Antineutrophil Cytoplasmic Autoantibodies: How Should the Biologist Manage Them?

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Auteur	Beauvillain, Céline [1], Delneste, Yves [2], Renier, Gilles [3], Jeannin, Pascale [4], Subra, Jean-François [5], Chevailler, Alain [6]
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Résumé en anglais	Antineutrophil cytoplasmic antibodies (ANCA) are directed against enzymes found in the granules of the polymorphonuclear (PMN) leukocytes. They are detected by indirect immunofluorescence microscopy assays on human ethanol fixed neutrophils. Three different fluorescence patterns can be distinguished: a cytoplasmic pattern (cANCA), a perinuclear pattern (pANCA), and an atypical pattern (aANCA). The use of other fixatives, e.g., formalin and methanol, allows differentiation between the pANCA and the antinuclear antibodies. ANCA specificity is determined by solid phase assays (ELISA, immunodot, and multiplex assay). ANCA with high titres and defined specificities (antiproteinase 3 [PR 3] or antimyeloperoxidase [MPO]) are proven to be good serological markers of active primary systemic vasculitis: c/PR 3-ANCA for Wegener's granulomatosis and p/MPO-ANCA for microscopic polyangiitis. The former have higher sensitivity and specificity for Wegener's granulomatosis than the latter for microscopic polyangiitis. ANCA with low titres and unknown specificity have been detected in a wide range of inflammatory and infectious diseases leading to a critical reappraisal of the diagnostic significance of ANCA testing. Physicians must keep in mind the possible occurrence of infectious diseases like subacute endocarditis that could be dramatically worsened by irrelevant immunosuppressive therapy. ANCA findings in certain manifestations, such as the pulmonary-renal syndrome in which massive pulmonary hemorrhage can quickly be life-threatening, warrant ANCA testing as an emergency test for patient care.

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