Panniculitis by leprosy: Case report of a diagnostic challenge

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ABSTRACT

Leprosy, or Hansen’s disease, is a chronic infectious-contagious disorder that affects skin and peripheral nervous system, caused by \textit{Mycobacterium leprae}, and remains as one of the most important neglected diseases. Panniculitis is an inflammatory disease of the subcutaneous adipose tissue that corresponds to a group of heterogeneous disorders which has clinical singularity. We report a case of an extremely unusual form of clinical manifestation of the disorder, panniculitis, as the only manifestation. The condition was solved with a combination of systemic corticosteroids and specific antibiotic treatment.

1. Introduction

Leprosy, or Hansen’s disease, is a chronic infectious-contagious disorder that affects skin and peripheral nervous system[1]. It is characterized as a high infectiousness and low pathogenicity illness, which advances slowly but can potentially generate severe disabilities and stigma. The condition is caused by \textit{Mycobacterium leprae} (\textit{M. leprae}), an obligate intracellular bacillus with tropism for macrophages and Schwann cells, transmitted by prolonged intimate contact with untreated bacilliferous individuals. Both nasal and skin secretions from these people can shed the bacterium into the environment. Leprosy presents an extensive variety of clinical, histological and immunological appearances, which are determined by the natural resistance of the host to invasion of \textit{M. leprae}[1,2].

Panniculitis is an inflammatory disease of the subcutaneous adipose tissue that consists to a group of heterogeneous disorders which has clinical singularity[3]. The clinical morphology presents as erythematous nodules generally located on the lower limbs.

2. Case report

A 27-year-old female patient was hospitalized for eye swelling and fever complaint that started about two months, on suspicion of facial cellulitis. She reported that, approximately 2 years ago, she had had cutaneous lesions in plaque (two in the left arm, one in the right arm and other in the left thigh), of hardened consistency associated with local erythema. Initial lesions reverted spontaneously with localized depression. About 2 months later, the patient presented pain and tough edema in the right periorbital region and in the right shoulder. It evolved with progressive worsening and companied by frequent episodes of fever and headache. In 15 days after admission, the periorbital edema led to ocular opening restriction. It has been used clindamycin and oxacillin, without clinical improvement or remission of fever.

Serological tests for HIV and hepatitis were negative. A negative ANF was performed. The erythrocyte sedimentation rate was above 100 (Figures 1 and 2).

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It was performed a tomography of the sinuses, revealing the thickening of the skin of the right hemiface associated to densification of the subcutaneous cellular tissue and adjacent muscular planes, extending from the right mandibular region to the homolateral periorbital region. The laboratory was normal, despite mild anemia. The search for BAAR in the lymph was negative.

Figure 1. Clinical appearance of the right periorbital region and the right shoulder, which was biopsied.

The initial histopathological evaluation evidenced epidermis without significant changes and dermis with dense perivascular, perianexial and perineural lymphohistiocytic infiltrate, with some plasma and multinucleated giant cells. Adipose tissue was scarcely represented and xanthomatous macrophages were noted in the deep dermis (probable replacement of the subcutaneous tissue by fibrosis) (Figure 3).

Posteriorly, additional staining with Fite-Faraco was performed, which resulted positive for BAAR (Figure 4).

The patient was treated with the pattern combined therapy instituted by World Health Organization, with rifampin, isoniazid and clofazimine, besides prednisone. It has been remission of the lesions described, with no recurrence until the present moment (Figure 5).

3. Discussion

Leprosy is one of the most important neglected diseases, causing the greatest number of permanent disabilities among the infectious disorders. In 2015, 210,758 new cases of leprosy were diagnosed in 136 countries and territories worldwide. In Brazil, at the same year, it has been detected 26,395 new cases of leprosy, which corresponds to the second highest number of cases in the world and represents 13% of the global new cases. Among the new cases detected, 67.9% (17,913 patients) were multibacillary[5]. It is still relevant that Brazil is the only country in America that has not been able to control leprosy (defined as prevalence < 1 case per 10,000 inhabitants)[2].

Ridley and Jopling[6], in 1962, based in the many clinical manifestations of leprosy, subdivided the disease into the following groups: tuberculoid (T), borderline tuberculoid (BT), borderline (B), borderline lepromatous (BL), and lepromatous (L). For diagnosis and definition of the treatment regime with polychemotherapy, World Health Organization recommended an additional classification, that are based on the number of cutaneous lesions, per the following criterion: paucibacillary cases (PB) with up to five skin lesions;
multibacillary cases (MB) with more than five skin lesions[7,8].

Leprosy presents, clinically, in several manners. The initial lesion, the indeterminate form, corresponds to defined hypoesthesia associated or not with a visible lesion - the most common is one or some hypopigmented macules of ill-defined borders. The tuberculoid form presents cutaneous lesions with pronounced borders, single or small, and asymmetrically distributed through the integument. It presents, when macula, as hypochromic or erythematous lesion, delimitated by micropapulas, and, when plaque, as erythematos or coppery lesion, diffusely infiltrated, or with central tendency to planing. The lepromatous form involves diffusely extensive areas of the tegument, multiple nerve trunks, and even other organs. It presents with lesions marked erythema, infiltration, light skin, orange peel type, and overlapping areas of papules, nodules and tumors. The borderline form is characterized by immunological instability and, within the possible spectrum of cutaneous lesions, we can observe from macules, erythematous, in light skin, to hypochromic, in dark skin, which sometimes assumes coppery tonality, being also common the presence of papules, nodules, tumors and plaques[1,7].

The patients of Hansen’s disease may, still, develop hypersensibility host reactions. These are known as leprosy reactions. There are two kinds of them. Type 1 or reversal reaction is a late cellular hypersensibility reaction type IV. Previous lesions become gradually more swelled, reddened and even ulcerate. Type 2 reaction or erythema nodosum leprosum (ENL) is an acute inflammatory condition involving TNF-alfa and immune complex mediated immune response with infiltration of Th2-cells[8].

ENL presents as a multisystem disease, classically represented by painful, reddish, confluent papules or subcutaneous nodules, accompanied by systemic symptoms such as fever, prostration and leukocytosis. Being an immunocomplex mediated disease, it can have renal, ocular manifestations and in several internal organs. It represents a compelling cause of morbidity and physical disabilities[9].

In histopathology, ENL presents polymorphonuclear lymphocytic infiltrate in the deeper layers of the dermis, and may involve subcutaneous tissue, being a cause of panniculitis. At this juncture, the name erythema nodosum is inadequate, since leprosy does not present the sepal pattern characteristic of that disorder[8,10].

Pepler et al.[8] reviewed the clinical and histopathological findings in 19 cases of so-called ENL, and believed that it existed an unlike clinical manifestation that they preferred to call panniculitis nodosa leprosa (PNL), which was misnamed over time. It was characterized by showers of dusky red nODULES, 0.5–2 cm in diameter, which can coalesce to form plaques. It affects areas that are seldom or never affected in erythema nodosum, as the face, limbs, and, less often, trunk. Recurrence is the rule. In the histopathological findings, PNL is characterized for the involvement of subcutaneous tissue and, in chronic cases, evidences of its fibrous replacement[8].

Ramos e Silva[11] reported two cases that presented similar presentation to the related case, mainly the reactive sclerosis and the development of interstitial fibrosis in these patients. They assigned the title of “Hypodermite Nodular Lepromatosa Recidivante”[11].

Walzer and Nataro[12] described a case of a patient diagnosed with leprosy that presented fever and nodules on the extensor aspects of the legs. The histologic examination showed granulomatos reaction extending into the subcutaneous. After a review of the literature, they preferred to name this case a lepromatous panniculitis[12].

M. leprae can be, therefore, a cause of panniculitis, even in its classic form, with painful subcutaneous nodules and possibly associated vasculitis - cases in which the lesions may be accompanied by necrosis. The evaluation of the subcutaneous involvement, and not just the dermis, is clinically indistinguishable and, essentially, histopathological, and could be evidenced in the case by the replacement of the adipocytes by fibrosis. That said, it could be interpreted as an atypical form of ENL or PNL, depending on the reference.

Despite the treatment preconized for leprosy erythema nodosum was thalidomide, some situations demand the use of systemic corticosteroids[2]. In this specific case, the decision of how adjuvant therapy would be used was difficult. We decided that, as the component of inflammation predominated and the clinical was compatible with panniculitis more than ENL, it was opted for prednisone besides the pattern combined therapy. The response was complete and there were no relapses.

Conflict of interest statement

We declare that we have no conflict of interest.

References