中文題目：紫斑症併發急性腎衰竭：案例報告

英文題目：Acute kidney injury in a Patient with purpuric rash

作　　者：盧律衡[1]，連培因[2]，郭美娟[3]

服務單位：[1]高雄醫學大學附設醫院內科部，[2]高雄醫學大學附設醫院病理科，[3]高雄醫學大學附設醫院腎臟內科

Introduction:

The production of cryoglobulins is often the consequence of an underlying disorder that needs further evaluation. We reported a case of a 65 year-old-female with purpuric rash over bilateral legs accompanied with active deterioration of renal function.

Case Report:

A-65-year-old female had past medical history for chronic kidney disease(baseline serum creatinine of 1.6 mg/dl), hypertension and liver cirrhosis with portal vein hypertension. She was presented to our emergency room due to progressive bilateral lower legs edema for 6 months. Dyspnea and purpuric papules over legs(Figure 1) were also noted.

Initial laboratory test showed serum creatinine level of 5.01 mg/dl, microscopic hematuria and heavy proteinuria(24 hours urine protein of 1.3 g). She developed progressive serum creatinine level to 10.84 mg/dl with uremic symptoms and pleural effusions in days. Skin biopsy revealed leukocytoclastic vasculitis with C3 complement deposition. Additional laboratory data demonstrated positive serum cryoglobulin, elevated serum anti-dsDNA level of 44 IU/ml, elevated serum antiphospholipid level of 56.5 unit, low serum C3 level of 78.3 mg/dl but normal serum C4 level of 21.4 mg/dl. Serum anti-nuclear antibody, rheumatoid factor, antiphospholipid antibodies, anti-glomerular basement membrane, anti-neutrophil cytoplasmic autoantibody, hepatitis C antibody and hepatitis B surface antigen were negative. Kidney biopsy revealed type I membranoproliferative glomerulonephritis(MPGN) with immune complex deposits. Serum immunofixation showed no monoclonal gammanopathy.

Pulse therapy with 500mg Methylprednisolone for 3 days was performed, followed by low dose Cyclophosphamide 25mg per day. Follow-up renal function and proteinuria improved. Cryoglobulins also normalized.

Conclusion:

Cryoglobulinemia may be asymptomatic and may be detectable on healthy persons. Mixed cryoglobulinemia syndrome (MCS) refers to type II or type III cryoglobulinemia, which can present as a systemic vasculitis. Manifestations range

from purpura, arthralgia, and mild neuropathy to more severe neurologic and kidney involvement. Chronic Hepatitis C Virus infection leading cause of MCS and autoimmune disease only accounts for 10 percent of these cases .

About 20% of patients with MCS present with nephropathy at diagnosis (1). MPGN is the commonest type in Cryo GN. The rest are other types such as endocapillary proliferative GN, mesangial proliferative GN, and rare crescentic GN.

In our case, kidney biopsy didn't reveal rapidly progressive crescentic pattern. Neither immunofluorescence microscopy nor electron microscopy revealed specific pattern of MCS such as IgM deposition or fingerprint pattern. The diagnosis of systemic lupus erythematosus was questionable if using 2019 EULAR/ACR criteria. The mechanism of MCS may be related to undefined autoimmune disease which further induced cutaneous vasculitis and type I MPGN.

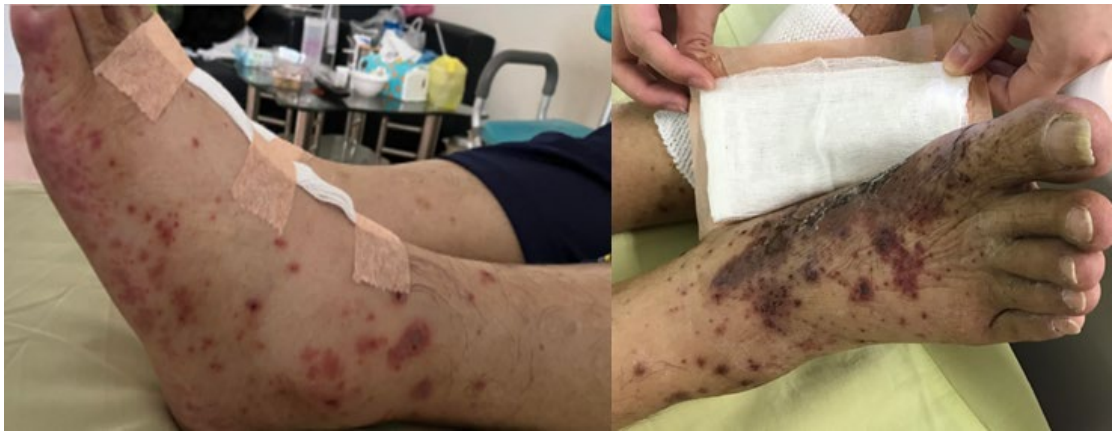

Figure 1. Purpuric rash over bilateral feet at admission