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Podocyte Infolding Glomerulopathy As The First Renal Manifestation In Systemic Lupus Erythematosus

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Objectives : The clinical and pathologic manifestations of lupus-associated kidney disease are diverse, with features of immune complex glomerulonephritis (GN) being the most common. Rarer entities have been recently described. We report a case of podocyte infolding glomerulopathy (PIG) as the first kidney manifestation in a south-east Asian patient with systemic lupus erythematosus (SLE).
Methods : -
Results : A 48-year-old Chinese female was diagnosed with SLE after presenting with serositis and alopecia, which was associated with low C3, positive anti-ANA, anti-dsDNA, anti-Ro antibodies and sub-nephrotic to nephrotic range proteinuria (peak uPCR 3.12g/g; serum albumin 39g/L). There was no hematuria. Serum creatinine was 71µmol/L (eGFR 87ml/min/1.73m^2). Hepatitis virologies were negative. Kidney biopsy performed for persistent proteinuria showed focal, mild mesangial and endocapillary hypercellularity, with trace granular mesangial / paramesangial staining for immunoglobulins, C3, C1q and C4 on immunofluorescence. Interestingly, ultrastructural study demonstrated lack of electron dense deposits, but there was infolding of podocyte cytoplasmic projections, forming microspheres or microtubules. Findings were most consistent with PIG, without evidence of conventional lupus nephritis subclasses. Rapid improvement of proteinuria was observed with prednisolone (PRL) 40mg OD and mycophenolate mofetil (MMF) 2500mg OD. Patient remained in full remission at 19 months of follow-up (PRL 3mg OD; MMF 1000mg OD).
Conclusions : The occurrence of PIG lesions in SLE patients is increasingly reported. Our case, together with prior reports, expands the spectrum of renal lesions in SLE. The pathomechanism of PIG is poorly understood, although podocyte cytoskeleton dysregulation has been implicated. Remission after immunosuppression (IS) in our patient suggests the beneficial role of IS, at least in a subset of patients, although the optimal regimen is presently unclear. Remission with supportive therapy alone has also been described. Given its rarity, pooled review of literature is necessary to improve our understanding of this entity.