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CASE REPORT

Hansen's disease, a condition to be remembered

Hanseníase, uma doença a não esquecer

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Abstract

Leprosy is a chronic granulomatous disease caused by *Mycobacterium leprae*, which mainly affects the skin, mucous membranes, and the peripheral nervous system. Despite the marked global reduction in the prevalence of the disease, the latest available data, referring to 2019, report six cases of leprosy in Portugal. In this report, we describe a case of borderline tuberculoid leprosy that was diagnosed initially as dermatophytosis and psoriasis. With the increase in international travelling, the disease can arise in any country. This case highlights the value of clinical suspicion in countries where the disease is less prevalent, particularly in individuals from endemic regions.

Keywords: Leprosy/diagnosis. Leprosy. Tuberculoid. *Mycobacterium leprae*.

Resumo

A hanseníase é uma doença granulomatosa crónica, causada pelo *Mycobacterium leprae,* que afeta principalmente a pele, as mucosas e o sistema nervoso periférico. Apesar da acentuada redução global da prevalência da doença, os últimos dados disponíveis, referentes a 2019, reportam 6 casos de hanseníase em Portugal. Neste artigo, descrevemos um caso de hanseníase tuberculóide (BT) limítrofe que foi inicialmente diagnosticada como dermatofitose e psoríase. Com o aumento das viagens internacionais, a doença pode surgir em qualquer país. Este caso destaca o valor da suspeita clínica em países nos quais a doença é menos incidente, particularmente em indivíduos provenientes de regiões endémicas.

Palavras-chave: Lepra/diagnóstico. Lepra. Tuberculoide. *Mycobacterium leprae.*

Introduction

Leprosy is a chronic granulomatous disease that mainly affects the skin, mucous membranes, and the peripheral nervous system. It is caused by *Mycobacterium leprae* transmitted from person to person through inhalation of infectious droplets and has a long incubation period[1]. The condition has a worldwide distribution, but it is found most frequently in Africa, Brazil, India, and Southeast Asia[2]. Despite the global reduction in

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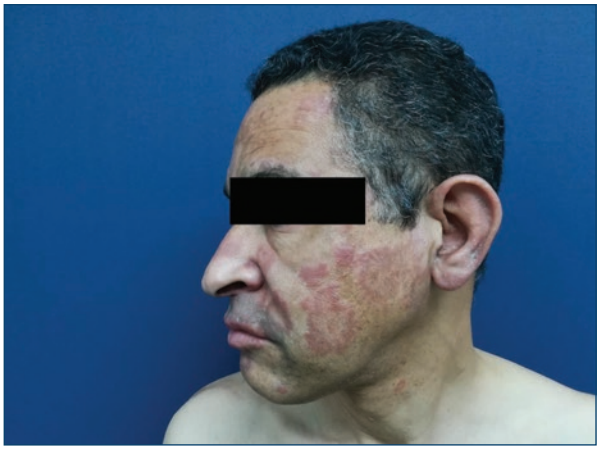

Figure 1. On the face ill-defined, homogenous, erythematous papules, and plaques, with slightly scaly edges measuring 1-3 cm, giving an edematous aspect, but without erasing the wrinkles and skin ridges.

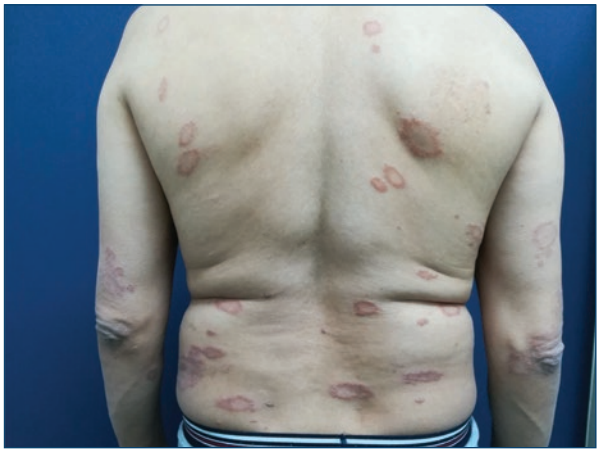

Figure 2. Erythematous/coppery colored plaques, measuring between 2 and 5 cm, with well-defined and elevated slightly scaling borders and central clearing on the trunk and limbs, but without atrophy.

its prevalence, six cases of leprosy were reported in Portugal in 2019, making it a differential diagnosis to keep in mind[3]. In this report, we describe a case of borderline tuberculoid (BT) leprosy that was initially misdiagnosed as dermatophytosis and psoriasis.

Case report

A previously healthy 48-year-old man, born in Brazil (Tocantins) and resident in Portugal for a year, presented to our hospital in March of 2021 due to skin lesions present for 10 months. Lesions initially appeared on the hands and feet as small erythematous macules and papules, with subsequent generalization to the trunk, remaining portion of the limbs, and face. He complained of dysesthesia, numbness, and electric shock sensation in the distal region of the limbs, and difficulty in distinguishing between hot and cold water over the past 6 months. The patient was otherwise well and denied the introduction of new drugs, changes in personal hygiene habits or contact with animals before the appearance of the lesions. From the family medical history, he referred the diagnosis of leprosy of his mother about 4 years ago.

Three months before his primary health care physician assumed the diagnosis of dermatophytosis prescribed oral and topical antifungals for 1.5 months and as there was no improvement, topical corticosteroids were initiated for a possible diagnosis of psoriasis.

At admission in our hospital, we observed a generalized maculopapular dermatosis mostly with isolated annular lesions affecting predominantly the face, trunk, and limbs, including palms and plants. On the face, mainly at malar, frontal and temporal region, and lesions were characterized by ill-defined, homogenous, erythematous papules, and plaques, with slightly scaly borders measuring 1–3 cm, giving an edematous aspect, but without erasing wrinkles and skin ridges (Fig. 1). The ears and scalp were spared. On the neck, trunk, and limbs, we observed, 2–5 cm, erythematous/coppery colored plaques with well-defined and elevated slight scaling borders and central clearing, but without atrophy (Fig. 2 and 3). At the dorsum of the hands, he had erythematous papules with little infiltration and scaling and in the palms and plants, diffuse ill-defined, and erythematous-violet macules, which occupied almost the whole plantar region. In total, there were more than 30 lesions.

The fine sensitivity assessed by a cotton swab was decreased inside the lesions and preserved outside. The cervical major and cubital nerves were palpable. We assumed BT leprosy as the most likely diagnosis. The slit-skin smear collected from the periphery (elevated border) of a skin lesion and ear lobe was negative. Anatomopathological examination revealed a normal epidermis and a dermal granulomatous reaction, consisting of epithelioid granulomas predominantly in the papillary dermis, with foamy epithelioid histiocytes and frequent lymphocytes, but also with deep periadnexal and perivascular involvement (Fig. 4 and 5). The histochemical study with Fite-Faraco techniques did not show micro-organisms. The immunohistochemistry study with antibodies to S100 and

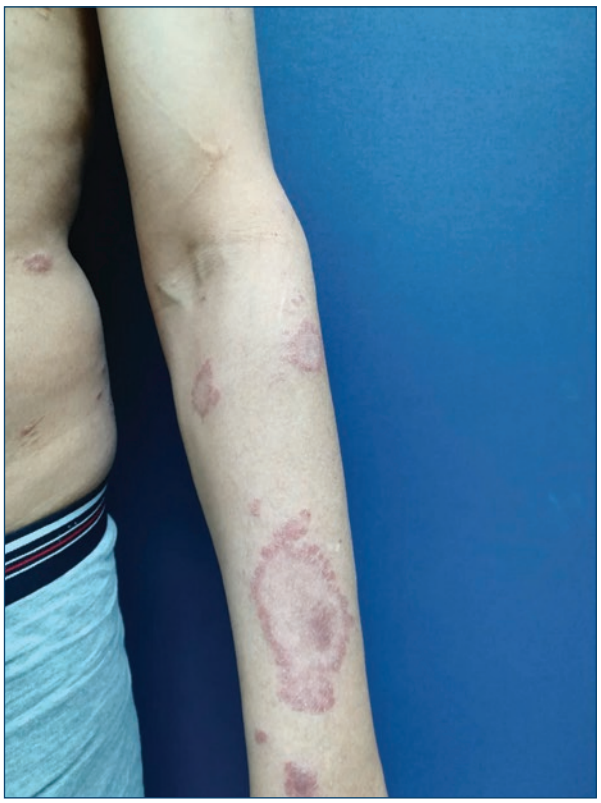

Figure 3. Detail of one of the annular lesions in the left forearm.

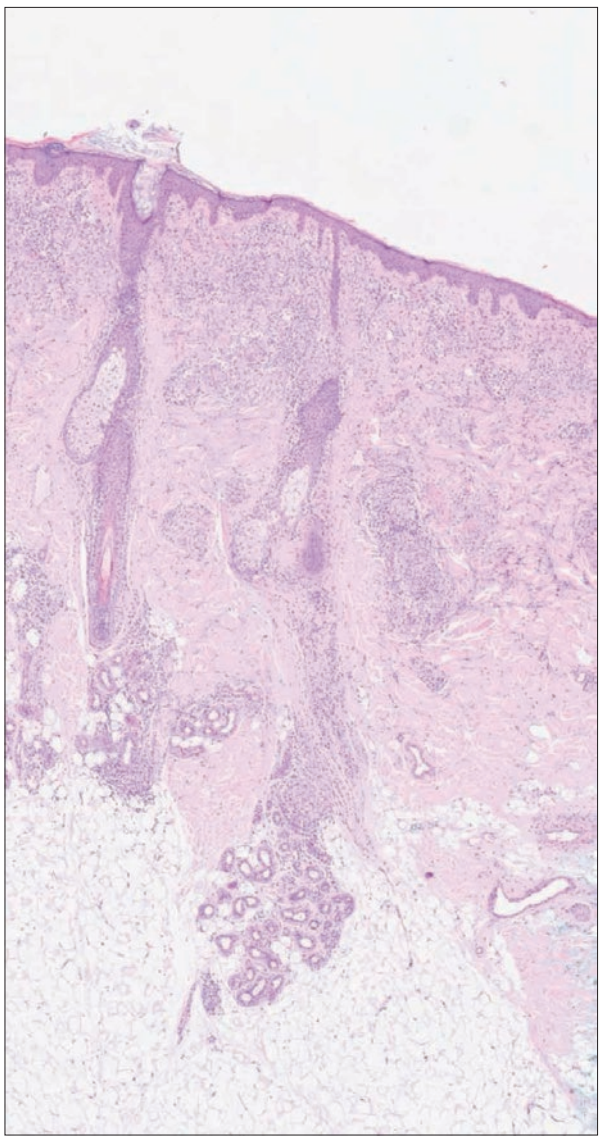

Figure 4. Histopathology of the skin biopsy with a dermal granulomatous reaction, predominantly superficial, with periadnexal and perivascular involvement (H&E ×40).

neurofilaments highlighted occasional nerve fibers, rarely in relation to granulomas. The polymerase chain reaction (PCR) test was negative. The nerve conduction study and electromyography were compatible with sensory and motor polyneuropathy, of axonal pattern. Therefore, the diagnosis of multibacillary leprosy was assumed based on the clinical, neurological findings, and epidemiology.

Treatment with dapsone 100 mg/day, clofazimine 50 mg/day, rifampicin 600 mg/month, and clofazimine 300 mg/month was started. Prednisolone 60 mg/day plus pantoprazole 40 mg/day was also initiated for neurological damage prevention. The patient was followed in dermatology, infectiology, and neurology consultations. There was a significant improvement of neurological manifestations, and resolution of the cutaneous lesions with post-inflammatory hyperpigmentation.

Discussion

Leprosy has not been fully eradicated and still represents a public health problem in modern societies. In 2019, more than 200,000 cases of leprosy were diagnosed in 118 countries worldwide[2]. The WHO Global Strategy for Leprosy 2021–2030 follows these epidemiological modifications. Contrary to previous strategies aimed at "eliminating leprosy as a public health problem," this new approach concentrates on disrupting transmission and reaching zero autochthonous cases. However, with increasing migrating fluxes and international travelling, the disease can present in any country[2]. As such, it is still essential to preserve the ability to diagnose the disease and keep surveillance in lower incidence countries (such as Portugal), to avoid misdiagnosis. This case highlights the value of clinical suspicion, particularly in subjects whose

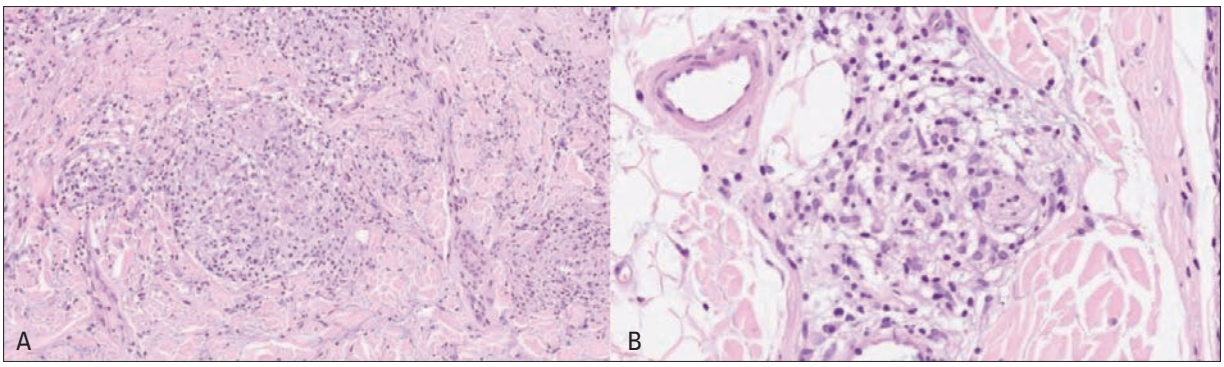

Figure 5. A: dermal granulomas made up of epithelioid histiocytes with pale cytoplasm, accompanied by lymphocytes and without necrosis. **B:** one involving a nerve bundle (H&E ×100).

original country is endemic for leprosy and manifest typical or atypical cutaneous lesions.

In 2018, the global leprosy detection rate was 1.93 cases/100,000 inhabitants. Countries such as Indonesia, India, and Brazil were responsible for 79.6% of reported cases[4]. In the particular case of Brazil, in the same year, the detection rate of new cases was 12.94 cases/100,000 inhabitants (about 93% of the total cases reported in the Americas)[4,5]. Despite the decrease in the number of cases in recent years in Brazil, some regions, such as the North, Center-West, and Northeast (Mato Grosso, Tocantins, Rondônia, Pará, and Maranhão) still have high incidence rates, and the patient reported lived recently in a highly endemic region of Brazil[6]. It is also relevant to remind that *M. leprae* has a prolonged incubation period that could be larger than two decades[7].

At present, the diagnosis of leprosy is established through clinical history and physical examination and, when possible, by slit-skin smear[8,9]. Based on the WHO, clinical diagnosis is made with one of the following three characteristics: (i) A pale (hypopigmented) or reddish skin patch with permanent sensitive loss; (ii) an enlarged or thickened peripheral nerve with loss of sensation; and (iii) the presence on the skin smear of alcohol-resistant bacilli[10].

Leprosy can be classified according to the Ridley-Jopling and WHO classifications, which define the different forms of disease presentation and guide its diagnosis and treatment. The Ridley-Jopling classification includes clinical manifestations, bacteriological index, and histopathological characteristics, whereas the WHO classification considers the number of skin lesions or the bacteriological evaluation index[11,12]. According to the WHO 2018 guidelines, no additional tests are recommended beyond the examination of slit skin smear[1]. However, a skin biopsy can assist in the classification of leprosy and evaluate the treatment response. The PCR test can also be performed to support clinical diagnosis[1]. Nevertheless, in tuberculoid forms, skin biopsy usually demonstrates very few (or absent) acid-fast bacilli and the positive PCR rates decrease to a sensitivity of 50%[1].

Regarding treatment, the present WHO guidelines recommend the use of dapsone, rifampicin, and clofazimine for 6 and 12 months to treat paucibacillary and multibacillary leprosy, respectively[13]. The main cause of morbidity seen in leprosy is peripheral neuropathy, which is responsible for most of the deformities and deficiencies seen in leprosy[14,15]. Nerve damage occurs at the level of motor, sensory, and autonomic fibers. There is a subacute/chronic inflammatory infiltrate of epithelioid cells or macrophages in the epineurium, endoneurium, and perineurium[16]. This causes progressive impairment of the neural fibers, with the subsequent nerve destruction[17]. Treatment at an early stage is extremely important to cure the disease and prevent sequelae. Corticosteroids are indicated for the treatment of impaired nerve function and leprosy reactions[18]. Usually, leprosy neuropathy responds to prednisone, tapering being slow over 4–6 months. Early treated patients can recover 60% of neural impairment and nerve function[19]. Nevertheless, most recent reviews report a doubtful role for prophylactic corticosteroids in preventing nerve involvement[19].

What does the study add?

In this report we discuss a case of borderline tuberculoid leprosy (BT) that was initially diagnosed as dermatophytosis and psoriasis, despite all neurological complaints and maternal history of leprosy. Diagnosis of leprosy needs to be remembered even in countries

like Portugal which have a low disease prevalence, particularly in patients coming from endemic regions.

Authors' contributions

E. Freitas, J. Reis, A. Coelho, G. C. Velho, and M. Selores had the idea for the article, performed the literature search and data analysis, and drafted and critically revised the work.

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Conflicts of interest

The authors have no conflicts of interest to declare.

Ethical disclosures

Protection of people and animals. The authors declare that for this investigation no experiments were carried out on humans and/or animals.

Data confidentiality. The authors declare that no patient data appears in this article.

Right to privacy and written consent. The authors declare that no patient data appears in this article.

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