Tzu Chi Medical Journal 27 (2015) 91

Contents lists available at ScienceDirect

Tzu Chi Medical Journal

journal homepage: www.tzuchimedjnl.com

Pathology Page

Pauci-immune crescentic glomerulonephritis

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ARTICLE INFO

Article history:
Received 10 September 2013
Received in revised form
24 September 2013
Accepted 27 September 2013
Available online 26 October 2013

A 76-year-old woman presented with a corzea-like syndrome for 30 days followed by bilateral leg edema for 20 days. Urinalysis showed occult blood (4+), protein (+/−) and 60–65 red blood cells/high-power field. Laboratory data showed blood urea nitrogen 137 mg/dL and creatinine 8.6 mg/dL. Serology showed perinuclear antineutrophil cytoplasmic antibodies (P-ANCA) 157 U/mL. Histopathology of a renal biopsy specimen showed focal necrotizing glomerulonephritis with crescent formation (Fig. 1). An immunofluorescence stain failed to show positive results, indicating P-ANCA-positive-associated crescentic glomerulonephritis. Although the patient received hemodialysis, she died of respiratory failure after a 1-week course.

Rapid progressive glomerulonephritis (RPGN) is a clinical syndrome and not a specific etiological form of glomerulonephritis (GN). It is characterized by progressive loss of renal function, laboratory findings typical of nephritis syndrome, and often severe oliguria. If untreated, it leads to death from renal failure within a period of weeks to months.

The characteristic histological finding associated with RPGN is the presence of crescents (crescentic GN). There are three etiologies of crescentic GN. About 12% of patients have antiglomerular basement membrane (anti-GBM) antibodies, 44% have immune complex deposits, and the remaining 44% have antineutrophil cytoplasmic antibodies (ANCAs) with pauci-immune crescentic GN. ANCA-associated crescentic GN is a component of systemic vasculitis such as microscopic polyangitis (p-ANCA positive) or Wegener's granulomatosis (cytoplasmic-ANCA positive). The histopathology of crescentic GN shows crescent-shaped proliferative parietal cells and macrophages in the glomeruli. Pauci-immune crescentic GN is defined by the lack of anti-GBM or of significant immune complex deposition, such as our case. The onset of RPGN is much like that of the nephritic syndrome, except that the oliguria and azotemia are more pronounced. Proteinuria sometimes approaching that in nephrotic syndrome may occur. Some affected persons become anuric and require long-term dialysis or transplantation. Plasma exchange is of benefit in those with anti-GBM antibody GN and Goodpasture disease, as well as in some patients with ANCA-related pauci-immune crescentic GN.

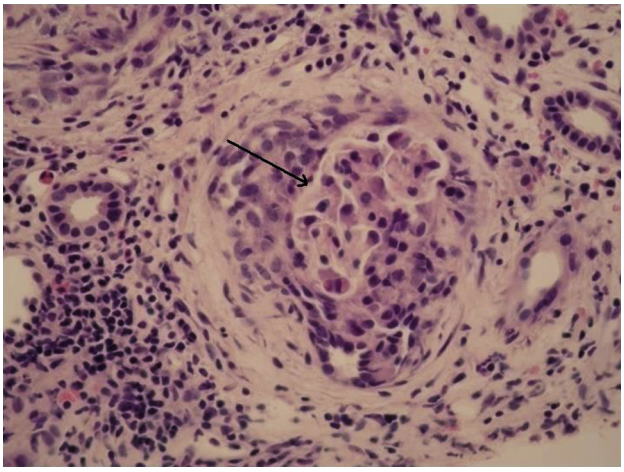

Fig. 1. Histopathology shows crescent formation (arrow) in the glomeruli (hematoxylin and eosin, 400×).

Further reading

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Conflicts of interest: none.

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http://dx.doi.org/10.1016/j.tcmj.2013.09.016