Look for Skin Lesions in ANCA-Positive Patients

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BUENOS AIRES — A positive test for antineutrophil cytoplasmatic antibody may be associated with a vasculitis of the skin, but the test is usually not specific, so pa-

tients with other diseases may test positive, Dr. Jeffrey Callen said at the 21st World Congress of Dermatology.

To support a diagnosis of the ANCA-associated vasculitides, a positive ANCA test should be correlated with other clinical symptoms. “Skin disease is a rel-

atively common feature in ANCA-associated vasculitis and can be an initial manifestation,” said Dr. Callen of the University of Louisville (Ky.).

ANCA-associated vasculitis affects small- to medium-sized vessels of the skin, whereas purpura and polyarteritis nodosa (PPAN) are typically associated with ANCA-positive vasculitides in Wegener’s granulomatosis, microscopic polyarteritis, and Churg-Strauss syndrome. Wegener’s granulomatosis and microscopic polyarteritis are both charac-
terized by pauci-immune necrotizing vasculitis with crescentic glomerulonephritis and pulmonary capillaritis. A significant difference between the two is the presence of granulomas in Wegener’s patients.

Churg-Strauss syndrome is a systemic vasculitis often tied to eosinophilia, allergic rhinitis, and asthma. Drug-induced vasculitides, like minocycline-induced disease, are also sometimes ANCA positive, he said.

ANCAs are autoimmune antibodies di-

rected against antigens in the cytoplasmic granules of neutrophils and monocytes. Most are IgG associated. They are grouped according to histochimical staining; cytoplas-
mic ANCA (c-ANCA) and perinuclear

ANCAs (p-ANCA). c-ANCA against protein

teinase 3 is specific for Wegener’s. One of the major p-ANCA’s, directed against myeloperoxidase, is seen in microscopic polyarteritis. c-ANCA is also found in ulcerative colitis, Sweet’s syndrome, polyvinylchloride-induced vasculitis, and minocycline-induced vasculitis, said Dr. Callen.

Skin lesions, primarily ulcers and palpable purpura, are a common feature of microscopic polyarteritis, and cuta-
nan manifestations have been reported up to 78% of patients with Churg-

Strauss syndrome. Skin lesions in Churg-

Strauss include subcutaneous nodules and macular or papular erythema or urticaria, but the most common is palpable purpu-

ra, seen in about half of the patients who have cutaneous manifestations. Other possible cutaneous manifestations include ul-

ceration and livido reticularis. A Churg-

Strauss granuloma on the skin is quite rare but highly specific, Dr. Callen said.

In a review of 90 patients with Churg-

Strauss syndrome at the Mayo Clinic be-

tween 1976 and 1995, investigators found 36% (40%) had cutaneous manifestations (J. Am. Acad. Dermatol. 1997;37:199-203), most frequently purpura and petechiae on the lower extremities and cutaneous nodules and papules on the elbows.

Wegener’s granulomatosis skin lesions are similar to those in Churg-Strauss syn-
drome. In addition, pyoderma gangrenosum—like lesions can occur in Wegener’s. A recent report described cutaneous manifestations of Wegener’s in 17 patients (J. Cutan. Pathol. 2007;34:739-47). Three had skin disease before systemic disease; one patient’s ANCA test was initially negative. Six had concurrent onset of skin and sys-
temic disease, and eight developed skin dis-
ease after systemic disease was diagnosed.

In a meta-analysis (JAMA 2007;298:655-

69), patients with localized disease were recommended an antibiotic like crom-
moxazole, with or without glucocorticoids. Methotrexate plus corticosteroids was recommended for generalization, non-organ-threatening disease and pulse cyclophosphamide plus corticosteroids was touted for patients with generalized organ-threatening disease. For severe renal vasculitis, plasma exchange might also be included. High-dose cyclophosphamide plus pulse methylprednisolone was recommended for diffuse pulmonary hemo-

orrhage, possibly with plasma exchange.

Dr. Callen has consulted for and ac-

cepted speaker fees from several makers of biologics used in rheumatic disease.