Granulomatosis with polyangiitis (GPA) is characterized by the granulomatous inflammation of the upper and lower respiratory tracts, necrotizing vasculitis of small and medium-sized blood vessels and necrotizing glomerulonephritis. Both cellular and humoral immune system are involved in the disease. The production of ANCA specific for the serine protease PR3 or for MPO. The cytokine-primed neutrophils and monocytes express PR3 and MPO on their cell surface membranes. ANCA binds to the cell surface and activates the neutrophils which release oxygen radicals, proteolytic enzymes, and inflammatory cytokines. Furthermore patients with active vasculitis have a lower proportion of Bm1 cells whereas patients in remission have higher proportions of CD25+ (the α-chain of the interleukin 2 receptor) and CD86+ (co-stimulatory molecule) B cells suggesting that B cells may play a regulatory role in the pathogenesis of GPA. ANCAs also induce the release of BLyS from activated neutrophils that support B cell survival in vitro. BLyS is detectable in the serum of patients with active disease suggesting that it plays a role in B cell activity and survival.

The standard glucocorticoid and cyclophosphamide treatment for GPA is often not satisfactory. Rituximab (RTX) is a chimeric monoclonal anti-CD20 antibody which causes a selective depletion of B lymphocytes. The rituximab for ANCA-associated vasculitis, which involved 197 ANCA-positive patients with GPA or microscopic polyangiitis, found that RTX therapy was not inferior to daily cyclophosphamid treatment in inducing remission and that it may be superior in relapsing disease.