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ANCA-Associated Vasculitis: Translating Pathogenesis into Targeted Therapy

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Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) is a pauci-immune small-vessel vasculitis characterized by necrotizing inflammation of blood vessels, frequently affecting the kidneys and lungs. Over the past two decades, advances in experimental and clinical research have established AAV as a disease driven by pathogenic interactions among ANCAs, neutrophils, and the alternative complement pathway. Animal studies demonstrated that anti-myeloperoxidase (MPO) IgG alone can induce necrotizing crescentic glomerulonephritis and systemic vasculitis, providing direct evidence for the pathogenic role of ANCA. Recent studies further suggest that distinct clinical phenotypes, including microscopic polyangiitis, granulomatosis with polyangiitis, and eosinophilic granulomatosis with polyangiitis, may arise from the same MPO-ANCA in the presence of different synergistic immune stimuli. Neutrophil activation is central to disease pathogenesis. Cytokines and C5a prime neutrophils for ANCA-mediated activation, resulting in reactive oxygen species production, degranulation, NET formation and ultimately endothelial injury. Activated neutrophils further amplify inflammation by activating the alternative complement pathway, generating a self-sustaining C5a-mediated feedback loop. These mechanistic insights have led to targeted therapies. Rituximab effectively suppresses ANCA production through B-cell depletion, while the C5a receptor antagonist avacopan enables glucocorticoid sparing. Emerging therapies targeting plasma cells, Fc receptor signaling, NETosis, and proximal complement components may further improve outcomes. AAV is therefore best viewed as an ANCA–neutrophil–complement amplification disease, providing a framework for the development of increasingly precise therapies.

Keywords: ANCA, Complement, Vasculitis, Pathogenesis