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What’s the diagnosis?

A 53-year-old man presented with progressive lower extremity edema and acute kidney injury with serum creatinine 9.6 mg/dl (baseline 2.5 mg/dl). Urinalysis/urine sediment was strongly positive for proteinuria (3+) and mild hematuria without dysmorphic RBCs or RBCs casts. Twenty-four-hour urine protein was 7.1 g. Kidney ultrasound was normal. AntiPLA2R antibody assay was strongly positive with a titer of 1500 relative units/ml. Complement levels were normal. All other serologies were negative with no monoclonal gammapathy and negative malignancy workup. Kidney biopsy demonstrated evidence of membranous nephropathy along with crescents. The patient was diagnosed with primary crescentic membranous nephropathy. He was started on lisinopril along with prednisone monotherapy. Patient declined using any other immunosuppression agents. The patient’s serum creatinine returned to baseline and protein/creatinine ratio improved to 2.4 g/24 hr within four months.

Light microscopy showed mesangial matrix expansion, thickened of the capillary walls, and active cellular crescents (Figures 1 and 2). Electron microscopy demonstrated diffuse effacement of the podocyte foot processes involving 80—90% of the capillary surfaces with subepithelial deposits (Figure 2).

Primary crescentic membranous nephropathy with negative serology is a rare disease and was reported in only a handful of prior case report/series. Patients usually present with nephrotic range proteinuria, hematuria and acute decline in kidney function. Kidney biopsy confirmsthe diagnosis. Currently there are no guidelines regarding the ideal treatment in these cases. Unlike typical membranous nephropathy, crescentic membranous nephropathy usually progresses long-term, even if patients experience initial positive response to therapy.

(Images and text provided by Mohamad Hanouneh, MD, Steven Menez, and Duvuru Geetha, MBBS, MD, Johns Hopkins University, Department of Medicine, Baltimore, Maryland)

Reference