Annular Lesions in a 70-Year Old Austrian Man: Answer

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ANSWER
Borderline lepromatous leprosy

DISCUSSION
Leprosy or Hansen’s disease is a human chronic infectious disease associated with damaging inflammatory lesions in the skin and peripheral nerve caused by Mycobacterium leprae. Despite the claim of the WHO that it would not be a public health problem anymore after the year 2000, leprosy burden has been reduced but leprosy is far from being eliminated with more than 200,000 new detected cases each year in the last 5 years.2,3

Leprosy’s clinical manifestations are determined by a dynamic interaction process between M. leprae and the cell-mediated immunity (CMI) of genetically predisposed subjects. Leprosy patients are placed into a spectrum of clinicopathological manifestations with polar tuberculoid (TT), lepromatous (LL), and middle types of borderline tuberculoid (BT), mid-borderline (BB), and borderline lepromatous (BL) leprosy. The spectrum is determinate by the balance between CMI and bacilli: high CMI response means low number of bacilli (paucibacillary leprosy: TT and part of BT).1,4 Low CMI response means high number of bacilli (multibacillary leprosy: LL, BL, BB, and part of BT). Leprosy reactions [type 1 reaction (T1R) and type 2 reaction (Erythema Nodosum Leprosum)] are severe acute episodes that are common in the immunologically unstable borderline patients, and involve an upregulation of the host response to M. leprae antigens. T1R are responsible of nerve damage, leading to impairments and permanent disabilities.4

TT is characterized by single or a few anaesthetic, asymmetric lesions (macules, papules, plaques) with well-defined edges and dry surface. LL is characterized by numerous symmetric lesions (macules, plaques and nodules) with smooth surface and preserved sensitivity. BT leprosy has anaesthetic asymmetric lesions. BB form is characterized by symmetric lesions with polymorphous morphology and a high immunological instability with incidence of T1R. The BL form has symmetric arrangement of macules, plaques, and nodules, but fewer than in LL.3,5

Leprosy pathogenesis has not been definitively understood. M. leprae is the solely bacterium with neurotropismus for peripheral nerves and it is not cultivable, the exact mode of transmission is not known. The exact incubation time is unknown, and it can vary from few months until 20 years or more.3

Our patient was a challenge for 3 reasons. First, the rarity of leprosy in Europe, makes physicians not to think on leprosy in their differential diagnosis; this is even more true in cases like ours when the patient is not original from an endemic country. In Austria, only one case of leprosy in an African immigrant has been diagnosed in the last 10 years.6 Due to the increase in the number of immigrants from endemic areas, leprosy cases might be more frequently observed also in nonendemic areas like Europe.2 For instance, in Italy, 59 leprosy cases have been

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diagnosed in immigrants coming from endemic areas. Moreover, due to the globalization process and travels, leprosy can also be observed in European citizens working or living in endemic areas. Our patient had been living for 25 years between Brazil and Paraguay (still endemic countries for leprosy) and referred a contact with a Brazilian woman whose child was affected by lepromatous leprosy.

Second, from a clinical point of view, our patient simulated well multilocular erythema migrans (especially in an endemic area like Austria), inflammatory stage of morphea, Jessner’s lymphocytic infiltration, subacute cutaneous lupus erythematosus, erythema figuratum and mycosis fungoides. BL leprosy is characterized by bilateral symmetric macules, plaques, or nodules with red or coppery colour or slight hypopigmentation and vague edges. Lesions’ surface is smooth. Peripheral nerves may be thickened at palpation. In the early phases of disease, skin smear is negative in nasal discharge and becomes positive in advanced disease. Skin smear carried out on lesions reveals a large amount of bacilli, which are sometimes arranged in small globi. The bacterial index varies from 4+ to 5+.3–5

Last, our patient represented mainly an histopathological challenge. In fact, the first biopsy showed only a discrete mainly perivascular superficial and deep lymphohistiocytic infiltrate with few plasmacells and without epidermal changes, a major reaction pattern that has a broad histopathological differential diagnosis from lupus tumidus to syphilis, erythema migrans, scleroderma, or drug reactions among many others. Only the correlation with clinical features and anamnesis allowed to perform a Fite stainings that disclosed AFB in the nerve. In leprosy, a superficial and deep perivascular infiltrate without granuloma formation is observed mainly in the indeterminate form.3,8 Interestingly, this pattern can be also observed in active lesions of borderline patients, mainly in BL.8,9 Contrarily to TT and BT that show epithelioid granulomas, BL leprosy is characterized by a macrophage granuloma like LL but with more numerous lymphocytes filling a segment of the granuloma to its periphery. The infiltrate can be diffuse, patchy nodular, perivascular or peridendal but always separated by a typical narrow zone of collagen from the epidermis. Macrophages have foamy changes, especially in regressing lesions but no vacuoles. Nerves show an onion-skin perineurium with lymphocytes forming a cuff around a nerve bundle. AFB (4-5+) are present inside macrophages, Schwann cells, endothelial cells, and arrector pili muscles. In some cases, as the 1 herein presented, lymphocytes predominate over macrophages and BL may simulate histologically indeterminate leprosy.1,4,8

Dermatologists, dermatopathologists, and pathologists should be aware of leprosy also in nonendemic areas like Europe, to manage properly and timely these patients.

REFERENCES