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Case Report

A Case of Lepromatous Leprosy in Oregon

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The cases of leprosy occur rarely in North America and a health care professional may not readily consider a diagnosis of leprosy. We encountered a 23-year-old female with hypopigmented macules and painful nodules along with peripheral neuropathy in Oregon, who had immigrated from Micronesia. A skin biopsy confirmed the diagnosis of leprosy. Patient developed a type 2 reaction, Erythema nodosum leprosum, upon initiation of the multidrug therapy. It is vital to recognize the signs and symptoms of leprosy and associated reactions so the patient can be treated appropriately to prevent debilitating and stigmatizing neurocutaneous and systemic illness.

Keywords : Leprosy, Erythema Nodosum Leprosum, Lepromatous Leprosy, Hansen's disease

Introduction

Leprosy is a chronic and progressive granulomatous disease affecting the skin and nerves that is caused by *Mycobacterium leprae* (Aftab et al 2016). Transmission is thought to be mainly through nasal and oral droplets from close contact with the bacilliferous patient and less often from eroded skin (Sehgal 1994). Transmission of disease depends on the infectivity of the contagious patient (Sehgal 1994, Bhatia et al 1993). There is about 25% chance of acquiring the disease from household contact with a majority of new cases in children and young adults from close relatives with the disease in endemic countries (Canizares and Adrianns 1992). The incubation period varies between months to 30 years but is usually 4 to 10 years (Cohen 2003). Only 5-10% of those infected develop disease (Sugita 1995). Although considered curable and rare in North America, it is important to recognize the presentation of leprosy especially when encountering patients have emigrated from endemic areas in tropical and warm temperature regions (Britton and Lockwood 2004).

Case Synopsis

A 23-year-old female who recently immigrated to the United States from Micronesia presented with a 1-week history of painful nodules on her left lower extremity. She also complained of decreased sensation throughout her upper and lower extremities. Associated symptoms include intermittent fever and night sweats. She had been previously seen by infectious disease and was prescribed cephalexin for possible cellulitis before being referred to dermatology. Further history revealed that she

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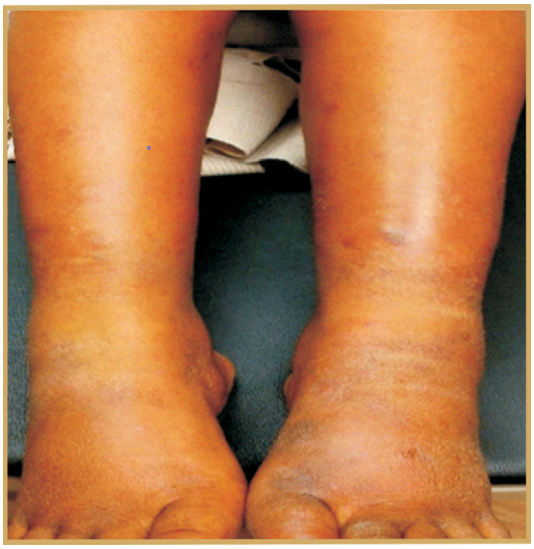

Fig 1 : Hyperpigmented nodules and significant leg edema

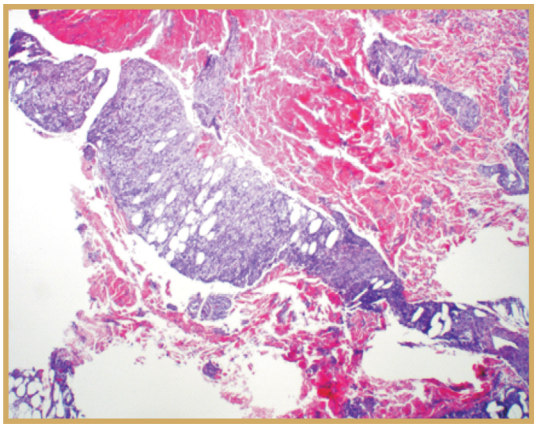

Fig 2a : Histopathology of Leprosy case, H&E

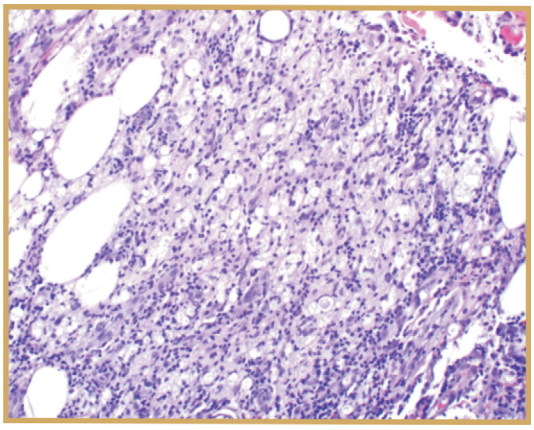

Fig 2b : Histopathology of Leprosy case, H&E

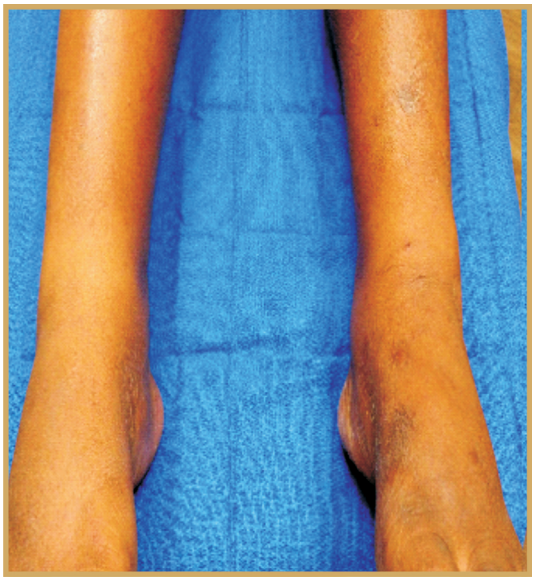

Fig 3 : Improved Edema and skin lesions 10 days into treatment

had been empirically treated with unknown medications for 9 months from August 2016 at the Leprosy clinic in Micronesia for similar nodules prior to her move to the United States. After she moved to the United States, she failed to continue her medications. She did not have a history of immunodeficiency and was not on any medications other than Ibuprofen as needed. Dermatologic examination revealed bilateral leg edema and erythema. Painful subcutaneous nodules varied in size from 0.5 to 2.0 cm. Further examination demonstrated hypopigmented macules on the bilateral upper arm, legs, and trunk (Fig. 1). Initial labs demonstrated a leuko-

cytosis and marked anemia. Her Quantiferon Gold was negative and her complete metabolic panel was grossly normal. An 8 mm punch biopsy was performed on the lateral aspect of her left leg. Histological examination demonstrated non-caseating granulomatous inflammation with numerous bacilli in the granulomas highlighted by the Fite stain (Figs 2a, 2b). Acid-Fast Bacilli smear further confirmed the diagnosis of Leprosy.

The patient was subsequently referred back to an infectious disease specialist who initiated treatment with rifampin, dapsone, folic acid, minocycline, and prednisone. Her symptoms were resolving (Fig. 3) as she tolerated the treatment. However, a month into the treatment, she developed a lump on her submental area along with painful nodules in her legs, which were consistent with a type 2 erythema nodosum leprosum reaction. A prednisone burst was started at 70 mg daily for 4 days followed by 50 mg for the next 4 days and then tapered. The patient was also referred to Ophthalmology for a retinal exam for consideration of initiating clofazimine, which is available only through the National Hansen's Disease Program as an investigational drug (Program NHsD 2017).

Discussion

Leprosy is ancient disease dating back to 1550 B.C. in Egypt (Hulse 1972) that is now primarily seen in subtropical climate of Central and South America, South Asia and Africa and rarely in North America (Canizares and Adrianns 1992). However, the number of new cases in the United States has not declined compared to that of the previous decade (Program USDoHaHSHRaSANHsD 2015).

Leprosy presents a spectrum of clinical presentation. It can be classified into two major forms, tuberculoid and lepromatous leprosy, based on histological and immunological features defined by Ridley and Jopling; there are also hybrid forms which includes borderline tuberculoid (BT), borderline leprosy (BB) and borderline lepromatous leprosy (BL) (Ridley and Jopling 1966). Tuberculoid Leprosy is characterized as a predominantly Th1 reaction with paucibacillary skin lesions which may either be one large erythematous patch or a few asymmetric hypopigmented macules or patches and can be accompanied by significant nerve damage with anesthesia or dysesthesia (Britton and Lockwood 2004, Ridley and Jopling 1966). In tuberculoid leprosy, lepromin test is usually positive which indicates a T-cell mediated, delayed type hypersensitivity reaction (Canizares et al 1992). On the other hand, the Lepromatous form presents with more acute onset. Small, multiple ill-defined, hypopigmented macules may be the earliest presentation (Longo et al 2011). Plaques or nodules may form later or they may infiltrate the skin and hair follicles, especially on the face which may result in loss of eyebrows with "leonine facies." (Ernst 2012, Kustner et al 2006)

There are variants of Leprosy reaction that are rare and therefore the pathogenesis is not completely understood. Patients may develop a delayed hypersensitivity reaction, a type 1 "reversal" reaction also called an "upgrading reaction" which may occur in borderline leprosy states (Cuevas et al 2007). Type 2 reaction, Erythema nodosum leprosum, which presents as a sudden onset acute inflammatory response due to the formation of immune complexes in association with an excessive humoral reaction (Th2 response) (Sugita 1995). Type 2 lepra reactions may develop during the natural course of disease, during antimicrobial therapy as in our case presentation, or even after completion of antimicrobial treatment of infection (Ramos-e-Silva and Rebello 2011). In Type 2 reaction, the antibodies combined with *Mycobacterium*

leprae antigen form immune complexes that circulate and deposit in various tissues, activate complement, and damage the tissues (Vashisht and Das 2014) and presents with erythematous, tender, nodules and/or plaques. About 15-50% of Leprosy patients develop erythema nodosum leprosum more often within the first year of the therapy (Ladhani 1997). Increased disease burden and a Th2 response in erythema nodosum leprosum can result in more systemic symptoms, such as neuropathy, lymphadenopathy, iritis, orchitis, glomerulonephritis and etc, along with multiple skin lesions (Longo et al 2011) Histologically, we may observe neutrophilic infiltrate with lepromatous macrophage granuloma with vascular endothelial edema (Anshu et al 2002). Severe erythema nodosum leprosum may develop ulcerative skin lesions, erythema necroticans (Davis et al 2002). Two other clinical variants of type II reaction include erythema polymorphous and Lucio's phenomenon, which is designated as either type II or type III reaction (Cuevas et al 2007).

Standard treatment of leprosy consists of multidrug therapy with rifampicin and dapsone, with or without clofazimine (Program NHsD 2017). For paucibacillary or tuberculoid leprosy cases, the recommended treatment duration is 12 months whereas for 24 months treatment is recommended for multibacillary or lepromatous leprosy cases (Program NHsD 2017). Multidrug regimen should be continued through the Erythema nodosum leprosum reaction. A type 2 reaction can be treated with prednisone 0.5 to 1 mg/kg daily with a slow tapering of 6 months or more to prevent potentially irreversible cutaneous and nerve damage (Anshu et al 2002, Lebwohl et al 2017). Thalidomide is known as the drug of choice for men and postmenopausal or surgically sterile women (Haslett et al 2005). Other drugs for type 2 reaction include clofazimine, pentoxyfylline, methotrexate, azathioprine, infliximab, and etanercept (Lebwohl et al 2017, Worobec 2009). Lucio's phenomenon, erythema necroticans, is characterized as extensive painful skin ulcerations and can be treated with systemic steroid with clofazimine and intensive supportive care (Sehgal 2005).

Conclusion

Leprosy remains a rare disease in North America. Therefore, a health care professional may rarely consider the diagnosis of leprosy, which can delay diagnosis and treatment. Leprosy should be included in a differential diagnosis when patients from endemic areas present with skin and neurological findings even years after immigration. Early diagnosis and prompt treatment of leprosy with rifampicin and dapsone with or without clofazimine is vital to halt the disease progression and prevent further transmission of disease. If *erythema nodosum leprosum* occurs, patient should be promptly started on appropriate therapy, such as prednisone or thalidomide as they are the first line therapy, to prevent permanent sensory, motor, or autonomic or peripheral nerve damage. It may also be prudent to consider prophylactic treatment in close contacts of leprosy patients (Smith and Smith 2000) and the role of chemoprophylaxis should be studied further.

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