A 43-year-old female was admitted to the Department of Nephrology at Jinling Hospital (Nanjing, China) in January 2017 complaining of edema for 3 months with urine abnormalities. Her father had renal disease (with no biopsy performed) when he was 40 years old and died of uremia at 56 years old. Her mother and brother were healthy; however, her daughter and nephew (her brother’s son) had slightly high microalbumin levels in routine urine screenings. Her daughter’s urinary protein level was weakly positive, whereas her nephew’s urinary protein level was negative. At admission, the patient had a normal mental status. Her height, weight, body temperature, pulse, and respiratory rate were 154 cm, 49.4 kg, 36.2°C, 95 beats/min, and 15 breaths/min, respectively, and her blood pressure was 127/96 mmHg. Her daily urinary protein, serum albumin, blood urea nitrogen, creatinine, and estimated glomerular filtration rate levels were 2.67 g/day, 40.4 g/L, 12.6 mg/dl, 0.61 mg/dl, and 111.38 ml·min⁻¹·1.73 m⁻², respectively. The following blood tests were also performed: liver function tests; blood lipid levels; immunoglobulin (Ig) G (IgG), IgM, IgA, antistreptolysin O, rheumatoid factor, complement C3 and complement C4 levels; testing for the presence of antinuclear antibodies (ANAs), anti-double stranded-DNA antibodies, anti-phospholipase A2 receptor antibodies, c-anti-neutrophil cytoplasmic antibodies, and p-anti-neutrophil cytoplasmic antibodies; the ratio of blood monospecific free light chain; immunosassay electrophoresis; and hepatitis B and C serology. A renal ultrasound indicated that the kidney size was normal. An ANA spectrum indicated RO-52++, and other tests such as for anti-SSA antibody and anti-SSB antibody were negative.

The use of a renal biopsy light microscope indicated one abandoned glomerulus out of the 37 glomeruli obtained from the kidney biopsy samples. The findings indicated that the glomerular volume had increased, and all glomeruli exhibited moderate-to-severe mesangial cell proliferation, mesangial hyperplasia, segmental mesangiolysis, and a modular formation. The capillary loops opened well, and large periodic acid-Schiff-positive (PAS) materials were distributed in the subcutaneous glomeruli. Numerous PAS and Masson Trichrome-positive deposits were identified in the subendothelial and mesangial areas. The case of tubular disease appeared to be mild, tubulointerstitial was scattered in the mononuclear cell infiltration, medullary interstitial fibrosis was identified, and the smooth muscle of the small arterial showed vascular degeneration. Immunofluorescence microscopy testing (for IgG, IgA, IgM, C3, C1q, κ, and λ) indicated completely negative results. Antifibronectin immunostaining demonstrated that the mesangial deposits were positive for fibronectin [Figure 1]. Fibrillar high electron density deposits (diameter of approximately 10 nm at a higher magnification) were identified in the glomerular subendothelial and mesangial areas.

Fibronectin glomerulopathy is an autosomal dominant genetic disease. In 1995, Assmann et al. showed the presence of extensive deposits in the mesangium and subendothelial space with strong immune reactivities to fibronectin. They named the disease “familial glomerulonephritis with
Fibronectin deposits. Several case reports subsequently appeared in the literature. The disease can manifest at any age, with reports of cases that range from 3 to 88 years. The clinical features of the disease include proteinuria, microscopic hematuria, hypertension, a decreased glomerular filtration rate, and Type IV renal tubular acidosis. Most patients progress to end-stage renal disease (ESRD) in the second to sixth decades of life. Familial morbidity is another feature. The patient in our case had a family history of renal disease, as her father had renal disease, and her daughter had slightly high microalbumin levels.

Fibronectin glomerulopathy was established on the basis of the kidney biopsy findings. Using light microscopy, the volume of the glomerulus was increased, and substantial amounts of PAS and fuchsinophilic materials were present in the mesangial area. The Congo-red and silver staining results were negative. IgG, IgM, C1q, C3, and C4 deposition and complement staining were typically negative or faint. These findings demonstrate the strong immunoreactivity of fibronectin. Electron microscopy indicated focal fibrils or fibrillar subendothelial deposits with 12–16 nm fibrils, and immunoelectron microscopy confirmed that colloidal gold particles marked fibronectin assembled in the electron-dense deposits in the mesangial matrix and along the glomerular capillary loops.

To date, the genetic background of fibronectin glomerulopathy has been unclear. In 2008, Castelletti et al. first reported a link between fibronectin glomerulopathy at the fibronectin 1 gene (FN1) locus at 2q34 and three heterozygous missense mutations within the heparin-binding domains of FN1 (p.Tyr973Cys, p.Trp1925Arg, and p.Leu1974Arg). In 2016, Ohtsubo et al. identified six FN1 mutations and first reported five mutations (p.Pro969Leu, p.Pro1472del, p.Trp1925Cys, p.Lys1953_Ile1961del, and p.Leu1974Pro) within the integrin-binding domain. They determined that p.Tyr973Cys, p.Pro1472del, and p.Leu1974Pro mutations exist in multiple families, and their founder mutations include p.Pro1472del and p.Leu1974Pro. Regrettably, the patient in our case did not agree to genetic testing.

Currently, there is no known effective and specific treatment. Most researchers believe that fibronectin glomerulopathy is not an immune disease; thus, treatment with hormone and immunosuppressive therapies is inappropriate. Basic therapy includes blood pressure control and the administration of angiotensin-converting enzyme inhibitors or angiotensin II receptor blockers. Patients can undergo hemodialysis or peritoneal dialysis as they progress to ESRD.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity; however, anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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