Sixteen-Year-Old Boy From Nigeria With a Rash

Craig A. Shapiro,1 Douglas Parker,2,3 and Joseph A. Hilinski1
1Division of Pediatric Infectious Diseases, and Departments of 2Pathology, and 3Dermatology, Emory University School of Medicine, Atlanta, Georgia

Corresponding Author: Joseph A. Hilinski, MD, Division of Pediatric Infectious Diseases, Emory University School of Medicine, 2015 Uppergate Dr NE, Rm 534, Atlanta, GA 30322.
E-mail: joseph.hilinski@emory.edu.

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Case Presentation
A previously healthy 16-year-old young man returned from a 4-year residence in Awka, Nigeria, with a rash on his face, back, and upper and lower extremities that had become more prominent over the past 4 months. The rash, first noted on his lower extremities 12 months earlier, was pruritic. The rash that involved his upper extremities and face had developed over the past 4 months and was nonpruritic and painless. He denied any fever, weight loss, or visual or neurologic deficits. He reported no underlying medical conditions, and he had no known allergies. He was not taking any medications.

He was born in Nigeria but immigrated to the United States with his family when he was 2 years of age. He attended a boarding school in Awka, Nigeria, for 4 years prior to presentation. He reported no classmates or teachers with similar dermatologic symptoms. He made periodic trips to his grandmother’s residence in a rural village where he bathed in and drank water from a local stream and had contact with chickens, goats, and cows. He denied being sexually active.

His vital signs and examination were normal except for 2+ pitting edema up to his knees bilaterally. Lichenified skin was visible on the anterior aspect of both lower extremities. There were papules, nodules, and plaques on his face, ears, and upper and lower extremities with sparing of the trunk (Figures 1 and 2). Hypopigmented patches were noted on the thoracic
and lumbar regions of his back. The temporal, ulnar, and popliteal nerves were visually prominent and easily palpable. His neurologic examination was grossly intact. Sensory examination, including fine touch and proprioception, was normal.

On evaluation of laboratory values, the white blood cell count was 6840 cells/μL (reference range, 4500-13 500 cells/μL), with the following cell percentages: neutrophils, 68%; lymphocytes, 22%; monocytes, 4%; and eosinophils, 6%. The hemoglobin concentration was 12.6 g/dL (reference range, 13.0-16.0 g/dL), and the platelet count was 279 000 cells/μL (reference range, 150 000-450 000 cells/μL). Results of a complete metabolic panel were normal, including transaminase levels. The results of a urinalysis were also normal. Chest radiographs did not reveal any pulmonary opacities or mediastinal lymphadenopathy; the heart size was normal. An enzyme-linked immunosorbent assay for human immunodeficiency virus was negative.

Punch biopsies were obtained from the ear, upper arm, and elbow. Hematoxylin-eosin staining demonstrated a histiocytic infiltration of the dermis and specifically the superficial nerves (Figure 3). Ziehl-Neelsen staining did not identify any acid-fast organisms, but additional stains were subsequently performed that identified the organism.

**Diagnosis: Lepromatous Leprosy (Hansen’s Disease)**

The diagnosis of lepromatous leprosy (Hansen’s disease) was confirmed on the basis of the histological appearance of numerous bacilli on Fite-Faraco staining (Figure 4) and the invasion of histiocytes into peripheral nerves (Figure 3). The patient was treated with dapsone, rifampin, and clofazimine under the guidance of the National Hansen’s Disease Program in Baton Rouge, Louisiana. The lesions noticeably decreased in size and distribution within 6 months of therapy initiation. A repeat biopsy will be performed after 1 year of therapy to help guide duration of therapy with a minimum 24-month treatment course.

Leprosy is a slowly progressive disease caused by the partially acid-fast bacillus *Mycobacterium leprae*. Based on surveillance data reported by the World Health Organization, in 2010 there were 230 000 new cases of leprosy worldwide, with 11% originating in Africa. It is a relatively rare disease in the United States; only 150 cases were reported to the Centers for Disease Control and Prevention in 2008 [1]. The rarity of this condition contributes to delays in diagnosis, especially in the pediatric population, with an average time to diagnosis of 2 years [2]. Transmission is complex and susceptibility is related to the host’s cell-mediated immune response to mycobacterial antigens. Family members who are genetically related to an infected person have a greater probability of becoming infected themselves. As such, it is recommended that all genetically related family members living with the index case have a thorough dermatologic exam performed at least twice a year for signs of disease. Spread is believed to be through nasal droplets, although contact with open lesions may also transmit bacilli.

Leprosy should be considered in the differential diagnosis of anyone traveling from an area where the disease is endemic who has slowly progressive non-healing skin lesions, which can occur over 2-5 years.
Neurologic deficits are very helpful in making a diagnosis of leprosy and occur in 12%-55% of cases [4]. These most commonly manifest as loss of sensation to temperature and touch, which is caused by damage to peripheral nerves. Firm, palpable peripheral nerves are another hallmark of disease. Clinical presentations can vary due to the broad spectrum of immunological responses to *M. leprae*. Other infections that can cause similar-appearing lesions include cutaneous onchocerciasis and infections with other mycobacterial species including *Mycobacterium marinum* and *Mycobacterium ulcerans*. Cutaneous leishmaniasis should also be on the differential diagnosis if ulcerative lesions are present. The anesthetic nature of the lesions, however, is specific to *M. leprae* infections. The diagnosis of leprosy should be made in the context of epidemiologic exposures and physician examination findings, and it should be supported by histopathological analysis. The disease is categorized into 2 main forms, lepromatous and tuberculoid, on the basis of a classification system developed by Ridley and Jopling in 1966 [5]. In the lepromatous form of leprosy, characteristic facial features including thickening, furrowing, and coarsening of the skin, known as *leonine facies*, are often seen (Figure 1). Diffuse nodular and papular lesions characteristically develop on the elbows, knees, and ears because of the organism’s affinity for areas where the body surface temperature is lower. Tuberculoid leprosy is characterized by hypopigmented anesthetic plaques. The type of leprosy that develops is determined by the host’s cell-mediated immune response, with a stronger response predisposing toward tuberculoid forms of the disease. Thickening of the peripheral nerves and anesthetic areas, where invasion of the mycobacteria into the superficial nerve endings has occurred, is a hallmark of either form of the disease.

Biopsy and histopathologic examination of the lesion confirm the diagnosis [6]. Because these mycobacteria are only partially acid-fast, a modified acid-fast bacilli staining method known as Fite-Faraco, which uses peanut oil, is necessary to identify organisms under direct microscopy (Figure 4). Visualization of invading organisms into the peripheral nerves is pathognomonic for an *M. leprae* infection (Figure 3) [7].

For the tuberculoid form of the disease, therapy involves the use of 2 drugs, rifampin and dapsone. In lepromatous cases a third drug, clofazimine, is added to reduce resistance leading to recurrence of disease. Clofazimine is also used to reduce the risk of reactions: type 1 (reversal reaction) and type 2 (erythema nodosum leprosum), which are more commonly seen in patients who have started treatment. The National Hansen’s Disease Program recommends up to 24 months of therapy for lepromatous disease. When multidrug therapy is initiated early, the long-term morbidity and disfigurement associated with leprosy can be prevented. The recurrence rate is <3% [8]. Secondary complications can be prevented by prompt diagnosis and early initiation of therapy.

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References


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