Diagnostic Significance of Anticytoplasmatic Antibodies (ACPA/ANCA) in Detection of Wegener's Granulomatosis and Other Forms of Vasculitis

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Sir,

The presence of ACPA has facilitated the diagnosis of Wegener's granulomatosis (WG) [1,2]. This is also documented in our experience. Whereas we could identify 20 patients with WG from 1980 to mid-1986 [3], this number has dramatically increased to 51 patients up to now. This increase supports our opinion that WG is much more frequent than hitherto recognized and is only partially explained by our position as a referral center for the diagnosis of WG. All 51 patients showed the typical pattern of ACPA in leukocytes, as shown in figure 1a.

This pattern is, however, not pathognomonic since one further patient with a severe pulmorenal syndrome in need of hemodialysis turned out to have the classical histological aspect of WG, but revealed a completely different immunofluorescent pattern (fig. 1b) with fluorescence of the outer nuclear membrane as typically seen in systemic lupus erythematosus (SLE). SLE serology, however, was completely negative in this case [to be published]. In addition, we could identify 6 other patients with biopsy-confirmed severe systemic hypersensitivity vasculitis – all with renal involvement – who showed a more patchy, rosette-like distribution of ACPA as shown Fig. 1a ACPA/ANCA antibodies in a patient with typical WG exhibit a diffuse finely granular intracytoplasmic staining. 1:40. × 270. b Cytoplasmatic and punctate nuclear binding of ACPA/ANCA in a patient with WG. 1:20. × 270. c Rosette-like staining pattern in a patient with microscopic polyarteritis. 1:40. × 270.

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in figure 1c. However, one additional patient with this pattern displayed clinical and histological signs undistinguishable from typical WG.

The presence of ACPA is helpful in the diagnosis of necrotizing vasculitis. In contrast to previous authors [1, 2], immunofluorescent patterns of ACPA are heterogeneous and may allow the diagnosis of various forms of vasculitis supporting the findings of Lockwood et al. [4] who were able to identify different antigens in WG. Absence of the typical distribution of ACPA, however, does not exclude WG. Therefore, histological confirmation should be obtained in all cases of WG or related disorders.

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References

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