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LETTER TO THE EDITOR

Episcleris, arthrofasciitis and hypereosinophilia: primary hypereosinophilic syndrome or atypical Wegener's granulomatosis?

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Sir,

The differential diagnosis of hypereosinophilic states can be complex. It includes a variety of conditions such as allergic diseases, infections, leukemias, eosinophilic fasciitis (EF), vasculitis, eosinophilia-myalgia syndrome, and the idiopathic hypereosinophilic syndrome (IHES) [1]. We herein describe a case of a patient with systemic disease whose diagnosis is a matter of debate.

The patient, a 37-year-old Caucasian male, bank employee, had been complaining of cyclic arthralgias for the last 9 months. Severe episcleritis of the left eye was diagnosed 6 months before admission, with partial response to oral corticotherapy. Episcleritis had recently recurred. The patient was admitted to Hospital with arthritis of the second proximal interphalangeal joint of the right hand, right olecraneous bursitis, arthritis of the right wrist, arthrofasciitis of the left elbow, and fasciitis of the right thigh.

Blood cell count revealed marked eosinophilia (from 3,000 up to 11,000 eosinophils) and erythrocyte sedimentation rate of 12 mm in the first hour. Liver and renal function tests were normal. Testing for human immunodeficiency virus, as well as for B and C hepatitis virus, were negative. Antinuclear antibodies were absent and the rheumatoid factor (RF) test was weakly positive (34 UI/ml). The antineutrophil cytoplasmic antibodies (ANCA) test was positive (C-ANCA, titer 1/40). The chest X-ray and ecochardiogram were unremarkable. A fasciomuscular biopsy of the right thigh demonstrated an eosinophilic and plasmocytic infiltrate in fascia and muscle; vasculitis and an incomplete granuloma formation were also seen in muscle. The patient showed an excellent response to a combined intravenous "pulse" of cyclophosphamide/metilprednilone (1 gram each). The current therapy includes mycophenylate mofetil 2 g daily and prednisone 10 mg daily for control of a persistent episcleritis.

At a first sight, the patient might have EF [2]. Yet we found no description of ocular disease and C-ANCA in this circumstance. Of note, EF, myositis and arthritis were early manifestations of a T-cell lymphoma according to a recent report [3].

Our patient could potentially have IHES [4, 5] (hypereosinophilia, fasciomuscular and articular disease, eosinophilic infiltrate in fascia/muscle). Characteristic features of IHES, such as pulmonary and endomyocardial disease [4, 5] were lacking. Episcleritis, present in our patient, has been previously reported in IHES [6]. As regards to ANCA, a Japanese report dated from 1999 accounted for the presence of P, but not C-ANCA, in a man with IHES manifested by liver, lung and skin eosinophilic infiltration [7]. Thus, an incomplete subset of IHES might be a possible diagnosis for our patient.

An atypical (by sparing kidney and lung) form of Wegener granulomatosis (WG) [8] is to be strongly considered in this case. A recognized characteristic of Churg–Strauss syndrome [9], hypereosinophilia is only eventually found in WG [10]. The musculoskeletal findings (fasciitis excluded) and episcleritis seen in our patient can be part of

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the WG context. Moreover, the presence of C-ANCA and of granulomatous infiltration of muscle, as well as the response to cyclophosphamide, are suggestive of WG. Lastly, we could alternatively postulate two diagnosis for this case (EF plus a localized form of WG).

In summary, we describe a case of a young patient with episcleritis, arthrofasciitis, hypereosinophilia, miofascial granulomatous/eosinophilic infiltration, low RF titers, and C-ANCA antibodies. The case is illustrative of how complex the etiologic definition of a chronic hypereosinophilic state may be.

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