Large Infiltrated Erythematous and Scaly Plaque on the Cheek and Thickened Earlobe

(See pages 1015–1016 for the Photo Quiz.)

Figure 1. Large infiltrated erythematous, scaly plaque on the patient’s cheek. Satellite erythematous papules on her neck (lower arrow) and thickened earlobe (upper arrow) were observed.

Diagnosis: multibacillary borderline lepromatous disease.

Biopsy specimens obtained from the erythematous, scaly plaque on the patient’s cheek (Figure 1) revealed diffused epithelioid granulomas with a peripheral rim of lymphocytes distributed throughout the dermis and subcutis and elongated along the course of the vessels (Figure 2). There were abundant acid-fast bacilli singly or in clumps inside the bubbly histiocytes visible on Fite-Faraco staining (Figure 3). No nerves were identified by hematoxylin-eosin and S100 protein staining.

Chemical testing revealed normal proteinemia and serum immunoelectrophoretic results. Urinalysis revealed no abnormalities. No abnormalities were visible on a chest radiograph. Bacterial, mycobacterial, and fungal cultures had negative results. Acid-fast bacilli testing of an earlobe biopsy specimen showed a bacterial index of 4+. The patient refused electroneurographic studies.

One year of therapy with clofacimina (50 mg per day), rifampicin (600 mg per month), and dapsone (100 mg per day) resulted in a marked improvement in the lesions. Nevertheless, the treatment will be continued for 1 additional year.

Leprosy is a chronic granulomatous infection that principally affects the skin and peripheral nerves and is caused by the obligate intracellular organism *Mycobacterium leprae*. Leprosy is a protean disease in which there is an inverse correlation between the bacterial index and cell-mediated immunity and in which patients present with different clinicopathological manifestations (namely, polar tuberculoid or lepromatous, borderline tuberculoid or lepromatous, mid-borderline, and pure neuritic forms) [1, 2].

Borderline leprosy involves skin lesions that are intermediate between the 2 polar forms. The morphology of lesions may be macular, papulonodular, plaquelike, annular, or with a geographic appearance [3]. Infiltration with or without nodulation over the pinna, mainly the helix, is characteristic of lepromatous
leprosy. External ear involvement may be minimal, with diffuse infiltration or discrete nodules, or extensive. Ulceration with a “nibbled” or “rat-bitten” defect may be seen [4]. Megalobule of the ear, which is characterized by a greatly elongated and wrinkled earlobe that hangs loose, may occur [5]. Auricular chondritis as part of a reactional state has been described [6].

The formation of small granulomas is characteristic of borderline leprosy, and the granulomas become more diffuse as the condition develops from borderline tuberculoid to borderline lepromatous disease. There are abundant acid-fast bacilli in lepromatous lesions, but acid-fast bacilli are not seen in tuberculoid lesions. Detection of acid-fast bacilli is useful in making a diagnosis, because the borderline states are immunologically unstable, and laboratory test results can be complicated by reactions (eg, type 1 lepra reaction, type 2 lepra reaction, and Lucio reaction) [7].

Demonstration of bacilli in a slit-skin smear is the bacteriological cardinal sign used to make a definitive diagnosis of leprosy. Involvement of common cutaneous nerves with thickening and/or tenderness is the second clinical cardinal sign used to diagnose leprosy. Skin smears can identify patients with multibacillary disease, which is the most infectious form of leprosy. When slit-skin smears are unavailable, multibacillary disease can be identified as present in patients with ≥6 skin lesions [3].

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