

Antineutrophil cytoplasmic antibodies (ANCA) and vasculitides

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Antineutrophil cytoplasmic antibodies (ANCA) have been linked to a broad spectrum of vasculitides since they were first described in 1982 in an infection caused by Ross River virus. A prototype of these settings is granulomatosis with polyangiitis (Wegener's), or GPA. A wide array of descriptions about the ANCA associated vasculitis (AAV) now include microscopic polyangiitis (MPA), pauci-immune glomerulonephritis, Churg-Strauss syndrome, and others. To detect ANCA, 2 types of assays are currently in wide use, the indirect immunofluorescence assay, using alcohol fixed buffy coat leukocytes and enzyme-linked immunosorbent assay (ELISA), using purified specific antigens, with the former more sensitive and the latter more specific. Two types of antigens are targeted by ANCA which are reported in AAV, the proteinase 3 (PR3) and myeloperoxidase (MPO). Both PR3 and MPO are located in the azurophilic granules of neutrophils and the peroxidase-positive lysosomes of monocytes. The staining pattern of them are thus called PR3-ANCA or MPO-ANCA, respectively. Since the former is staining diffusely throughout the cytoplasm, it is also called c-ANCA. On the other hand, antibodies directed MPO stain around the nucleus and they are called perinuclear ANCA or p-ANCA. Atypical ANCA patterns may be observed on immunofluorescence staining in patients with immune-mediated conditions other than AAV. These include systemic autoimmune diseases such as systemic lupus erythematosus, inflammatory bowel disease, or autoimmune hepatitis. Such patterns may be confused with p-ANCA patterns. In addition to be a specific and peculiar mark for systemic vasculitis, ANCA are considered to play significant pathogenetic role in the development of AAV. This will be briefly discussed in the current presentation.