Antiproteinase 3 antibody in adult-onset Still's disease

Sirs,

Adult-onset Still's disease (ASOD) is a rare condition that typically affects young adults aged 16-35 years. Although it is characterized by febrile illness, typical rash and arthropathy, other systems may also be involved by the disease process. The diagnosis rests on the combination of clinical and laboratory findings. Diagnostic criteria have been developed by various investigators and those established by Cush et al. (1) and by Yamaguchi et al. (1992) (2) are frequently used for this purpose. Typically, rheumatoid factor (RF) and antinuclear factor (ANF) are negative in patients with ASOD (1, 2).

Moreover, two groups of antineutrophil cytoplasm antibodies (ANCA) are now recognized: perinuclear (pANCA), which is non-specific for a single disease and is seen in a wide spectrum of conditions, and cytoplasmic (cANCA) (3). The major autoantigen for cANCA is proteinase 3 (PR3). The antibody to PR3 is known to be highly sensitive. He also complained of intermittent sore throat with occasional fever spikes at home and during his admission to hospital a week earlier. He had been regularly taking antibiotics for a presumed bacterial throat infection. The patient had been on atenolol for the last 10 years.

On physical examination he was in moderate respiratory distress, but was not cyanosed or febrile. The BP was 145/90. His throat looked congested. Moderate features of bilateral synovitis were noticed in the shoulders and knees. A pleural rub was also present. The rest of the examination was unremarkable. There was no family history of similar illnesses.

The chest radiograph revealed bilateral, plate-like atelectasis in the lower zones, but spiral CT showed pericardial effusion, a thickened pericardium and mild pleural collection on the left side, in addition to alveolar infiltrates in both lower lobes consistent with inflammatory alveolitis. Pleural fluid was not accessible for tapping. The echocardiography was confirmatory of pericardial disease.

Laboratory findings included Hb 11 g/dl, neutrophil leukocytosis (695 of 18.5 x 10^3/uL), thrombocytosis (523 x 10^3/uL), CRP (179 mg/L, N<10), ESR (129 mm/hr), serum ferritin (1548 ng/mL, N<30-400), albumin (2.3 g/dl), hypergammaglobulinaemia, ALT 186 U/L, N 10-55, urea (109 mg/dl), creatinine (1.6 mg/dl), 24 hour urinary protein (0.750 g/day, N 0.05-0.1), creatinine clearance (24.2 ml/min/m²) and sodium in spot urine test (53 mmol/L, N<10). The ANF, RF and anti-PR3-antibody were negative in patients with ASOD (1, 2).

The anti-myeloperoxidase (MPO) ELISA (IgG) was negative. The anti-proteinase 3 (PR3) ELISA (IgG) using the International Reference Laboratory (Euroimmune, Lubeck, Germany) (5) was positive at levels of 380 and 400 u/ml (N<15) on separate occasions. The ANCA (cANCA) was negative.

The patient showed no evidence at any point during his illness or the follow-up to suggest Wegener's granulomatosis (WG) according to the 1990 ACR classification criteria of Leavitt et al. (10). We therefore felt strongly that the presence of the highly specific PR3-ANCA did not justify revising the initial diagnosis of ASOD. Nearly, 10 months have passed in the follow-up without the patient developing new physical manifestations. Apart from minimal proteinuria, renal and hepatic functions, ESR, CRP and serum ferritin have been normal on subsequent testing. Concomitantly, PR3-ANCA has significantly dropped to 80 u/ml, while the MPO-ANCA, ANF, anti-dsDNA and RF remained negative.

In our opinion this case falls within the category of clinical heterogeneity of antiPR3-associated diseases, in which the high specificity of anti-PR to WG is challenged. Perhaps this finding is peculiar to non-Caucasian populations so far. In a recent interesting report by Lee et al., among a group of predominantly Chinese patients with positive PR3, only 22% had WG. The diagnosis in the remaining patients varied from systemic vasculitis to renal disease possibly related to underlying vasculitis, livedo reticularis, amyloidosis, relapsing polychondritis, mononeuropathy and some unclassified cases (11). Therefore, it appears that testing for c-ANCA and anti-PR3 can certainly provide valuable and supportive information for the diagnosis of WG, but they alone should not lead the clinician to such diagnosis in the absence of the respective criteria. To the best of our knowledge, we are not aware of any similar associations having been reported in the English literature.

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References

Letters to the Editor


Catastrophic antiphospholipid syndrome presenting with renal thrombotic microangiopathy and diffuse proliferative glomeru-lonephritis

Sirs.
The catastrophic variant of the antiphos-pholipid syndrome (APS) is an unusual but often lethal form of presentation of this syn-drome characterized by a rapid develop-ment of multiorgan failure, mainly due to thrombotic microangiopathy in several organs (1). Since the early description of the catastrophic APS (1), more than 300 cases have been collected, being the kidney one of the more commonly affected organs (70%) (2). However, there are no previous reports of the simultaneous presence of dif-fuse proliferative lupus glomerulonephritis and renal thrombotic microangiopathy (TMA) as the first manifestation of cata-strophic APS.

A 29-year-old Caucasian man was admitted at the Emergency Department in June 2004 due to the appearance of generalized oede-ma in the last 4 weeks accompanied by decrease in urine output. He had been diag-nosed as having systemic lupus erythematosus (SLE) in 2002 due to a history of Evans' syndrome, recurrent leg ulcers, per-etebral effusion in a 50-year-old woman. PMJ 2001; 77: 347, 355-7.

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