

HANSEN'S DISEASE MIMICKING A SYSTEMIC VASCULITIS

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Abstract

Hansen's disease, caused by *Mycobacterium leprae*, classically presents with cutaneous and neurological manifestations. Rheumatologic manifestations present in 1 to 5% of the patients, and include arthritis, arthralgias, Charcot arthropathy, erythema nodosum and vasculitis. We report a case of a 86 year old woman with polyarthritis, subcutaneous nodules and leg ulcers whose differential diagnosis included primary vasculitis and diffuse connective tissue diseases and ended to be leprosy in a non endemic country.

Keywords: Hansen Disease; Leprosy; Vasculitis; Arthritis

Introduction

Leprosy is a chronic granulomatous infectious disease caused by *Mycobacterium leprae*, with predominant involvement of skin and nerves¹. It is an endemic disease that presents as a spectrum of clinical manifestations depending on the immune response of the host: lepromatous, tuberculoid, borderline lepromatous, borderline tuberculoid and mid borderline^{1,2}. Leprosy reactions are acute inflammatory states that commonly precede diagnosis or occur after the initiation of appropriate chemotherapy.

The classical presentation of leprosy is in the form of macules or papulo nodular lesions and paresthesias or sensori-motor mononeuropathy, mononeuritis multiplex or polyneuropathy^{1,2}.

Musculoskeletal manifestations range from 1 to

5% for patients with leprosy but may increase to 50 to 70% during reactional states³. They include arthritis, arthralgias, Charcot arthropathy, erythema nodosum and vasculitis⁴.

We present a clinical case with polyarthritis, subcutaneous nodules and leg ulcers that simulated a rheumatic disease.

Case Report

A 86 years old, caucasian female, presented with a five years history of symmetric polyarthritis of metacarpophalangeal, proximal interphalangeal, wrists and elbows, treated with prednisolone 5mg id and naproxen 500mg bid. In 2003 she reports painless subcutaneous nodules in the legs, and one year ago the spread of this nodules, to the upper and inferior extremities, ulcerative lesions in the legs and distal paresthesias of both upper and inferior extremities. She denied infectious diseases or journeys to endemic countries for leprosy.

She was admitted to our hospital, presenting painful nodular lesions on the extremities (Figure 1), some of them ulcerated, leg ulcers (Figure 2), symmetric polyarthritis of proximal interphalangeal and metacarpophalangeal, ulnar deviation of fingers, and distal and symmetric hypoesthesia of the upper and lower limbs. She was afebrile, with palpable and symmetric peripheral arterial pulses, and normal cardiovascular, respiratory and abdominal examination.

Laboratory blood tests revealed anaemia (Hgb 10,6 g/dL), elevated erythrocyte sedimentation rate (62 mm) and C-reactive protein (7,6 mg/L), with normal renal and liver function, urinalysis and immunoglobulins. Rheumatoid factor, anti-nuclear antibody, and anti-neutrophil cytoplasmic antibodies were negative.

Microbiology examination of leg ulcer isolated a *Staphylococcus aureus* sensitive to ciprofloxacin

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Figure 1. Nodular lesions in the arm

which was instituted for 15 days.

Chest x-ray was normal.

Hands and feet radiographs revealed bone erosions in the first interphalangeal of the left hand (Figure 3).

The electromyography revealed mononeuritis multiplex.

Biopsy of a forearm nodule revealed a dermal lymphohistiocytic and plasma cell nodular infiltrate. Histochemistry staining (PAS, Grocott, Fite-Faracco) showed macrophages containing abundant *Mycobacterium leprae* (Figure 4).

The diagnosis of reactional lepromatous leprosy with skin lesions compatible with Lucio's phenomenon was made, and started rifampin 600 mg id, dapsone 100mg id and clofazimine 50mg id. This treatment continued for thirty days, with progressive amelioration of skin lesions, leg ulcers and arthritis. Rifampin was reduced to 600 mg per month, with dapsone 100mg id, clofazimine 50mg id and prednisolone 5mg id, maintaining a good response after six months of follow up.

Discussion

Leprosy is a chronic infectious disease caused by *Mycobacterium leprae*. The route of transmission



Figure 2. Leg ulcer with 12x2cm

remains uncertain and may be multiple: nasal droplet infection, contact with infected soil, and even insect vectors have been considered the prime candidates. In endemic countries 50% of leprosy patients have a history of intimate contact with an infected person, while for unknown reasons leprosy patients in nonendemic locales can identify such contact only 10% of the time.

The husband of this patient lived in Africa forty years ago during 5 years, and had leprosy in that



Figure 3. Hand x-ray. Periarticular osteoporosis in the metacarpophalangeal, joint space narrowing, geodes on the first and second metacarpo, and erosions in the first interphalangeal of the left hand.

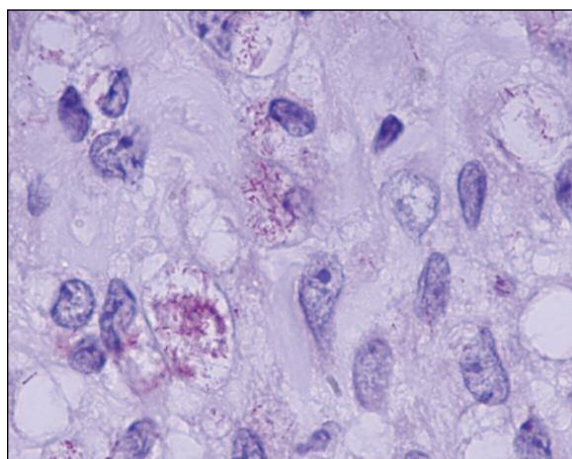


Figure 4. Subcutaneous nodule biopsy (Fite-Faracco staining): macrophages with abundant *Mycobacterium leprae*

period. He was treated and considered cured.

Lepromatous leprosy is characterized by a reduced immune response with a predominance of T CD8 cells and a large number of bacilli in the tissues. This form of leprosy has some clinical and serologic similarities with rheumatic diseases, such as erythematous macules, subcutaneous nodules, arthralgias, polyarthritis and sometimes rheumatoid factor, antinuclear antibody or anti-neutrophil cytoplasmic antibodies^{4,5}.

Lucio's phenomenon is an unusual reaction that occurs in untreated lepromatous leprosy, characterized by recurrent crops of large ulcerative lesions, particularly on the lower extremities, without significant general symptoms. These lesions are histologically characterized by ischemic necrosis of the epidermis and superficial dermis, and heavy parasitism of cells with acid-fast bacilli⁵.

Arthritis in leprosy can be acute or chronic. Acute arthritis is associated with reactional states and is characterized by a symmetric polyarthritis involving the knees, ankles, wrists, elbows, proximal interphalangeal, metacarpophalangeal and metatarsophalangeal, without radiographic abnormalities. The synovial fluid is inflammatory and sterile, and synovial biopsy can show lymphoplasmocytic infiltrate with bacilli. Chronic arthritis is the commonest form, and is characterized by an insidious symmetric polyarthritis involving the wrist, small joints of the hands and feet, and knees. Radiographs can rarely show bony erosions⁶. In most cases polyarthritis manifests concomitant to the skin lesions, although in some ca-

ses it can precedes the cutaneous manifestations, as in this clinical case, and have a slow progression⁷.

Vasculitis can be seen in Lucio's phenomenon and is characterized by endothelial proliferation, ischemic necrosis, mononuclear infiltrate and bacilli in small vessel walls^{4,5}.

In this case, the patient had chronic polyarthritis, with few bone erosions, without RF and subsequently developed painful subcutaneous nodules, ulcerative lesions in both legs and mononeuritis multiplex, in a non endemic country, mimicking rheumatic diseases.

We suspected of primary vasculitis, like polyarteritis nodosa, or vasculitis secondary to connective tissue disorders like rheumatoid arthritis. The biopsy was the main exam that allowed the correct diagnosis and treatment of leprosy, avoiding the institution of immunosuppressive drugs that would have led to high morbidity⁸.

The therapeutic regime instituted was based on the World Health Organization recommendations for the chemotherapy of leprosy. It classifies patients as multibacillary if they have six or more skin lesions, and as paucibacillary if they have fewer. The WHO recommends that paucibacillary adults should be treated with dapsone 100mg id and rifampin 600mg monthly for 6 months, and multibacillary adults be treated with dapsone 100mg and clofazimine 50mg daily unsupervised, and rifampin 600mg and clofazimine 300mg monthly supervised, for 1 to 2 years⁹.

In this case as the clinical presentation was a severe form of multibacillary, it was decided to start with rifampin 600mg daily during one month, since it is the only bactericidal drug, and maintain clofazimine 50mg/d during all the treatment, with gradual improvement.

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References

1. Britton WJ, Lockwood DNJ. Leprosy. *Lancet* 2004; 363: 1209-1219.
2. Moschella, SL. An update on the diagnosis and treatment of leprosy. *J. Am. Acad Dermatol* 2004; 51: 417-426.

3. Gibson T, Ahsan Q, Hussein K. Arthritis of leprosy. *Br J Rheumatol* 1994; 33: 963-966.
4. Albert D, Weisman MH, Kaplan R. The rheumatic manifestations of leprosy. *Medicine* 1980; 59:442-448.
5. Pardillo FE, Fajardo TT, Abalos RM et al. Methods for the classification of leprosy for treatment purposes. *Clin Infect Dis* 2007; 44: 1096-1099.
6. Yens D, Asters D, Teitel A. Subcutaneous nodules and joint deformity in leprosy – case report and review. *J Clin Rheumatol* 2003; 9: 181-186.
7. Alves Pereira HL, Euzébio Ribeiro SL, Sato EI. Manifestações Reumáticas da Hanseníase. *Acta Reumatol Port* 2008;33:407-414.
8. Genta M, Genta R, Gabay C. Systemic Rheumatoid Vasculitis: a review. *Semin Arthritis Rheum* 2006; 36: 88-98.
9. World Health Organization. WHO-recommended MDT regimens. World Health Organization. Available at <http://www.who.int/lep/mdt/regimens/en/>.

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