Is the Proteinuria from Lupus or a Solitary Kidney?

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Abstract

Podocyte infolding glomerulopathy (PIG) is a primary podocytopathy characterized by the infolding of podocytes into the glomerular basement membranes leading to the formation of microspheres and microtubules. Podocyte infolding glommerulopathy is frequently associated with autoimmune disease, most commonly lupus and mixed connective tissue disorder. The typical presentation of the patient is with proteinuria, hematuria, with or without signs of systemic autoimmune disease. The etiology of PIG is currently unknown, but recent studies have shown a decrease in the protein ACTN4 which serves as an actin crosslinking cytoskeletal protein for maintaining normal podocyte structure. This entity usually responds favorably to steroids.

Case Presentation: LH is a 52 year-old female with a past medical history significant for CKD Stage 3 (solitary kidney S/P right nephrectomy 1993) who presented with nephrotic syndrome, hydronephrosis (with nephrostomy tube placement), and a creatinine of 1.85. She has lupus which has been treated with Plaquenil.

Renal Biopsy Findings: The biopsy showed glomerulomegaly, secondary focal segmental glomerulosclerosis, and moderate interstitial fibrosis consistent with the patient's history of a solitary kidney. Immunofluorescence microscopy was negative within the glomeruli by both routine and protease digested paraffin-embedded tissue. Electron microscopy showed extensive infolding of cytoplasmic processes into the glomerular basement membranes. Membrane bound microspherules and microtubules were readily identified. Complete podocyte foot process effacement was noted.

Clinical Follow-up: LH has only received a few doses of steroids + rituximab. She has been admitted to the hospital several times for nephrostomy tube issues. She is currently on dialysis and the rituximab and steroid treatment is resuming as an outpatient.