中文題目：抗嗜中性球細胞質抗體合併抗腎絲球基底膜抗體造成快速進展的腎衰竭 ： 個案討論及文獻回顧

英文題目：Young Female Present With Rapid Progressive Kidney Injury – A Case Report

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Background: Concurrent positive anti-GBM and ANCA renal vasculitis is a rare clinical entity with un-clarified pathophysiological mechanism. Controversy existed regarding the response to immunosuppressive therapy survival prognosis.

Hospitalization Course:

We herein report a 23-year-old young lady, initially manifested as dizziness, paroxysmal nocturnal dyspnea and gross hematuria for 1 week. Laboratory investigation disclosed anemia and impaired renal function accompanied with increased infiltrate over bilateral lower lung fields. Otherwise, mild weight gain and mild elevated temperature were also noted.

Laboratory Results:

Numerous RBC were found over urine analysis though no obvious cast formation. Serology showed normal complement level, negative viral screening include HIV, HBV and HCV. Nevertheless, immunofluorescence assay detected positive perinuclear anti-neutrophil cytoplasmic antibodies (P-ANCA) with high titer, 1:640(+), and further confirmed as myeloperoxidase(MPO) ANCA with titer 134IU/ml (cut-off value, >5IU/ml) by fluorescence enzyme immunoassay (FEIA). In addition, antiglomerular basement membrane(anti-GBM) antibodies was positive at 27U/ml (cut-off value, >10U/ml). Further renal biopsy revealed extensive crescent glomerulonephritis with coexistence of various-staged crescents and prominent peri-glomerular inflammation. Immunofluorescence studies showed strong deposition of IgG in linear pattern along the basement membrane. Electron microscopy revealed no electron-dense deposition around glomerular capillary or mesangial area.

All together, the final diagnosis of crescentic glomerulonephritis with concurrent presence of anti-glomerular basement membrane antibodies (anti-GBM) and anti-neutrophil cytoplasmic antibodies (ANCA) was therefore made. Treatment was started with pulse methylprednisolone and cyclophosphamide. Double filtration plasmapheresis was performed daily for 11 sessions using 60ml/kg per exchange. Mycophenolate mofetil 1gm daily was substituted as further induction and maintenance immunosuppressive therapy instead of cyclophosphamide in concideration of possible fertility toxicity. She returned ordinary activity with modest renal insuffiency.

Discussion:

Delayed diagnosis and under-treatment contribute to ominous outcome in the past time. Our patient achieved rapid and sustained remission despite convincing data for MMF as induction and maintenance agent still lack. Further large scale observation study would be needed to confirm non-inferior prognosis of double positive glomerulonephritis and role of Mycophenolate.