Acute kidney injury in a patient with systemic sclerosis: Looking beyond scleroderma renal crisis

Sir,

A 32-year-old female patient of diffuse cutaneous systemic sclerosis (SSc) for 9 years, presented with effort intolerance, and progressive fatigue for 2 months without any history of pedal swelling, orthopnea, paroxysmal nocturnal dyspnea, any worsening of baseline shortness of breath, puffiness of face or oliguria or fever. Initial clinical examination was unremarkable except for mild pallor and tachycardia (pulse rate 110/min). Blood pressure was 100/70 mm of Hg. Laboratory examination showed hemoglobin 9.2 g/dl, total leukocyte count 4500/μL, platelets 170,000/μL, normal liver function tests and lipid profile, creatinine was 2.2 mg/dl (previous creatinine 3 months ago was 0.8 mg/dl) and urine routine examination showed microscopic hematuria and pyuria. Urine culture and Z/N stain, tuberculosis (TB) polymerase chain reaction and BACTEC TB culture were negative. Ultrasound examination of both kidneys showed normal size and echotexture. Normotensive scleroderma renal crisis (SRC) was the presumptive diagnosis, and peripheral blood smear was ordered, which did not show any schistocyte and her reticulocyte count was also normal. Immune serology showed positive antinuclear antibody and Scl70, normal complements and negative lupus anticoagulant, antineutrophil cytoplasmic antibodies (ANCA) positive (3+, p – ANCA pattern, Euroimmune), anti-myeloperoxidase (MPO) strongly positive – 195 (normal <20, enzyme-linked immunosorbent assay). Renal biopsy [Figure 1] showed: cellular crescents in 6/10 glomeruli, increase in mesangial matrix and cellularity in all glomeruli, hyaline, granular and red blood cell casts in tubules along with endothelial prominence, focal fibrinoid degeneration, and vasculitis. Immune staining was negative for IgM, IgA, IgG, C3 and C1q. In view of no other systemic manifestations, especially otorhinolaryngological and pulmonary manifestations, presence of pauci-immune glomerulonephritis in presence of MPO – ANCA positivity suggested isolated renal ANCA – associated vasculitis (AAV) in this patient with diffuse cutaneous scleroderma. She was treated with pulse cyclophosphamide according to CYCLOPS protocol with gradual improvement.

ANCA- associated vasculitis is a rare complications of SSc, occurring in 0.4% cases. Majority of such cases occur

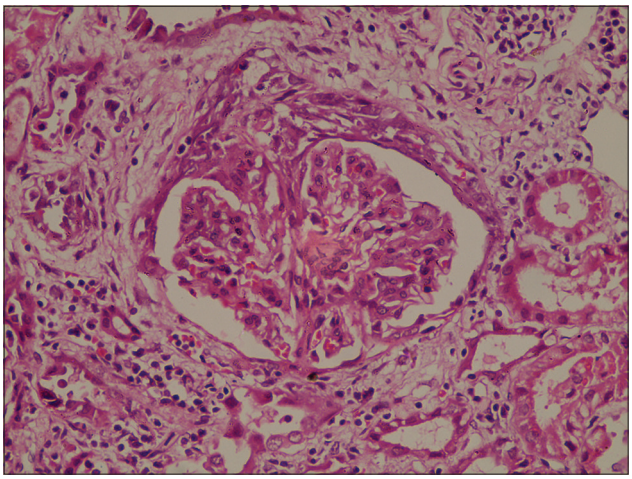

Figure 1: Renal biopsy (light microscopy): Showing cellular crescents, increase in mesangial matrix and cellularity

in the limited rather than the diffuse cutaneous group, as opposed to our patient. However, anti-MPO ANCA is the most common subtype and clinically granulomatous disease is rare, and the majority of patients present with glomerulonephritis, renal arteritis or pulmonary fibrosis.[1] Yamashita *et al.* reported a case of AAV in 72-year-old lady with diffuse cutaneous SSc with cellular crescent on renal biopsy and mononeuritis multiple.[2] In another study with 12 patients of systemic vasculitis and associated SSc, AAV was found in nine patients. Renal limited vasculitis was found in only two out of these nine patients. Interestingly, unlike our patient, all patient of AAV in this series had limited type of SSc.[3] AAV can rarely be associated with SSc and may pose a diagnostic challenge to the physicians. In a patient of SSc with acutely worsening renal function AAV should be considered as a differential apart from SRC as the treatment protocol and outcome are different for these two conditions.

R. P. Goswami, S. Mondal, K. Basu, S. Das, P. Ghosh, A. Ghosh

Department of Rheumatology, Institute of Post Graduate Medical Education and Research, Kolkata, West Bengal, India

Address for correspondence:
Dr. Sumantro Mondal,
Department of Rheumatology, Doctor's Hostel 242, A. J. C. Bose Road, Kolkata - 700 020, West Bengal, India.
E-mail: drmsumantro@gmail.com

References

1. Derrett-Smith EC, Nihtyanova SI, Harvey J, Salama AD, Denton CP. Revisiting ANCA-associated vasculitis in systemic sclerosis: Clinical, serological and immunogenetic factors. Rheumatology (Oxford) 2013;52:1824-31.
2. Yamashita H, Takahashi Y, Kaneko H, Kano T, Mimori A. A patient with diffuse cutaneous systemic sclerosis complicated by antineutrophil-cytoplasmic antibody-associated vasculitis exhibiting honeycomb lung without volume loss. Intern Med 2014;53:801-4.
3. Quéméneur T, Mouthon L, Cacoub P, Meyer O, Michon-Pasturel U, Vanhille P, *et al.* Systemic vasculitis during the course of systemic sclerosis: Report of 12 cases and review of the literature. Medicine (Baltimore) 2013;92:1-9.

Access this article online	
Quick Response Code:	Website: www.indianjnephrol.org
	DOI: 10.4103/0971-4065.157005