

Letters to the Editor

IgA nephropathy associated with eosinophilic fasciitis: Report of a case

Sirs,

Several recent reports have described the association of eosinophilic fasciitis (EF) with autoimmune disorders such as thrombocytopenic purpura (1), thyroiditis (1) and Sjögren's syndrome (2), and with visceral involvement such as reactive hepatitis (3), multiple peripheral neuropathy (4), synovitis (5) and pulmonary disease (6). To our knowledge, there are only four cases of EF with nephropathy reported in the literature (7-10), including one case of IgA nephropathy (7). We report one additional case of EF associated with IgA nephropathy.

A 35-year-old woman with no past medical history was admitted to our hospital in March 2000 for progressive myalgias over the previous two months. Physical examination was normal except for edema in all extremities. The laboratory findings were: white cell count 16.3×10^9 cell/l (eosinophils, 32% or 5.000 cell/mm^3), lactate dehydrogenase 248 U/l (normal: 91-180), aldolase 17 U/l (normal < 7), creatin-kinase 27 U/l and serum albumin 30 gr/l. Urinalysis revealed mild proteinuria (0.7 gr/day). Immunology (antinuclear, antacentromere, anti-Scl70, anti-Jo1 and c- and p-ANCA antibodies, cryoglobulins, and complement levels) was negative except for high levels of circulating immunocomplexes (3.5 mcg/ml; normal < 1 mcg/ml). High levels of IgA were present (781 mg/dl; normal < 400 mg/dl). Examination for parasites was negative.

Renal ultrasonography and both thoracic and abdominal computed tomography (CT) were normal. A bone marrow aspirate showed the presence of 25% eosinophils and bone marrow biopsy showed no signs of myeloproliferative syndrome. Nuclear magnetic resonance of the lower limbs detected a hyperintense spot in the fasciae of the legs. A renal biopsy demonstrated preserved glomerular structures with moderate mesangial hypercellularity and mesangial IgA and C3 deposits. Forearm muscle, fascial and subcutaneous cellular tissue biopsy specimens showed fibrous thickness of the subcutaneous fat with lax collagen tissue that had spread over the muscular fascia and perimysium, chronic inflammatory infiltrate with the focal presence of eosinophils, medium vessel wall fibrosis, lymphocytic infiltrates in the small vessel walls, and proliferation of small lymphatic vessels in the subcutaneous cellular tissue. The patient was diagnosed as having both EF and IgA nephropathy and methylprednisolone treatment (60 mg daily) was started. The clinical response was excellent with

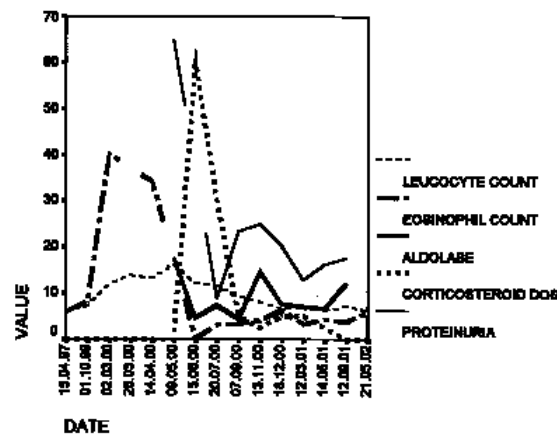


Fig. 1. Evolution of eosinophil and leucocyte count, aldolase and proteinuria levels before, during and after treatment with corticosteroids (we have transform International System of units to other units, in order to be able to put all parameters into the same graphic). UNITS: Aldolase - U/L; corticosteroids dose - dg/day; eosinophil count - cell number $\times 10^8$ /L; leucocyte count - cell number $\times 10^9$ /L; proteinuria - cg/day.

disappearance of the myalgias one month later. The methylprednisolone dose was tapered to 5 mg daily after three months of treatment (Fig. 1) and stopped one year later, without new clinical exacerbations of the disease.

To our knowledge, only four cases of renal disease have been previously described in association with EF (7-10). Caspi *et al.* (10) reported a patient with EF complicated by multisystem abnormalities, including proteinuria. Kirschstein *et al.* (8) reported renal involvement in a 17-year-old boy with EF. Renal biopsy showed ischemic collapse of the glomerular capillaries and cortical tubular atrophy. The third case was described by Janzen *et al.* (9). Renal biopsy demonstrated focal segmental glomerulosclerosis and subepithelial immune deposits. The fourth case, reported by Takeda *et al.* (7), was a 36-year-old Japanese male with a tubulointerstitial nephritis and IgA nephropathy. It is possible that tubulointerstitial involvement might be closely linked to EF, and that a delayed hypersensitivity reaction might be responsible for the impairment of renal function. On the contrary IgA nephropathy, the most common glomerular disease, is sometimes latent. The kidney biopsy of our patient showed no predominance of eosinophil cells in the inflammatory infiltrate. In addition, Takeda *et al.* (7) reported that a second renal biopsy of his patient, after corticosteroid treatment, showed no reduction in mesangial proliferation or IgA deposits despite a decrease of infiltrating cells in the interstitium. Whether this association between IgA nephropathy and EF is due to immunological defect or is a mere coincidence could not be further elucidated from these findings.

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