

中文題目：Amyloidosis 患者合併頑固性蛋白尿之個案報告

英文題目：Amyloidosis with refractory heavy proteinuria

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Introduction: Amyloidosis is a rare disease caused by extracellular deposition of pathological insoluble fibrillary protein in organs or tissues and may result in organ dysfunction. Most patients with amyloidosis will require biopsy of an involved organ or tissue to confirm the diagnosis. One third of the patients with amyloidosis have nephrotic syndrome.

Case presentation: This 47 year-old patient presented with refractory foamy urine and extremities pitting edema for half a year. Laboratory examination reported hypoalbuminemia (2.5 g/dL) and hypercholesterolemia (Cholesterol 740 mg/dL, LDL 563 mg/dL), hypertriglyceridemia (2903mg/dL). Serum/urine IFE both showed no monoclonal band. Elevation of lambda serum free light chain was observed. Urine analysis revealed no hematuria but heavy proteinuria (UPCR 34021.11 mg/g). Renal ultrasound revealed relatively increased size and increased cortical echogenicity of bilateral kidneys. She denied past history and her ADL was totally independent. Ultrasound-guided percutaneous renal biopsy was performed smoothly on 2022/2/10.

Special stain for congo red was positive and the pathology reported lambda light chain amyloidosis. The PYP heart scan showed equivocal suggestion of transthyretin amyloidosis (ATTR). Lambda light chain amyloidosis with renal involvement was diagnosed. We started treatment with steroid as Prednisolone 10mg QD (0.15mg/kg/day) and Endoxan 100mg QW. However, renal function deteriorated gradually with heavy proteinuria persisted. Here, we report a 47-year-old female with AL amyloidosis which is refractory to treatment.

Discussion: Refractory heavy proteinuria and dyslipidemia persisted despite optimal medical treatment. A few case reports mentioned of the role of plasmapheresis in patients with paraproteinemia. NHI double filtration plasmapheresis QW was applied for the indication of dyslipidemia. We regular followed-up the effect of plasmapheresis on dyslipidemia and paraproteinemia.

Conclusion: After double filtration plasmapheresis QW for one month, dyslipidemia significant improved. The patient's creatinine and UPCR mild improved. We will keep closely monitoring this patient's condition.