Autoaggressive Lepromatous Leprosy

TO THE EDITOR:

Autoaggressive leprosy was first described by Azulay ([1]) as a syndrome occurring mainly in lepromatous leprosy, and infrequently in borderline leprosy, characterized by clinical and immunologic features resembling autoaggressive connective tissue diseases. It is probably caused by the stimulation of B lymphocytes by antigenic complexes of *Mycobacterium leprae* plus autologous tissue along with dysfunction of T-suppressor lymphocytes ([2]). We describe here a lepromatous leprosy patient recently seen by us in whom there was clinical and laboratory evidence of collagen vascular disease.

A 45-year-old housewife presented with a history of edema of her hands and feet of 1 year's duration, arthralgia of the small joints of her hands and feet for 6 months, and a low-to-moderate grade intermittent fever of 4 months' duration. In addition, she complained of photosensitivity, malaise and weakness which had gradually been increasing over a period of 6 months. No history suggestive of erythema nodosum leprosum (ENL) lesions was obtained. She was diagnosed as a case of systemic lupus erythematosus (SLE), and was put on prednisolone 30 mg a day by a private practitioner. She stopped this treatment of her own accord after having taken it for about 2 weeks. No improvement was observed.

On examination, she was febrile (38.5°C) and moderately anemic with bilateral mild pedal pitting edema. A cutaneous examination revealed diffuse infiltration of the face, including the earlobes, with ciliary and superciliary madarosis. The skin over her whole body was apparently normal and only on very close inspection was it observed to be mildly infiltrated, but no infiltrated plaques or hypopigmented patches could be appreciated. The ulnar, radial cutaneous, and lateral popliteal nerves were moderately thickened bilaterally but were not tender. There was bilateral glove-and-stocking hypesthesia of 50% to all modalities. The power of the small muscles of her hands and feet was grade 4. Examinations of the cranial nerves, eyes, and mucosae were within normal limits. There was no organomegaly, and examination of the cardiovascular and respiratory systems was noncontributory. A clinical diagnosis of lepromatous leprosy was made.

On investigation, slit-skin smears from five sites revealed a bacterial index (BI) of 4+ and a morphological index (MI) of 1%. Histopathology of skin from the infiltrated skin over her back showed a thinned-out epidermis, subepidermal grenz zone, upper dermal dense lymphomononuclear and plasma cells and foamy macrophages. An acid-fast stain was positive. These features were supportive of a clinical diagnosis of lepromatous leprosy; a lepromin test was negative. A fine-needle aspiration from cervical lymph nodes showed acid-fast bacilli (AFB) suggestive of *M. leprae*. Serum electrophoresis revealed hypergammaglobulinemia. Antinuclear factor (ANF) was positive (particulate pattern); L.E. cell was positive while rheumatoid factor was negative. Serum complement C3 level was 107 mg/dl (normal 100 mg/dl). Serum biochemistry, urinalysis, chest X-ray, and X-rays of her hands and feet were essentially normal.

Blood culture on three occasions, blood for fungal culture, fungal serology, sputum for AFB, and Mantoux tests were noncontributory. A hemogram showed hemoglobin of 8.3 g/dl but other hematologic parameters were within normal limits.

The patient was put on WHO multibacillary treatment with dapsone 100 mg and clofazimine 50 mg a day along with once monthly 450 mg rifampin and 300 mg clofazimine. To control the autoaggressive phenomenon, 30 mg of prednisolone daily was added. After 1 month of treatment she was symptomatically much better with subsidence of fever and a general feeling of well being. She is presently on follow up with us and is responding to treatment. The BI and MI presently are 4+ and 0%, respectively, after 6 months of treatment.

Various manifestations of autoantibodies, i.e., ANF, L.E. cell, rheumatoid factor, and antithyroid antibodies, have been demonstrated in patients with lepromatous and borderline leprosy ([4–7]). However, autoaggressive leprosy encompasses a particular clinical presentation characterized by pro-

longed fever (37°C–40°C), anorexia, weight loss, asthenia, arthralgia, malar rash, and erythema multiform or necrotizing vasculitis-like skin lesions. In addition, one or more autoantibodies are present in the serum of these patients ([3]). Our patient had almost all these features for making a diagnosis of autoaggressive lepromatous leprosy.

The hypergammaglobulinemia and normal serum C3 level seen in our case is in corroboration with other observations ([2, 7]). The hypergammaglobulinemia could be due to polyclonal activation of B lymphocytes.

The lack of an adequate response to systemic corticosteroids in our case suggested that to control this ongoing autoaggressive phenomenon, corticosteroids alone are not as effective as a combination of antileprosy treatment and corticosteroids or thalidomide ([3]).

Although many of the above features are also present in type 2 lepra reactions, the lack of neuritis and iridocyclitis, and the presence of photosensitivity and associated autoantibodies make autoaggressive hanseniasis a distinct clinical and immunological entity.

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