DNAJB9-Associated Fibrillary Glomerulonephritis and Immunoglobulin A Nephropathy – A Rare Combination

Dear Editor,

A 67-year-old gentleman presented to us for evaluation of incidentally detected renal dysfunction. He had systemic hypertension for 10 years and was recently detected to have diabetes. The blood pressure was 150/90 mm of Hg and had no evidence of target organ damage related to diabetes or hypertension.

Urinalysis showed 2+ albuminuria with 10–16 red blood corpuscles/high-power field. The urine spot protein–creatinine ratio was 3.1 and serum creatinine was 1.6 mg/dl. C3 and C4 levels were normal. Serological testing for hepatitis B surface antigen, hepatitis C antibody, antinuclear antibody, antineutrophilic cytoplasmic antibodies, anti-glomerular basement membrane antibodies, serum cryoglobulins, and human immunodeficiency virus antibody were all negative. Serum protein electrophoresis with immunofixation was negative for monoclonal protein.

Renal biopsy showed histopathologic findings consistent with fibrillary glomerulonephritis and immunoglobulin A nephropathy dual glomerulopathy (in light, immunofluorescence, and electron microscopy), and all glomeruli stained positive for DNA-J heat-shock protein family member B9 (DNAJB9) [Figure 1].

His renin-angiotensin-aldosterone system blockade was enhanced to 80 mg daily of telmisartan. At 6 months follow-up, he is asymptomatic, and his blood pressure is well controlled with a serum creatinine of 1.4 mg/dl and urine spot protein–creatinine ratio of 1.6. He is advised to remain on close follow-up.

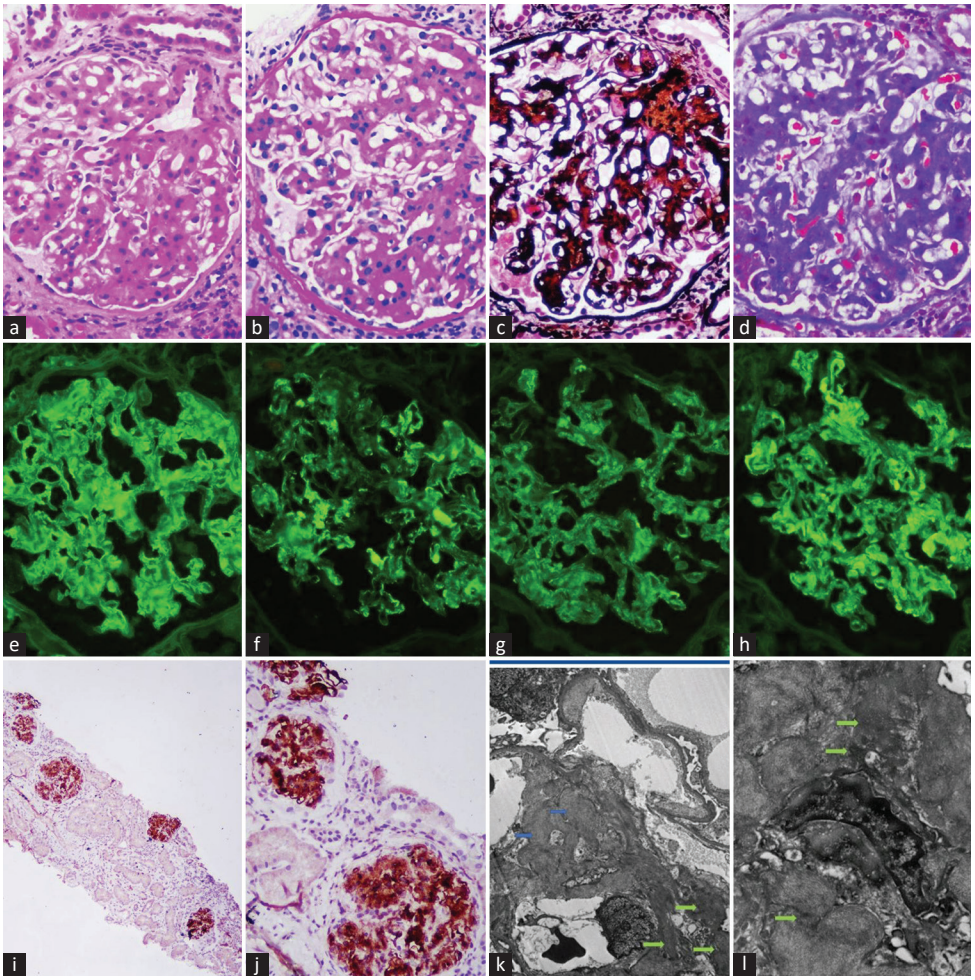

Figure 1: Renal biopsy showing mesangioproliferative pattern of glomerular injury on light microscopy (a–d). Immunofluorescence microscopy showing smudge positivity of IgG along the capillary loops with intensely positive IgA and C3 over mesangium (×400) (e–h). Glomeruli were intensely positive for DNAJB9 immunohistochemical stain (×600) (i and j). Electron microscopy confirmed nonbranching linear fibrils on capillary loops and mesangium, along with electron-dense deposits consistent (arrows) with IgA over mesangium (k and l) DNAJB9 = DNA-J heat-shock protein family member B9.

Coexistent FGN–IgAN glomerulopathy is an extremely rare form of glomerular disease.[1,2] FGN–IgAN has a similar clinical presentation as FGN and is associated with infections (hepatitis C), autoimmune diseases, or malignancy in 30% of cases.[3] In our patient, workup for secondary causes of FGN was negative. The discovery of DNAJB9 and immunohistochemical staining for DNAJB9 now make it possible to diagnose FGN even in the absence of ultrastructural evaluation.[3] Our report highlights a patient with idiopathic FGN–IgAN dual glomerulopathy, an extremely rare glomerular disease. To our knowledge, this condition has not been reported before from the Indian subcontinent.

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Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent.

Conflicts of interest
There are no conflicts of interest.

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Scrub Typhus in a Kidney Transplant Patient

Dear Editor,

Scrub typhus is a vector-borne disease transmitted by the bite of a trombiculid mite and is caused by the bacterium *Orientia tsutsugamushi*.[1] Although it is a common cause of fever in the Indian subcontinent, there is little information about scrub typhus in transplant recipients. To date, only one other case of scrub typhus in a kidney transplant recipient has been reported.[2]

We treated a 60-year-old male kidney transplant recipient who presented with a 15-day history of fever along with headache, dry cough, myalgia, and a rise in serum creatinine from 2 mg/dL to 4 mg/dL. Common possible causes of acute febrile illness, including, imaging with CT scan, cytomegalovirus serology, malaria, dengue, and enteric fever, were ruled out, and a test for scrub typhus IgM ELISA was positive with a value of 2.13 (normal < 0.9). He was started on tablet doxycycline 100 mg twice daily. His fever subsided within 24 h, and graft kidney function improved.

This case report highlights the importance of considering scrub typhus in the differential diagnosis of fever of unknown origin in kidney transplant recipients, especially in the Indian subcontinent. Typical painless eschar may be seen at the site of a bite in 40%–50% of patients; hence, the absence of eschar does not rule out this infection.[3] Diagnosis can be confirmed by scrub typhus IgM ELISA, but this may be absent in the early phase of the disease. Polymerase chain reaction can be done, with the sensitivity ranging from 80% to 90%.[4]

Clinicians should be aware of the clinical presentation of scrub typhus and should have a high index of suspicion in kidney transplant recipients with fever of unknown origin. Early diagnosis and treatment with doxycycline is essential to prevent severe complications. Azithromycin is an alternative agent that can be used.

Declaration of patient consent
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