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ANCA associated vasculitis in Korea

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The anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAVs) are a group of disorders involving severe, systemic, small-vessel vasculitis and are characterized by the development of autoantibodies to the neutrophil proteins leukocyte proteinase 3 (PR3-ANCA) or myeloperoxidase. According to clinical manifestations and pathological features, AAV is classified into three variants: microscopic polyangiitis, granulomatosis with polyangiitis, eosinophilic GPA as well as renal-limited vasculitis. AAVs have a complex and unique pathogenesis, with evidence for a loss of tolerance to neutrophil proteins, which leads to ANCA-mediated neutrophil activation, recruitment and injury, with effector T cells also involved. Renal involvement, in the form of glomerulonephritis, is a common complication of AAV and indicates a poor prognosis.
Overall, the incidence of AAV has been considered low, but recent data have shown that AAVs is the most common form of new-onset GN in adults over 50. Without therapy, prognosis is poor but treatments, typically immunosuppressants, have improved survival, albeit with considerable morbidity from glucocorticoids and other immunosuppressive medications. Significant morbidities such as end stage renal disease and frequent relapses lead to poorer survival.
In this talk, I with discuss the incidence and outcomes of pauci-immune necrotizing glomerulonephritis, based on KoGNET database from 18 hospitals in Korea, from 1979 to 2017. Briefly, the incidence of pauci-immune necrotizing glomerulonephritis was the third most common pathologic diagnosis among elderly over 70 who received kidney biopsy, following membranous nephropathy and IgA nephropathy. Kidney outcome was mainly determined by kidney function at the diagnosis. Two-year kidney survival rate was approximately 90% in the patients with eGFR (estimated glomerular filtration rate) over 30 mL/min/1.73m^2, at the diagnosis. In the patients with eGFR lower than 15 mL/min/1.73m^2, 2-year kidney survival rate was approximately 50%, and most of renal event occurred within 6 months after diagnosis. The major determinant of patients' outcome was age and mortalities also occurred within early 6 months. In very elderly over 80, the mortality was more common than kidney failure.
These findings suggest that prompt diagnosis is mandatory to improve kidney outcome in patients with AAVs and treatment decision making should be based on safety issue especially in elderly.