculitis, particularly those positive for MPO-ANCA, ranges between 4%–36% for MPO-ANCA and 2–4% for PR3-ANCA. In MPO-ANCA positive ILD, it was reported to be symmetrical in 50–100% of patients, affecting predominantly the lung periphery and lower areas and in cases without generalized involvement, UIP is also the most common abnormal pattern. It is consistent with the chest CT finding of this patient.

Conclusion

We encountered a patient who developed UIP and PRGN in the presence of MPO-ANCA. Though ILD is an uncommon complication of AAV, it is associated with poor prognosis and reduced survival. In addition, the ILD is more common in AAV with MPO-ANCA positive than PR3-ANCA ones. So, in the AAV with MPO-ANCA, ILD need to keep in mind.