中文題目：一名 Goodpasture's disease 病患早期診斷與治療的挑戰

英文題目： The challenges of early diagnosis and treatment in a patient with Goodpasture's disease

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Background: Anti-Glomerular Basement Membrane Disease (Anti-GBM disease) is a rare and life-threatening autoimmune disorder (0.6 per million/per year in Asian population). It targets the alpha 3 chain of the type IV collagen of glomerular and alveolar basement membranes. It presents with rapid progressive crescentic glomerulonephritis, and pulmonary hemorrhage in over half the patients, known as "Goodpasture's disease".

Case presentation: A 67 years old woman presented with decreased urine output and progressive bilateral legs edema over 4 days. She had underlying hypertension but otherwise had normal renal functions. Prior to this she had a common cold but denied taking NSAIDS or other medications. Laboratory workup showed acute kidney injury (Creatinine: 11.8 mg/dL) and hyperkalemia (K+ 5.9 mmol/L).

Due to persistently anuric status and hyperkalemia (K 6.7 mmol/L) refractory to medical treatment, dialysis therapy was initiated on the day 1of hospital admission. Workup for rapidly progressive renal failure including serology, ultrasound and then renal biopsy were sequentially performed.

Pathology showed diffuse (100%) crescent formations and immunofluorescence stain showed linear deposition of IgG (2+) and C3 (1+) along glomerular capillary walls. Serology workup for ANA, C3, C4, hepatic viral markers, P-ANCA, C-ANCA were within normal limits, whilst high titers of anti-GBM antibody (314 EliA U/mL) was found. We promptly initiated daily plasmapheresis therapy along with cyclophosphamide and prednisolone. Adjusted dose was prescribed for the concern of the infection due to uncertain presentation of pulmonary hemorrhage in chest X ray.

Initially, she had no respiratory distress or cough. However, respiratory distress and desaturation occurred after the second day of plasmapheresis therapy. Hemoptysis with progressive anemia ensued and O2 non-rebreathing mask support was required. Chest CT confirmed pulmonary hemorrhage. After 10 cycles of plasmapheresis her alveolar hemorrhage and respiratory distress subsided and titers of anti-GBM antibody were significantly decreased (314 $\rightarrow$ 24 EliA U/mL). However, the patient still required maintenance hemodialysis.

Conclusion: Goodpasture's disease can present as a life-threatening pulmonary-renal disorder that requires rapid diagnosis and intervention. Therapeutic regimen of plasmapheresis, corticosteroids and cyclophosphamide can improve patient survival, treat pulmonary hemorrhage and may improve renal outcome.