Glomerular Syndrome in Systemic Vasculitis

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Glomerular syndrome can result from idiopathic and acquired diseases, with a variety of signs and symptoms. The severity ranges from asymptomatic urinary abnormalities, glomerulonephritis, nephrotic syndrome, rapidly progressive glomerulonephritis (RPGN), and chronic glomerulonephritis.

RPGN is a clinical syndrome manifested by features of glomerular disease with extensive crescent formation and progressive loss of renal function over a comparatively short period of time. The term RPGN is usually due to one of the three broad mechanisms of glomerular injury, including anti-glomerular basement membrane antibody disease, immune complex disease, and pauci-immune glomerulonephritis.

Pauci-immune RPGN refers to a necrotizing glomerulonephritis, in which there are few or no immune deposits in the glomeruli on immunofluorescence and electron microscopy. The majority of pauci-immune RPGN patients are antineutrophil cytoplasmic autoantibody (ANCA)-associated vasculitis, including granulomatosis with polyangiitis, microscopic polyangiitis, eosinophilic granulomatosis with polyangiitis, and renal-limited glomerulonephritis. Some cases of ANCA-positive disease are induced by drugs. Most drug-induced pauci-immune RPGN patients are high-titer MPO-ANCA positive, especially in hydralazine and propylthiouracil. Many cases of ANCA-associated vasculitis are associated with constitutional symptoms, arthralgias/arthritis, cutaneous vasculitis, lung involvement and crescentic glomerulonephritis.

Untreated ANCA positive-associated crescentic glomerulonephritis typically progresses to end-stage renal disease over a period of several weeks. Early diagnosis with kidney biopsy and serologic testing and early initiation of treatment is essential to minimize the degree of irreversible kidney injury. Standard treatment in RPGN includes pulse corticosteroids, cyclophosphamide or rituximab, and plasmapheresis, especially if patients have lung hemorrhage, progressive kidney injury, and dialysis dependence.

Keywords: Antineutrophil cytoplasmic autoantibody (ANCA), Rapidly progressive glomerulonephritis, Crescentic glomerulonephritis