

CLINICAL IMAGE

Uncommon things to note about a common cause of nephrotic syndrome

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Tel: 352-294-8694
Email: akoratsla@ufl.edu**Key Clinical Message**

Primary membranous nephropathy is typically a disease of middle-aged white men but should be included in the differential diagnosis of nephrotic syndrome in patients of any age and race. Serological anti-PLA2R testing must be interpreted in the appropriate clinical context and histological PLA2R staining is recommended in seronegative patients.

KEYWORDS

biopsy, membranous nephropathy, PLA2R, THSD7A

1 | CASE

Primary membranous nephropathy (PMN) is the most common cause of idiopathic nephrotic syndrome in nondiabetic white adults. The mean age of incidence is 50-60 years and is most common in whites followed by Asians, Blacks, and Hispanics.¹ Most cases (70%-82%) of PMN have circulating IgG4 autoantibody to the podocyte membrane antigen, M-type phospholipase A2 receptor (PLA2R), and a minority have thrombospondin type 1 domain-containing 7A (THSD7A) antibodies. It is noteworthy that ~15% cases have biopsy evidence of PLA2R staining despite negative serum levels.^{1,2} We present a case of young Hispanic male with biopsy-proven PMN with positive PLA2R staining but negative circulating antibodies.

A 34-year-old Hispanic man presented with worsening generalized edema for 2 weeks. Laboratory data were consistent with nephrotic syndrome with a urine protein-creatinine ratio 11 g/g (Ref: <150 mg/g), LDL-cholesterol

279 mg/dL (<100) and serum albumin 1.1 g/dL (3.5-5). Work-up for HIV, viral Hepatitis, syphilis, and common vasculitides was negative. PLA2R and THSD7A serology were negative. Renal biopsy was consistent with PMN with a positive glomerular staining for anti-PLA2R (Figure 1). He was started on Losartan, statin, and diuretics and instructed to follow up with nephrology in a few months to determine the necessity for immunosuppressive therapy.

INFORMED CONSENT

Informed consent has been obtained for the publication of this clinical image.

AUTHORSHIP

All the authors have made substantial contribution to the preparation of this manuscript. DE: drafted the initial version of manuscript, attending nephrologist on the case; MA:

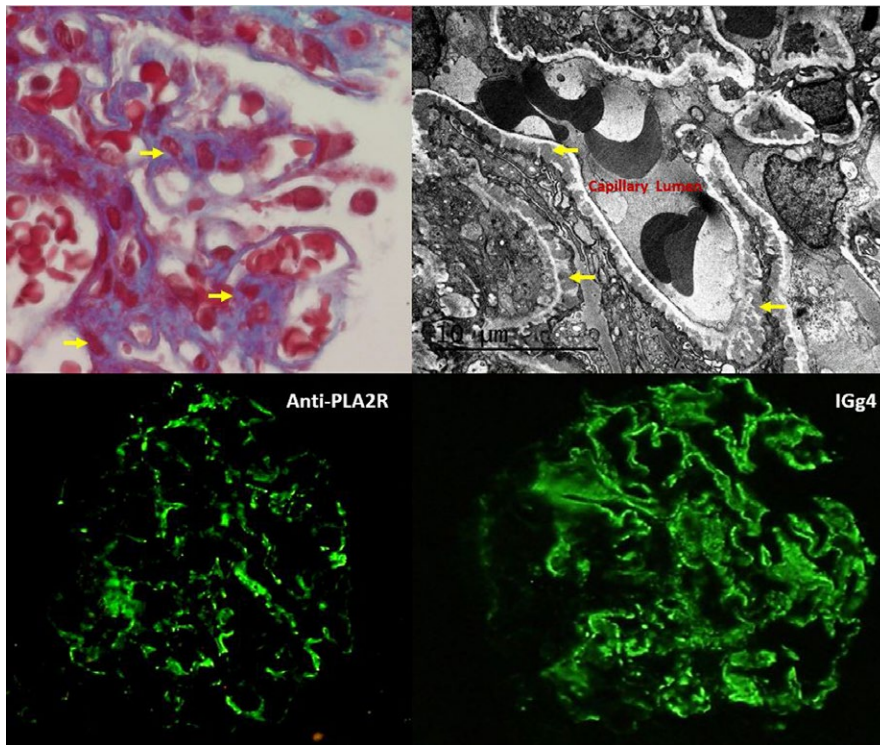


FIGURE 1 Kidney biopsy findings diagnostic of primary membranous nephropathy: [Upper Left Panel] Light microscopy with Masson's trichrome stain demonstrating fuscinophilic, red-orange subepithelial and mesangial deposits (arrows) [Upper Right Panel] Electron microscopy demonstrating subepithelial deposits [Lower Panels] Immunofluorescence microscopy showing positive staining for PLA2R and IgG4, respectively

provided pathology images and pertinent input, attending pathologist on the case; AK: Senior nephrologist who reviewed and revised the manuscript for critically important intellectual content.

CONFLICT OF INTEREST

The authors have declared that no conflict of interest exists.

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