中文題目：抗基底膜疾病: 罕見病例報告

英文題目：Anti-glomerular basement membrane disease: a rare case report

作　　者：彭梓晏[1], 彭渝森[1],

服務單位：[1]亞東紀念醫院內科部

***Case report**:*

Anti-glomerular basement membrane (anti-GBM) disease is a rare disease involving small vessels affecting kidneys and lungs. It is an archetypic autoimmune disease but rare in Taiwan according to previous investigation. [1] We here presented a typical case of this disease.

A 68 year old Taiwanese female, having history of coronary artery disease under medical control, hypertension and left femoral neck avascular necrosis status post total hip replacement, found to have little urine output and lower limb edema for 3 days at her our cardiac out-patient department follow-up. The laboratory result disclosed leukocytosis(WBC 20.84x103/uL), Normocytic anemia (Hb 9.0g/dL, MCV 84.1fL), hyperkalemia(5.9mmol/L), much elevated creatinine(11.80mg/dL) and high D-dimer level(>10000ng/ml FEU). VBG demonstrated metabolic acidosis with partial respiratory compensation(pH 7.291, pCO2 30.5 mmHg, HCO3 14.4 mmol/L). She was immediately referred to emergency department. Ceftriaxone was added empirically. Although delivered urgent potassium-lowering medications, the emergency staff noted the persisted hyperkalemia(5.6 mmol/L). Emergent and maintenance hemodialysis was started the next day. Renal ultrasonography found bilateral parenchymal renal disease and left renal cyst only. After stabilizing the patient, we arranged renal biopsy and checked autoimmune antibodies. The biopsy was performed 3 days after admission. The pathologist reported likely crescentic glomerulonephritis owing to anti-GBM disease. We then did plasmapheresis 5 times, double filtration plasmapheresis (DFPP) 5 times, and added prednisolone plus cyclophosphamide treatment. The patient then got better as the decreasing titre of the Anti-GBM. We kept cyclophosphamide for 4 weeks. Though renal function not recovered, there was residual urine output on discharge. The patient was discharged under improved condition after being treated in hospital for 40 days.

***Discussion**:*

This is a rare case of typical anti-GBM disease, also called Goodpasture syndrome, as named after the American pathologist Ernest Goodpasture describing the syndrome in his 1919 paper. Anti-GBM disease is a rare but fatal disease, usually presenting as rapidly progressive crescentic glomerulonephritis. The prevalence was estimated to be less than one case per million.[2] This is also true in the investigation from Taiwan Society of Nephrology[1]. Currently, standard treatment for anti-GBM disease include plasmapheresis, which able to remove the pathogenic autoantibody, as well as cyclophosphamide and corticosteroids which are inhibiting further autoantibody production and decreasing organ inflammation. The use of the combination is now still the recommendation of the latest Kidney Disease Improving Global Outcomes guideline for treating anti-GBM GN[3]. The plasmapheresis usage is supported by observational studies, which suggests

improved renal outcome and patient survival when compared to historical cohort treated by immunosuppression alone. [4] Our case demonstrated the feasibility of the treatment.

References

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