

What Is Your Diagnosis?



A 57-year-old Vietnamese woman presented with facial redness on the left cheek of 6 months' duration. She attributed the redness to an allergic reaction from eating seafood. Treatment with oral doxycycline was not effective. While living in Vietnam, she had been treated with a combination of clobetasol propionate and neomycin cream 0.05%.

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The Diagnosis: Hansen Disease (Leprosy)

The patient initially presented with facial redness (Figure 1). Examination 2 months later in Mississippi revealed annular erythema and a mild eczematous eruption on the lower leg (Figures 2–5). Treatment with desoximetasone cream 0.25% was not successful. A biopsy performed on a follow-up visit revealed a superficial and deep perivascular lymphocytic infiltrate. A drug eruption or viral exanthem was suspected. Testing for Lyme disease and serologic studies for lupus erythematosus were negative. Direct immunofluorescence was non-contributory. A biopsy submitted for routine histology revealed epithelioid cell granulomas, some surrounding nerves. Ziehl-Neelsen stain revealed acid-fast organisms within granulomas, confirming a diagnosis of Hansen disease (leprosy).

The earliest descriptions of leprosy are from India (600 BC). The disease spread to Europe in the 4th century with a peak incidence in the 13th century. *Mycobacterium leprae* was discovered by G.H. Armauer Hansen in 1873 and was the first of the mycobacteria to be associated with skin disease.¹ Although leprosy is relatively rare in the United States, millions of infected patients have been reported worldwide, with approximately 1 case per 10,000 individuals globally.² Frequent travel to and immigration from endemic areas in Asia, Africa, and South America (Brazil, India, Madagascar, Mozambique, Myanmar, and Nepal [in order of descending frequency]) have led to occasional cases encountered in New York, California, and Florida; endemic disease also has been reported in Louisiana, Texas, and Hawaii. Of note, indigenous cases of leprosy also have been reported in the Mississippi Delta.^{2,3} There are approximately 6500 patients with leprosy in the United States and most patients acquired the

infection outside the country; approximately 3300 of these cases require active medical management.⁴

Leprosy is thought to be an aerosol-transmitted disease spread by infected nasal secretions.⁵ The incubation period ranges from several months to many decades. Skin lesions and nervous system involvement are characteristic of leprosy.⁵ Because early lesions may be subtle in their manifestation, leprosy can be easily overlooked if it is not considered in the clinical differential diagnosis. Early cases may have a nonspecific clinical appearance; however, as lesions progress,



Figure 1. A 57-year-old Vietnamese woman with erythema on the left cheek of 6 months' duration.



Figure 2. Nonspecific erythema and mild eczematous eruption on the right lower leg.



Figure 3. Annular, nonspecific, erythematous eruption surrounding an area of hypopigmentation on the right posterolateral back. A linear erythematous lesion is visible beneath the annular eruption.



Figure 4. Circular nonspecific erythema.

World Health Organization Therapy Guidelines for Leprosy Treatment

Disease Classification	Treatment Regimen	
	Adults (50–70 kg)	Children (10–14 years)
Paucibacillary (single skin lesion)	Rifampicin (600 mg), ofloxacin (400 mg), and minocycline (100 mg) with one-time dose of all 3 medications together	Rifampicin (300 mg), ofloxacin (200 mg), and minocycline (50 mg) with one-time dose of all 3 medications together
Paucibacillary (2–5 skin lesions)	Dapsone (100 mg daily ^a) and rifampicin (600 mg once monthly) (supervised 6-month regimen)	Dapsone (50 mg daily) and rifampicin (450 mg once monthly) (supervised 6-month regimen)
Multibacillary (>5 skin lesions)	Dapsone (100 mg daily ^a), clofazimine (50 mg daily ^a), rifampicin (600 mg once monthly), and clofazimine (300 mg once monthly)(supervised 12-month regimen)	Dapsone (50 mg daily), clofazimine (50 mg every other day), rifampicin (450 mg once monthly), and clofazimine (150 mg once monthly) (supervised 12-month regimen)

^aSelf-administered in adults; administration must be supervised in children.

Data from the World Health Organization.⁴

hypopigmentation and numbness begin to develop.¹ Leprosy often involves the facial nerves. Commonly, it also occurs in the ulnar and median nerves, giving rise to clawhand; the common peroneal nerve, leading to foot-drop; and the posterior tibial nerve, resulting in clawfoot and plantar insensitivity. Well-developed

annular and hypopigmented lesions occur in association with granulomas.¹

Leprosy usually is treated with a multidrug therapy regimen instituted by the World Health Organization that consists of several effective chemotherapeutic agents against *M leprae* including dapsone, rifampicin,



Figure 5. Annular, nonspecific, erythematous eruption on the right upper lateral back.

and clofazimine.⁴ Rifampicin is the most effective bactericidal drug against *M leprae*. Clofazimine, which is minimally bactericidal with some anti-inflammatory properties, administered in combination with dapsons substantially decreases the risk for rifampicin drug resistance.⁶

The World Health Organization advocates for the treatment of paucibacillary disease with 2 to 5 skin lesions with a combination of 2 drugs, whereas multi-bacillary disease is managed with triple-drug therapy, as shown in the Table.⁴ For optimal results, careful follow-up is needed. Our patient was lost to follow-up for a short period of time. The stigma of leprosy often prevents patients from returning for follow-up.⁷

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