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ANCA Related Glomerulonephritis

Bum Soon Choi
The Catholic University of Korea, Seoul St. Mary's Hospital, Korea, Republic of

Antineutrophil cytoplasmatic antibodies (ANCA)-associated vasculitides (AAV) is a group of life-threatening diseases with unclear etiology, AAV comprise granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA), and eosinophilic granulomatosis with polyangiitis (EGPA). These diseases affect small-and medium-sized vessels and are characterized by multisystem organ involvement.

GPA is characterized histologically by necrotizing granulomatous inflammation in addition to vasculitis. Common clinical manifestations include destructive sinonasal lesions, pulmonary nodules, and pauci-immune glomerulonephritis. GPA is most commonly associated with cytoplasmic ANCA and antibodies to proteinase 3 (PR3). Among European populations, prevalence ranges from 24 to 157 cases per million, with the highest prevalence reported in Sweden and the UK. MPA is characterized histologically by vasculitis without granulomatous inflammation. Common clinical manifestations include rapidly progressive pauci-immune glomerulonephritis and alveolar hemorrhage. MPA is most commonly associated with perinuclear ANCA and antibodies to myeloperoxidase. The prevalence of MPA ranges from 0 to 66 cases per million among European countries and 86 cases per million in Japan. EGPA is characterized histologically by eosinophilic tissue infiltration in addition to vasculitis. Common clinical manifestations include asthma, peripheral eosinophilia, and peripheral neuropathy. Only 40% of patients produce detectable ANCA. The overall prevalence of EGPA in European populations has been estimated to range from 2 to 38 cases per million,

Immunomodulatory drugs such as glucocorticoids (GC), azathioprine (AZA), methotrexate (MTX), cyclophosphamide (CYC) and rituximab (RTX) have improved prognosis of AAV in general dramatically. More recently, two new treatment options, mepolizumab and avacopan, were approved for EGPA or GPA, respectively. Current treatment strategies for ANCA-associated vasculitis (AAV) are based on disease severity (severe vs. non-severe) and phase of treatment (remission induction vs. remission maintenance). Patients with severe disease, defined as having life-/organ-threatening manifestations, are treated with glucocorticoids as well as cytotoxic therapies such as cyclophosphamide or rituximab to induce remission. Medications with less toxicity, such as a lower doses of rituximab, methotrexate, or azathioprine are used to maintain remission. Patients with non-severe disease, defined as not having organ-/life-threatening manifestations, are often treated with oral medications such as methotrexate or azathioprine along with glucocorticoids.

Reference

1. 1. Sarah Goglin & Sharon A. Chung, New developments in treatments for systemic vasculitis, Current Opinion in Pharmacology 2022, Volume 66, Article 102270

2. Chung SA, Langford CA, Maz M, Abril A, Gorelik M, Guyatt G, Archer AM, Conn DL, Full KA, Grayson PC, et al., 2021 American College of rheumatology/vasculitis foundation guideline for the management of antineutrophil cytoplasmic antibody associated vasculitis. Arthritis Rheumatol 2021, 73:1366–1383