A 2-Phase Screening Process for Patients with Wegener's Granulomatosis: A Pilot Study

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ABSTRACT. A 2-phase screening process for patients with persistent upper respiratory tract manifestations for the detection of Wegener's granulomatosis (WG) was tested in 28 patients in this pilot study. One patient with WG was identified. A larger study is warranted. (J Rheumatol 2003;30:2420–1)

Key Indexing Terms:
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Wegener's granulomatosis (WG), a multisystemic necrotizing vasculitis associated with the presence of antineutrophil cytoplasmic antibodies (ANCA), may present in a relatively abrupt manner with clinical manifestations in classical target organs making its diagnosis relatively straightforward[1]. Often, however, patients with WG may have an insidious onset; unrelenting upper respiratory tract (URT) symptoms may be the initial clinical manifestation. A significant delay between the onset of URT symptoms and the diagnosis of WG when pulmonary, renal, and/or systemic involvement is lacking is not uncommon. Testing for ANCA under such circumstances was recommended by an international panel of experts a few years ago[2], and by others more recently[3-8]. We undertook a pilot study to determine whether testing for ANCA in patients with chronic non-seasonal URT symptoms presenting to an ear, nose, and throat (ENT) clinic is feasible and cost-efficient for the identification of patients with WG.

MATERIALS AND METHODS

Consecutive patients presenting to a university based ENT clinic with URT symptoms of 6 weeks' duration were eligible to participate. Clinical data were gathered using an ad-hoc questionnaire. Routine laboratory tests were carried out (complete blood cell count, urinalysis, and serum creatinine) and ANCA [perinuclear (p) and cytoplasmic (c) -ANCA] antibodies were obtained by immunofluorescence. ANCA specificity [myeloperoxidase (MPO) and proteinase-3] was confirmed by ELISA (Inova Diagnostics, San Diego, CA, USA). Patients positive for MPO or proteinase-3 were further evaluated with a full history, physical examination, and appropriate ancillary studies.

RESULTS

Twenty-eight of 32 eligible patients agreed to participate. Mean (± SD) age was 52.8 (± 12.1) years (range 29–80); 53% of the patients were women, 79% were Caucasian. Two patients exhibited p-ANCA antibodies, but were negative for MPO on ELISA; one patient exhibited c-ANCA antibodies proven to be against proteinase-3 by ELISA. This patient had by history, in addition to URT symptoms of about 10 weeks' duration, the recent onset of polyarthritis, malaise, weight loss, and persistent cough. Chest radiograph was normal, but a chest computerized tomography scan revealed a small peripheral cavity in the right lung. Serial urinalyses showed microscopic hematuria; serum creatinine rose from a baseline of 1.0 mg/dl to 1.3 mg/dl. Renal involvement was suspected, but a renal biopsy failed to show significant changes. This patient responded to oral administration of corticosteroids and oral cyclophosphamide with rapid resolution of all symptoms. The screening process, as described, added $200 US to each patient's assessment, or $5600 for the 28 patients.

DISCUSSION

The screening process we describe allowed identification of a patient with WG involving the upper and lower respiratory tract (and probably the kidneys as well). This patient had just started to have systemic manifestations (weight loss, arthralgias/arthritis) and it is probable that a diagnosis of a systemic disorder, specifically a necrotizing vasculitis, would have been suspected sooner or later. Nevertheless, he was diagnosed and treated before overwhelming, non-URT manifestations were clinically very severe, which, based on our experience in treating such patients, can be very costly, particularly if patients require ventilatory support and/or dialysis. Obtaining ANCA antibodies in the presence of persistent URT manifestations is in keeping with the guidelines recommended by experts[2,3,5,8]. This 2-phase screening process will have to be examined in a larger number of

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patients with persistent URT symptoms before it is implemented in busy multispeciality clinics.

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