Leprosy in Northwest Louisiana: A Case Series

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Abstract

Hansen’s disease, commonly known as leprosy, is a chronic mycobacterial infection caused by Mycobacterium leprae. Although generally uncommon in the United States, it is endemic in the Southern United States. We diagnosed and managed five leprosy patients from Northwest Louisiana, each presenting a distinct set of challenges. A retrospective study was performed to collect demographic, clinical, and laboratory data from our cases. The information was analyzed with a specific focus on associated factors, diagnosis, and management. The mean age at diagnosis was 67.6 years (range 56–83 years), and the average delay in diagnosis was 8.4 months (range 1–20 months). All five patients presented with nonhealing rashes, and three initially sought help from primary care providers. Only two patients developed subjective numbness. Leprosy was not suspected before skin biopsy in three cases, while noninfectious diagnoses were considered, including mycosis fungoides, erythema multiforme, vasculitis, and amyloidosis. In the other two cases, leprosy was in the initial differential diagnosis. Ultimately, the diagnosis of leprosy was established in all five individuals based on clinical presentation, routine histopathology, and tissue acid-fast staining. This case series highlights the importance of leprosy, especially in the Southern United States where its incidence is increasing.

Keywords: Hansen’s, leprosy, Louisiana, Mycobacterium leprae

Introduction

Leprosy is a chronic mycobacterial disease caused by the acid-fast bacillus Mycobacterium leprae. It generally affects the skin, peripheral nerves, eyes, and nasal mucosa, leading to rash and neurological manifestations involving the peripheral nerves. The disease has been known to mankind since biblical times, originating in ancient Egypt and Middle Eastern countries around 2400 BC. The number of new cases reported globally in 2015 was 211,973. In the United States, about 150–250 cases of leprosy are reported each year, with 178 cases reported in 2015. An epidemiological study performed on data regarding leprosy cases in the United States revealed that from 1994 to 2011, a total of 2323 new cases of leprosy were diagnosed with an average incidence rate of 0.45 cases per million persons. In the United States, leprosy is endemic in Texas and Louisiana, as well as states with substantial immigrant population such as Florida, Hawaii, Arkansas, California, and New York.

Cases of leprosy have been reported in Louisiana since the early eighteenth century. The incidence rate of leprosy in Louisiana in 2011 was reported to be 4.5/100,000 people. Leprosy was first reported to be endemic in the southern parishes in the late 1880s followed by reports from the northern parishes in 1988. The only inpatient hospital for the management of leprosy was functional from its founding in 1894–1999. Review of public health records indicated that the early inhabitants of Louisiana acquired leprosy from multiple sources; most prominent of them were settlers on the shores of the Gulf of Mexico.

Study

The study included a detailed description of five cases who were diagnosed and managed at a tertiary care hospital in Shreveport, Northwestern Louisiana. We diagnosed and treated five cases of leprosy from 2004 to 2011, some with unusual manifestations presenting significant diagnostic challenges. We performed a retrospective review of medical records from the five patients. Demographic, clinical, and laboratory data of each patient were collected.

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**Case Reports**

**Case 1**
An 82-year-old White male presented with diffuse rash all over his body in July 2004. The rash had started in January 2004 on his chest, back, and arms. There was no history of travel outside the United States. His social history was significant for killing and handling numerous armadillos. Physical examination was positive for an erythematous, patchy, confluent, raised skin rash over the chest, arms, and legs [Figure 1]. Neurological examination revealed decreased temperature sensation and proprioception in a stocking-glove distribution over the hands and feet, associated with decreased vibratory sensation involving the great toes. The patient was referred to dermatology for skin biopsy, with initial finding of nonspecific reactive process. He then developed numbness in distal upper and lower extremities, which prompted his dermatologist to request Fite stain showing acid-fast bacilli in foamy histiocytes within the perivascular infiltrates as well as around the nerves [Figure 2], consistent with multibacillary lepromatous disease. He was started on oral dapsone 100 mg daily, clofazimine 50 mg daily, and rifampin 600 mg daily and continued drug therapy for a total of 2 years. On follow-up, his rash was resolved, leaving residual hyperpigmentation in the areas of previous involvement. He had no complications or deformities. After 2 years, he was switched to dapsone monotherapy and later died of unrelated causes.

**Case 2**
A 60-year-old White male presented to an otolaryngologist in January 2008 for the excision of squamous cell carcinoma on the left nasal ala. He was referred to dermatology for an incidental finding of rash on his left elbow [Figure 3]. On his follow-up visit, we noticed that the rash had spread to both upper extremities. His medical history was notable for intermittent steroid treatment for sarcoidosis, and social history was positive for handling armadillos. Physical examination was notable for an erythematous, sharply demarcated, plaque-like lesion with irregular borders on the dorsum of forearms and hands and papular lesions on the chest and abdomen. Skin biopsy showed granulomatous dermatitis with numerous acid-fast bacilli in the collagen [Figure 4] but no evidence of neuritis. The patient was initially started on dapsone, rifampin, azithromycin, and ethambutol for empiric coverage against atypical Mycobacteria and M. leprae. Eight days later, his rash became painful, swollen, and warm, and the patient developed polyarticular inflammatory arthritis and leukocytoclastic vasculitis. This reaction responded well to a tapered course of systemic corticosteroids. Polymerase chain reaction (PCR) testing revealed M. leprae Indeterminate type of leprosy was diagnosed, and azithromycin and ethambutol were discontinued. Initial worsening of his rash and onset of inflammatory arthritis were considered to be consistent with a reversal reaction which responded to daily corticosteroid treatment. Steroids were subsequently tapered. The patient is currently receiving chronic suppressive monotherapy with dapsone with good results including flattening of all skin lesions.

**Case 3**
A 78-year-old White male was admitted to the hospital for the evaluation of syncope in August 2011. During his hospitalization, he complained of a 2-week history of rash involving both forearms. His medical history was notable for squamous cell carcinoma of the larynx, managed with radiation and surgery, as well as inflammatory arthritis treated with corticosteroids and methotrexate. Social history was significant for contact with armadillos. Physical examination revealed a scaly, pruritic, erythematous rash involving both forearms [Figure 5]. Biopsy showed perineural and periadnexal granulomas with acid-fast bacilli in the dermis. PCR was positive for M. leprae, and he was diagnosed with lepromatous leprosy. He was started on dapsone (100 mg daily) and rifampin (600 mg daily). He died 1 month later due to complications with other comorbidities unrelated to leprosy.

**Case 4**
A 64-year-old White male presented with a rash involving his left leg and ankle in January 2010. There was no history...
of malignancy or immunosuppressive treatment. Physical examination was positive for an erythematous, raised, irregular rash involving his left leg and ankle. The rash was treated with topical antifungals, but it worsened and spread to the chest, back, arms, and right leg. He was referred to dermatology in May 2010. Biopsy showed lymphohistiocytic perivascular and perineural inflammation with acid-fast bacilli visualized with Fite stain [Figure 6]. PCR was positive for \textit{M. leprae}, and he was diagnosed with lepromatous leprosy. He was started on dapsone (100 mg), rifampin (600 mg), and clofazimine (50 mg daily). His skin lesions promptly flattened. He received triple therapy for 2 years, followed by dapsone monotherapy for long-term suppression. During the months of subsequent follow-up, he had minor hyperpigmentation of previous lesions but no active lesions.

**Case 5**

A 54-year-old White male with no significant medical history presented with intermittent swelling of both hands in March 2008. Social history was significant for contact with armadillos. He subsequently developed rash, tingling, and numbness of both hands. This prompted his physician to do a skin biopsy in September 2009. The biopsy showed changes consistent with lepromatous leprosy, and he was started on dapsone and rifampin. When skin lesions worsened, he was referred to LSU Health Sciences Center, where numerous irregular erythematous macular lesions over his back and upper and lower extremities were observed [Figure 7]. He was referred to the National Hansen’s Disease Program (NHDP) for biopsy, PCR, and further management. PCR was positive for \textit{M. leprae}. Biopsy confirmed lepromatous leprosy with reversal reaction. He was started on prednisone (40 mg daily), rifampin (600 mg once monthly), clofazimine (50 mg daily), and dapsone (100 mg daily). Skin lesions improved after steroid treatment, but the patient was lost to follow-up.

**Results**

In our case series, all five patients were White males with a mean age at onset of 67.6 years, mean age at biopsy confirmation of 68.4 years, and mean delay in diagnosis of 8.4 months. Three patients had lepromatous leprosy, one had indeterminate, and one had borderline disease. All five individuals were natives of Northwest Louisiana, and only two had traveled outside the United States. Interestingly, four of the five patients recalled direct skin contact with armadillos. Malignancy and immunosuppression were present in two cases. The most common initial presenting symptom was rash. One of the patients developed numbness of his distal extremities 6 months after the onset of the rash, leading to the diagnosis of leprosy. Only one additional patient developed
sensory symptoms. In our series, three patients presented to their primary care providers, and one patient presented to his otorhinolaryngology specialist, while another patient was admitted to the hospital for the evaluation of syncope and rash. In only two of the five patients, leprosy was in the differential diagnosis before conducting a skin biopsy. Other conditions in the differential diagnosis were mycosis fungoides, vasculitis, Sweet’s syndrome, amyloidosis, sarcoidosis, and allergic drug eruption.

PCR for *M. leprae* was performed on four of the five skin biopsies at the NHDP, and diagnosis of leprosy was confirmed in all four patients. In contrast, mycobacterial cultures were uniformly negative. Leprosy was eventually diagnosed from clinical presentation, histopathology, and molecular techniques. All patients were followed up closely in the infectious disease clinic during anti-mycobacterial therapy with rifampin, clofazimine, and dapsone. Four of the five patients were successfully treated with multidrug therapy, while one patient died of causes unrelated to leprosy while on therapy. None of the patients in the study developed deformities or chronic complications involving eye, testes, or other organs.

**DISCUSSION**

The exact mechanism of transmission of the disease is not known precisely, due to long latency periods associated with the infection, sometimes as long as 20 years. However, it is widely postulated that droplet transmission is the most common possible mode of transmission. A recent study found a unique genotype of *M. leprae* among armadillos and patients from the same area in the Southern United States. By molecular methods, it was shown that leprosy could be acquired from armadillos in the Southern United States. Four of the five patients in the study had direct contact with armadillos, including hunting, kicking, and playing with armadillos. There have been recent reports of leprosy spreading eastward among armadillos. This raises the concern of occurrence of new cases in nonendemic areas.

Depending on the host response to the invading organism, leprosy can manifest either as a tuberculoid form of leprosy in patients with vigorous cellular response or lepromatous leprosy in patients with weakened cellular response. In the tuberculoid form of the disease, skin lesions and peripheral nerve involvement are the common presenting symptoms; in the lepromatous form, extensive skin lesions are the common presentation. Patients in the lepromatous spectrum tend not to present with anesthetic patch, whereas tuberculoid leprosy tends to present with numbness, nerve enlargement, and macular hypopigmented patch. Rash was the most common cause of presentation in this study. Physicians in many specialties may encounter patients having leprosy. Dermatologists should have a very low threshold to biopsy chronic skin lesions, especially in leprosy-endemic regions in the United States. Alerting the pathologist regarding suspicion of leprosy is crucial in doing acid-fast staining of the samples.

Two patients had a history of head and neck malignancy. One was treated with excision and the other with radiation and surgery. These two patients also had a history of systemic immunosuppressive treatment for sarcoidosis and inflammatory arthritis. Recent case reports suggested the possible occurrence of lepra reaction among patients treated with infliximab and HIV patients treated with antiretroviral therapy, suggesting that leprosy can be an asymptomatic latent infection. None of our patients presented with lepra reaction. Rash onset did not coincide with initiation of immunosuppressive treatment. However, considering its onset in the sixth and seventh decades of life, there is a need for further research on the effect of age, immunosuppression, and latency in leprosy.

In general, patients in the lepromatous spectrum were treated with a three-drug regimen for 2 years, followed by lifelong dapsone monotherapy. The three-drug regimen includes dapsone (100 mg oral daily), rifampin (600 mg oral monthly), and clofazimine (50 mg oral daily).

**Conclusion**

Leprosy is a re-emerging infection in the Southern United States, associated with compromised cell-mediated immunity and substantial delays in diagnosis. Our report supports the association between armadillo exposure and leprosy in the Southern USA described by Truman *et al.* Clinicians in the Southern United States should have a high index of suspicion for leprosy in patients with nonhealing rashes, even in the absence of numbness. When skin biopsies contain granulomatous inflammation, tissue acid-fast staining can lead to early diagnosis and treatment. Avoidance of delay is important because leprosy can lead to deformities. As more monoclonal antibodies, steroids, and immunosuppressive therapies are used, the incidence of lepra reaction may increase in the future.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other
clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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