RESIDENT ROUNDS: PART III

Case Report: A Papulo-Nodular Eruption With Systemic Signs and Symptoms

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ABSTRACT

This is a typical presentation of erythema nodosum leprosum in a patient with lepromatous leprosy who recently migrated from Micronesia. The clinical presentation, pathology findings, pathogenesis, and therapeutic options are reviewed here.

CASE REPORT

A 27-year-old male who had migrated from Micronesia 12 days previously presented with a 2-day history of rash, fever, weight loss (25 lbs), night sweats, and acute pain in his left leg that prevented ambulation. Upon examination, he was observed to have scattered discrete 5 mm to 15 mm erythematous papules and nodules on the trunk and extremities (Figure 1), skin-colored nodules on his earlobes bilaterally, and a 30 mm erythematous and tender left inguinal lymph node. He had no mucus membrane or neurologic abnormalities. Laboratory abnormalities included a leukocytosis and elevated serum lactate.

Punch biopsies were obtained from nodules on his right arm and elbow. The biopsies revealed dense granulomatous inflammation in the dermis, subcutaneous fat, and along nerves with admixed areas of acute inflammation and numerous globi (Figure 2). Periodic acid-Schiff (PAS) and Gomori methenamine silver (GMS) stains were negative, while acid-fast bacillus (AFB) and Fite stains highlighted acid fast bacilli present within globi (Figure 3), confirming the diagnosis of erythema nodosum leprosum (ENL).

DISCUSSION

Approximately 30% to 50% of leprosy patients will experience a secondary immune reaction.[3] Our patient experienced a type 2 lepra reaction (ENL) 1 year after completing the World Health Organization (WHO) treatment protocol for borderline lepromatous leprosy. An ENL reaction is fueled by a T helper type 2 cell and interleukin 4 response to antigen-antibody complexes precipitating in the blood, resulting in a small vessel vasculitis; and may occur before, during, or after treatment for leprosy. The diagnosis of ENL is made on a clinical basis, supported by histopathology and a confirmed diagnosis of leprosy. Scattered erythematous, tender, papulo-nodules develop acutely in concert with constitutional symptoms and lymphadenopathy. Myalgias, severe joint pain and swelling, hepatosplenomegaly, glomerulonephritis, and orchitis can also occur.[3]

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Although thalidomide is considered the treatment of choice, the WHO does not recommend thalidomide for initial treatment given its teratogenicity.[3] Instead prednisone is given initially at 1 mg/kg for 12 weeks, with the addition of 100 mg of clofazimine 3 times per day if necessary. Small maintenance doses are often given to keep the reaction in remission.[3] Our patient was placed on 60 mg of prednisone daily, but his ENL reaction persisted, so thalidomide 100 mg daily was added. He will be followed in one of the 16 National Hansen's Program's clinics in Dallas, Texas, where he will receive free care, medications, and monitoring.

FIGURE 1. Scattered ill-defined erythematous nodules and papules on the extensor forearm.

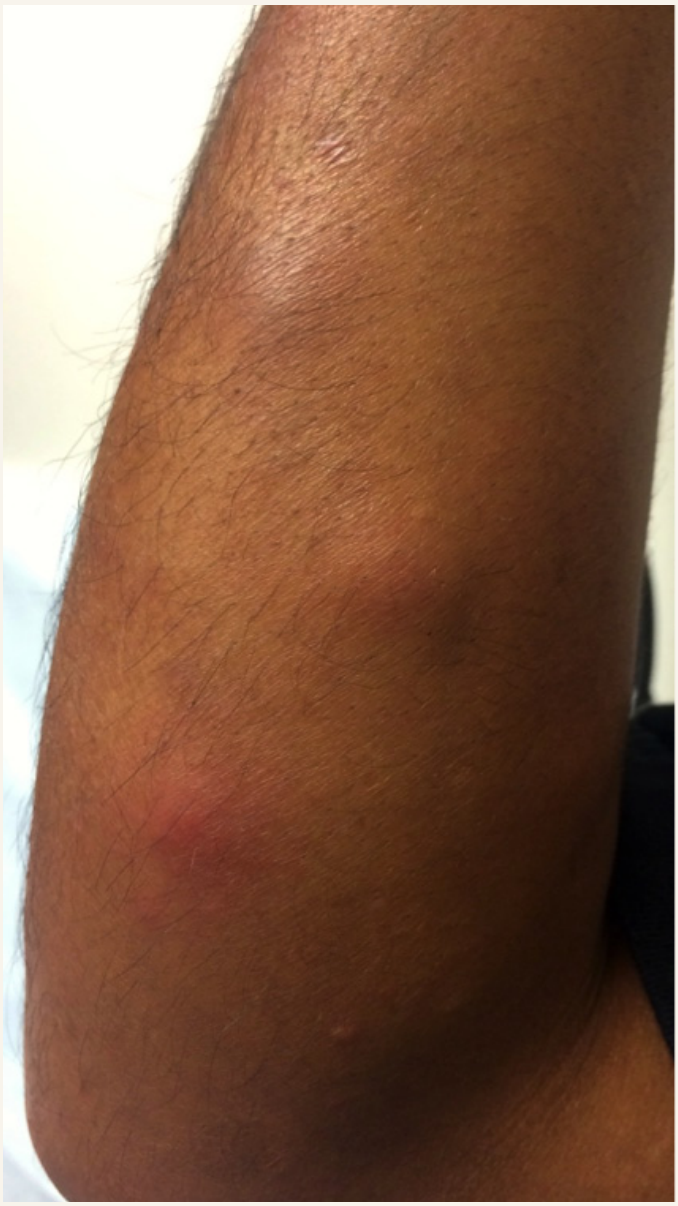


FIGURE 2. 4 mm punch biopsy from right arm with granulomatous inflammation tracking nerves (hemotoxylin-eosin, original magnification 4x).

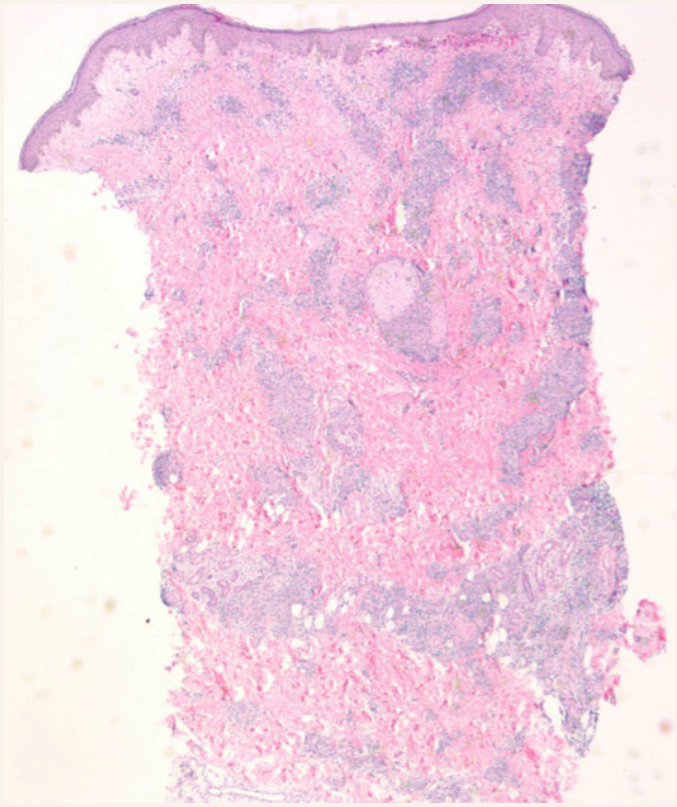
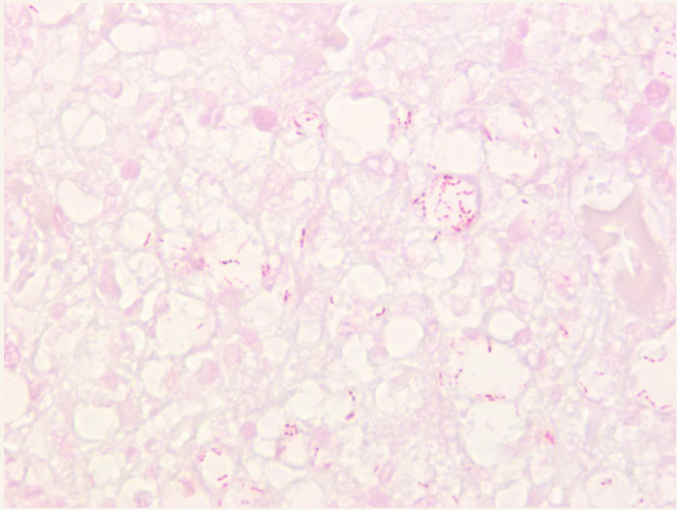


FIGURE 3. 4 mm punch biopsy from right elbow showing globi and mycobacteria (Fite's stain, original magnification 40x).



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DISCLOSURES

None of the authors has declared any relevant conflicts of interest.

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