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A case of lepromatous leprosy in South Carolina

Leslie Ching, OMS-IV, and R. P. Januschowski, DO

From Spartanburg Regional Medical Center, Spartanburg, SC

Hansen's disease, or leprosy, is a rare finding in the United States; the National Hansen's Disease Program reported 137 new cases in 2006.[1] Most of these cases are found in immigrants from Mexico, Southeast Asia, the Philippines, and the Caribbean, and in the states where there is a correspondingly high number of immigrants from these countries, i.e., California, Texas, New York, and Hawaii.[2] It is not uncommon for the diagnosis of Hansen's disease to be delayed because of health care providers' unfamiliarity with the presentation.[1] This case report presents a Mexican immigrant in a small South Carolina city who had a 2-year delay in treatment of Hansen's disease because of a delayed diagnosis and lack of follow-up by the patient, which possibly contributed to his increased morbidity.

Microbiology and epidemiology

The causative agent of Hansen's disease is *Mycobacterium leprae*, an obligate intracellular acid-fast bacillus that has only been demonstrated to be able to exist in humans, armadillos in specific areas, and sphagnum moss.[2] The primary route of transmission is currently thought to be respiratory, although there is also some evidence for transmission via infected soil.[2] Risk factors are continued close contact with patients with Hansen's disease, especially those with multibacillary leprosy; age (a bimodal distribution with higher risks from 5-15 years of age and over age 30); and impaired cell immunity.[3] *M. leprae* is not considered highly contagious; therefore, universal precautions only are recommended.[3]

Corresponding author. Ms. Leslie Ching, 5972 Westchester Park Dr. #201, College Park, MD 20740.

E-mail address: lching@vcom.vt.edu.

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Diagnosis and classification systems

The diagnosis of leprosy is generally a clinical one, although skin biopsies that display acid-fast bacilli are considered very helpful for the diagnosis and classification of the degree of pathology. *M. leprae* is notoriously difficult to culture in a laboratory, so identification by culture is not recommended.

There are two main classification systems of Hansen's disease: one is the Ripley-Jopling system (RJ) and the other is the World Health Organization system (WHO). RJ classifies Hansen's disease by clinical findings and histopathology into tuberculoid leprosy (TT) and borderline tuberculoid (BT); borderline leprosy (BB), mid-borderline, and borderline lepromatous (BL); and lepromatous leprosy (LL). TT is less severe and the effects are generally in the skin and peripheral nerves. LL is more severe and can result in the classic leonine facies and other neuropathies, but generally does not involve crucial organ systems. WHO classifies Hansen's disease by the number of skin lesions and how many bacilli are detected on skin smear in the lesions, into multibacillary and paucibacillary classifications.[2]

Further immunological complications are possible. Type 1 lepra reactions are more common in patients with borderline states of leprosy and involve edema within existing lesions. Type 2 lepra reaction, or erythema nodosum leprosum (ENL), is associated with lepromatous leprosy. ENL features fever, along with multiple erythematous tender nodules and various degrees of organ pathology, and can rarely be lethal.[2] A rare reaction, Lucio's phenomenon has been noted in patients from the Caribbean and Mexico with untreated diffuse lepromatous leprosy, resulting in ulcerations of the lower extremities that can become secondarily infected.

Clinical features

In humans, *M. leprae* is the only bacterium to invade the peripheral nervous system and it affects other organ systems

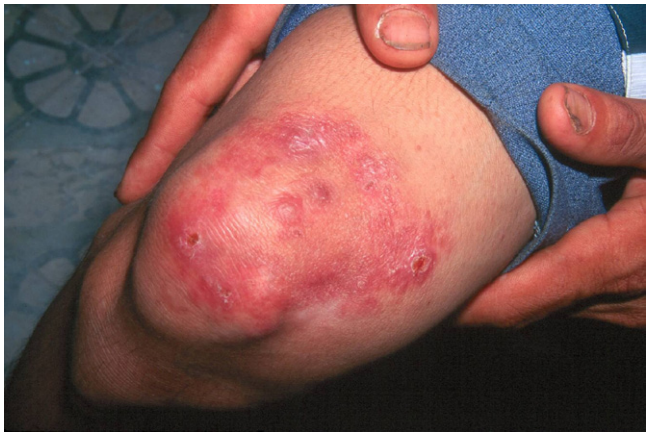

Figure 1 A representative picture of the chronic insensate lesions on the patient's trunk and arms. Image reprinted with permission from eMedicine.com, 2009.

as well. Deformation of the facial features and extremities are, of course, the most notorious complications since Biblical times: these would include saddle-nose deformity from nasal cartilage destruction, as well as extremity myopathy and neuropathy, which can lead to Charcot joints, plantar ulceration, foot-drop, clawing of the hands, and loss of digits. However, Hansen's disease can also result in corneal insensitivity, leading to trauma, secondary infections, and corneal ulcerations; orchitis, leading to elevations of LH and FSH, decrease in testosterone, and aspermia or hypospermia; amyloidosis, leading to hepatic or renal dysfunction; and nerve abscesses.[2]

Case report

The patient is a 28-year-old Mexican immigrant who had been in the United States for 5 years when he presented to the Spartanburg Regional Medical Center Emergency Department (SRMC ED) with a chronic rash over his entire body, sudden fever, generalized body aches, and malaise. He had made numerous ED and outpatient clinic visits over the past 2 years for this rash, which began at his head and now covered his entire body. He was referred to an area dermatologist who did skin biopsies on two separate occasions. He was subsequently lost to follow-up until this ED visit (Fig. 1).

He reported no known drug allergies and denied taking medications, tick exposure, or recent travel. His only past medical history is reported venous stasis ulcers of his lower extremities 7 to 8 months prior. He denied tobacco, alcohol, and illegal drug use. His twin brother, with whom he lived, was unaffected.

Review of systems was positive for recent onset of fever, night sweats, and chills. He reported fatigue and weakness for several months, and a 2-year history of rashes that started on his face, then spread to his trunk and his extremities. He denied pruritus but reported that the rash was somewhat painful and felt different than the rest of his skin. He noted a change in his facial features and denied any other symptoms (Fig. 2).

Physical examination showed a well-developed, well-nourished male who appeared older than stated age. He had coarse facial features and thinning of eyebrows more laterally than medially. Examination of his skin showed raised, erythematous, macular, and irregularly configured rashes throughout his body that had decreased sensation compared with the areas of skin without lesions. He had hyperpigmented skin lesions on his lower extremities, more so from the knees down, which were more prominent in the dependent portion and were historically related to the treatment he received at the wound center. There were no deformities of his ears, hands, or feet. The rest of his physical examination was normal (Fig. 3).

At this point, the differential diagnosis included Hansen's disease, complications of tuberculosis, other types of cutaneous infections, infections usually secondary to HIV, sarcoidosis, histiocytoma, and mycosis fungoides (cutaneous T-cell carcinoma).

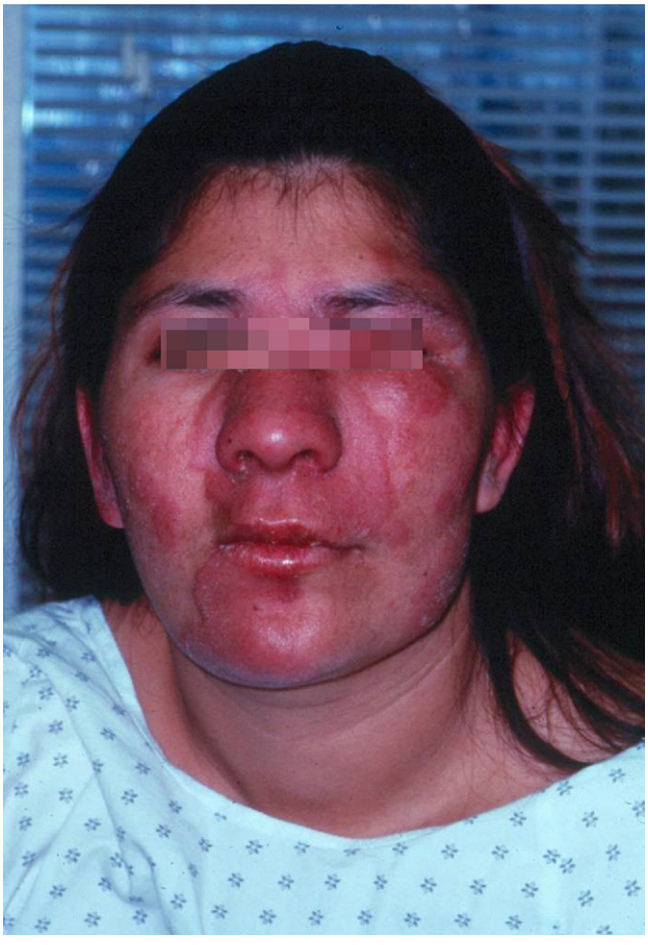

Figure 2 A woman with lesions from multibacillary leprosy and secondary erythema nodosum leprosum, which were also seen on the patient. Image reprinted with permission from eMedicine.com, 2009.

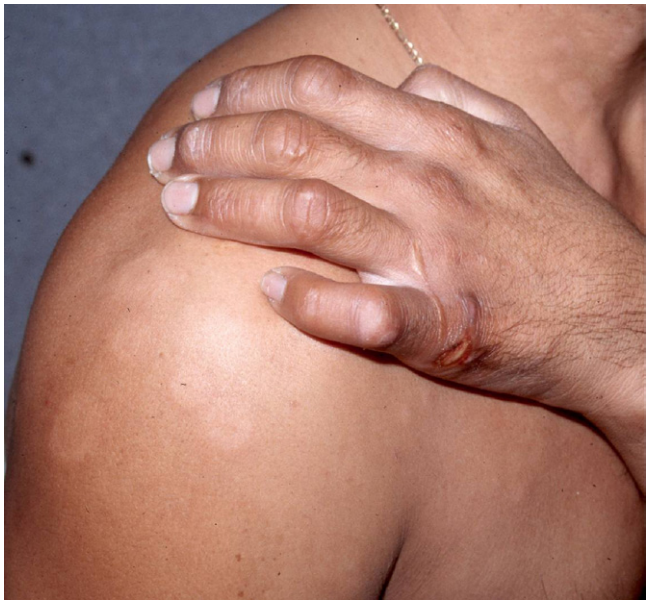

Figure 3 A representative picture of the hypopigmented lesions of multibacillary leprosy, which were also seen on the patient. Image reprinted with permission from eMedicine.com, 2009.

Pertinent data

- Complete blood count: white blood cells 18,1000 (65% segs, 14% bands, 11% lymphs, 5% monos, 4% eosinophils, 1% myelocyte), hemoglobin 11.2 g/dL, mean cellular volume 79.4 fL.
- C-reactive protein (CRP) 20. Metabolic panel and urinalysis were normal. HIV and RPR negative. Blood cultures negative to date. Tests done for tuberculosis were negative.
- Skin biopsies were found to have numerous beaded acid-fast bacilli. The presumptive diagnosis was leprosy.

Assessment/plan

Given the results of the laboratory studies and skin biopsies, the differential diagnosis was revised to include lepromatous leprosy with possible erythema nodosum leprosum and Lucio's phenomenon complications.

The diagnosis of lepromatous leprosy was based on the rash appearance, anesthetic quality of raised areas of rash and facial appearance (loss of lateral eyebrows and flattening of bridge of nose). This patient's presentation with multiple lesions and skin biopsies that showed multiple acid-fast organisms classified this as multibacillary. High CRP, leukocytosis, and microcytic anemia are likely secondary to this disease process. His unexplained fever and history of hyperpigmented areas on his lower extremities, coupled with his lack of treatment for lepromatous leprosy, made ENL and Lucio's phenomenon considerations in his differential diagnosis.

The plan for this patient was to refer him to the National Hansen's Disease Treatment Center in Louisiana for treatment and follow-up with the SRH Infectious Disease Clinic upon his return. The medication regimen for the patient's leprosy involved controlled medications, such as clofazimine and thalidomide. Standard treatment of multibacillary leprosy in the United States includes a multidrug regimen of dapsone 100 mg/day, rifampin 600 mg/day, and clofazimine 50 mg/day for 2 years.[3] Type 2 reactions are treated according to degree of severity, but can include prednisone, clofazimine, and/or thalidomide for as long as is clinically indicated.[3]

Discussion

As this case illustrates, leprosy should be considered in cases of rashes in various states of healing, with diminished sensation in the lesions, especially in immigrants from endemic countries. It is a disease that can affect many organ systems so although it is not as contagious as traditionally thought, it should be aggressively treated once identified because delayed treatment can cause increased morbidity and mortality, as in our patient. Prompt treatment with a multidrug regimen has been shown to decrease morbidity and mortality.[3]

Health professionals can discuss patient care and disposition with the National Hansen's Disease Program, which runs a medical center in Baton Rouge, Louisiana, and ambulatory care clinics in 11 states.[1] Hansen's disease is a nationally notifiable disease.

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