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Case presentation

Wade histoid leprosy revisited

Virgínia Coelho de Sousa, André Laureano, Jorge Cardoso

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Department of Dermatology and Venereology, Hospital de Santo António dos Capuchos – Centro Hospitalar de Lisboa Central, Lisboa, Portugal

Correspondence:

Virgínia Coelho de Sousa, MD
Hospital de Santo António dos Capuchos – Centro Hospitalar de Lisboa Central, Lisboa, Portugal
Alameda Santo António dos Capuchos, 1169-050 Lisboa, Portugal
virginiacoelhodesousa@gmail.com
+351926826029

Abstract

An 18-year-old man presented with a 4-year history of erythematous patches on the trunk, followed 2-years later by multiple nodules, mostly located on the limbs, and distal paresthesias. Two close contacts were treated for leprosy during his childhood. Histopathological examination revealed a histiocytic infiltrate with acid-fast bacilli on Ziehl-Neelsen stain. The slit-skin and nasal smears showed numerous acid-fast bacilli. The correlation between clinical, epidemiological, histopathological, and microbiological features allowed the diagnosis of lepromatous leprosy, histoid variant. Multidrug therapy as recommended by the WHO was initiated. A rapid and sustained improvement was seen. Histoid leprosy is a rare manifestation of lepromatous leprosy, first described by Wade in 1960. Since then few cases have been reported, the majority of them from countries with a high prevalence of the disease. Early recognition and treatment are of most importance to prevent neurological disabilities and achieve epidemiological control.

Keywords: Hansen's disease, histoid Leprosy, *Mycobacterium leprae*

Introduction

Leprosy or Hansen's disease is a chronic granulomatous infectious disease caused by *Mycobacterium leprae,* affecting the skin and the peripheral nervous system in the majority of cases. Histoid leprosy (HL) is a rare manifestation of multibacillary leprosy presenting with disseminated dermatofibroma-like papules and nodules. The clinical, histopathological, and bacteriological findings are characteristic. Most of the cases have been related to dapsone resistance in the context of long-term monotherapy. *De novo* cases, not associated with previous anti-leprosy treatment, have been less frequently reported.

Case synopsis

An 18-year-old man presented to our clinic with a 4-year history of asymptomatic coppery patches on the trunk, followed 2-years later by multiple papules and nodules located on the extremities and face. He also reported malaise, hand and foot paresthesias, and recurrent episodes of epistaxis. The patient lived with his sister and uncle in Minas Gerais, Brazil since he was 3-years-old. These relatives were then being treated for leprosy. Currently, he has been living in Lisbon, Portugal for 10 years.

Physical examination disclosed multiple, firm, round and well-demarcated nodules with a smooth bright surface, "dermatofibroma-like." The lesions had a symmetric distribution and some were clustered. An ulcerated nodule was present. Large coppery patches with vague edges and smooth surface were also observed on the trunk.

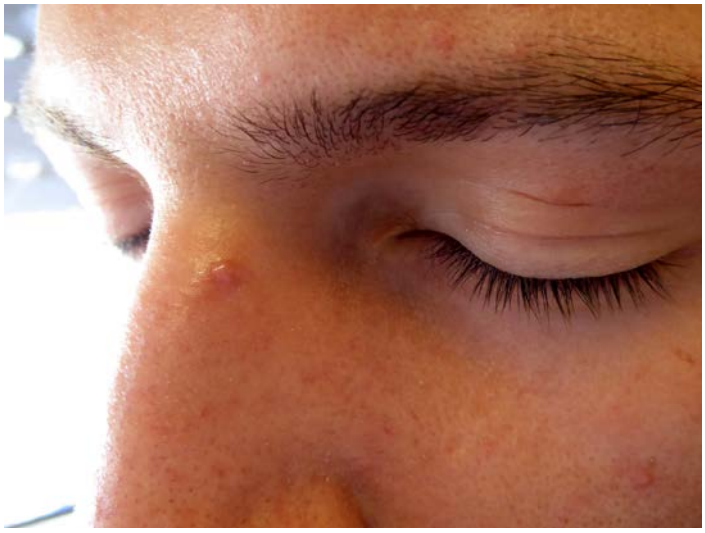


Figure 1. Firm, round and well-demarcated papule located on the nose

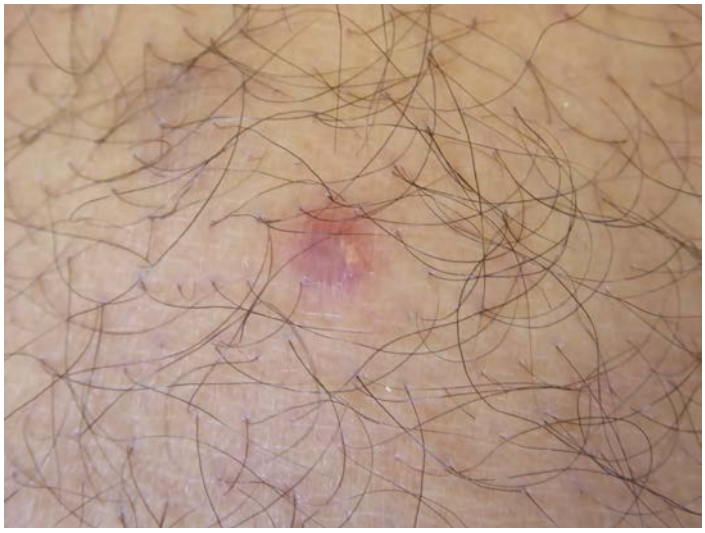


Figure 2. "Dermatofibroma-like" nodule on the right arm

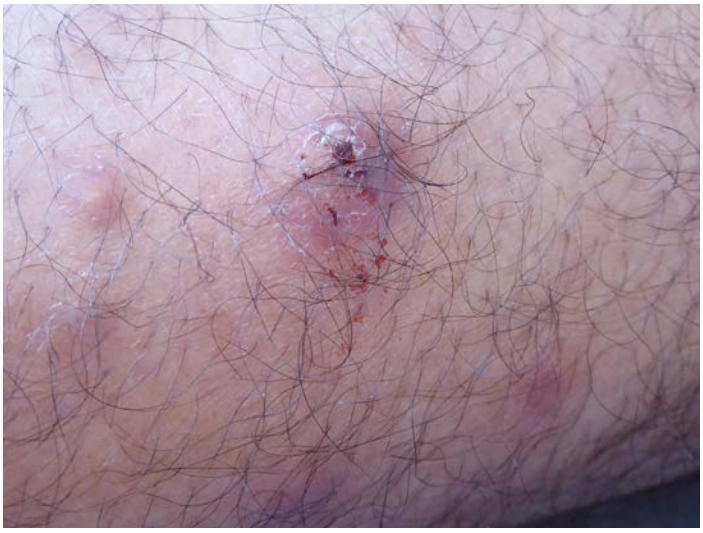


Figure 3. An ulcerated nodule was observed on the right leg.

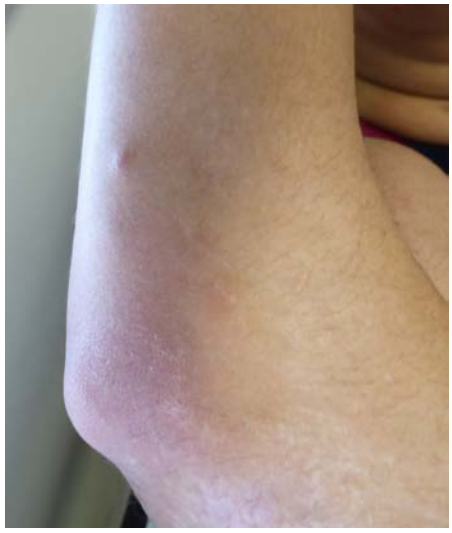


Figure 4. Histoid lesions show particular tropism over bony prominences.

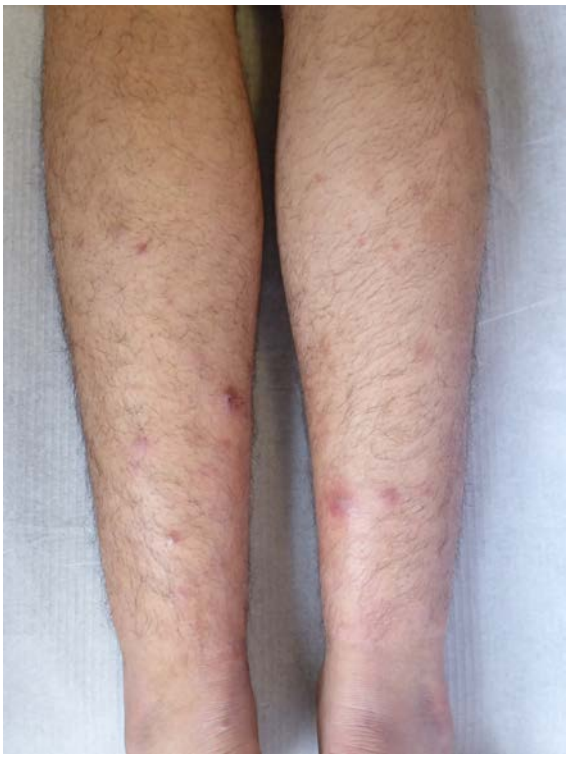


Figure 5. Multiple nodules, some grouped in crops, located on the legs

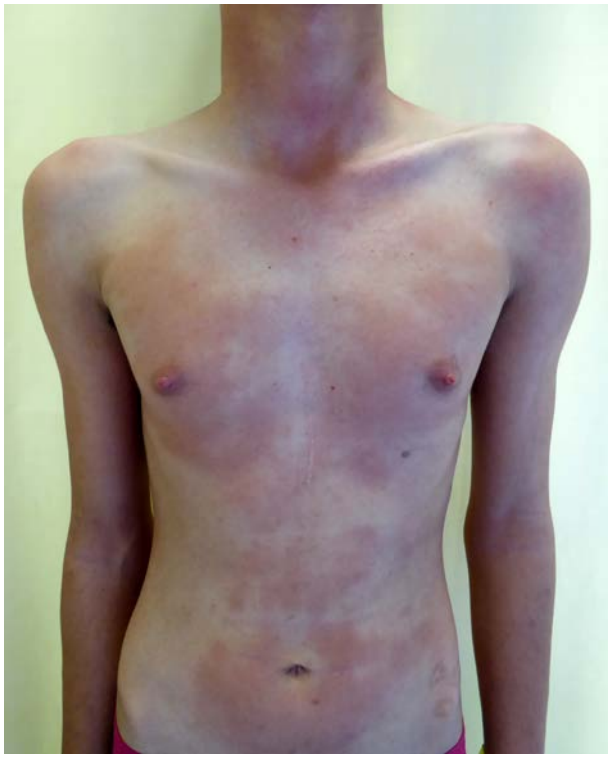
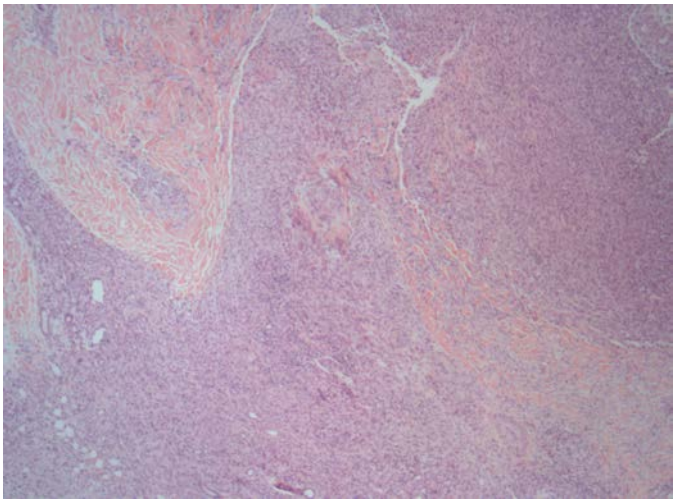


Figure 6. Coppery patches with vague edges and smooth surface were observed on the trunk.

Ulnar nerves were enlarged and decreased distal pain and tactile sensation of the limbs was also noted.

A clinical diagnosis of lepromatous leprosy was then considered, according to the Ridley-Jopling classification. Biopsy specimens from a nodule and a patch were taken. The histopathological evaluation from the nodule revealed an infiltrate of spindle shaped cells arranged in parallel bundles, similar to a dermatofibroma presentation. Multiple acid-fast bacilli were observed along the long axis of spindle cells on Ziehl-Neelsen staining. Nerve involvement was also seen.



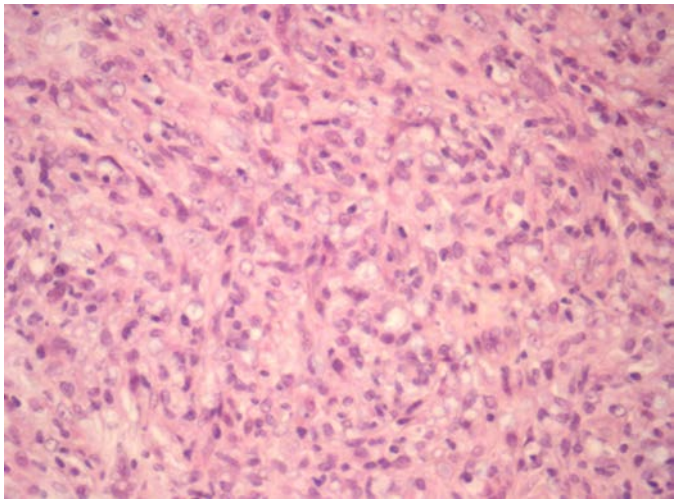


Figure 7. Histopathological evaluation from the nodule revealing an infiltrate of spindle shaped cells arranged in parallel bundles (Hematoxylin and Eosin, x200) **Figure 8.** Spindle shaped cells arranged in parallel bundles, similar to a dermatofibroma presentation (Hematoxylin and Eosin, x400).

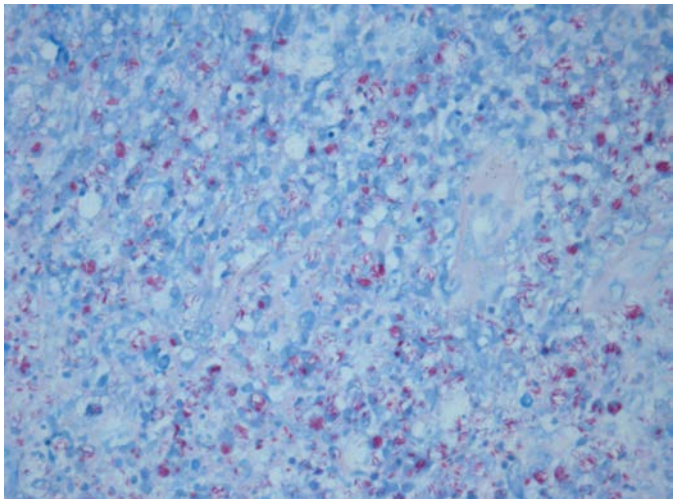
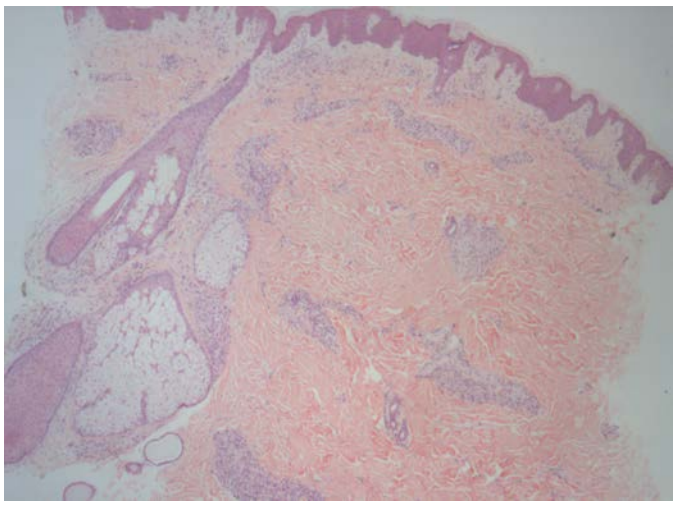


Figure 9. Multiple acid-fast bacilli along the spindle cell long axis on Ziehl-Neelsen stain (x200).

The histopathological examination from the patch also disclosed spindle cells in the deep dermis, surrounding nerves, and adnexal structures. Bacilli load was less pronounced on Ziehl-Neelsen staining.



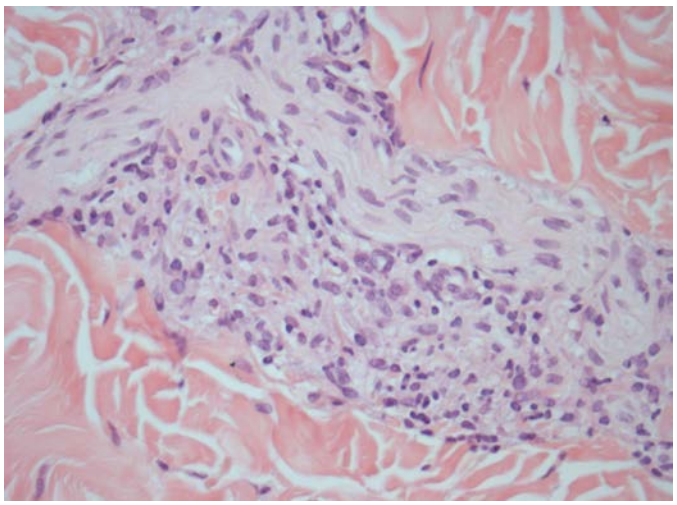


Figure 10. Histopathological examination from the patch disclosed spindle cells in the deep dermis (Hematoxylin and Eosin, x200).
Figure 11. Spindle cell infiltrate surrounding a nerve (Hematoxylin and Eosin, x400)

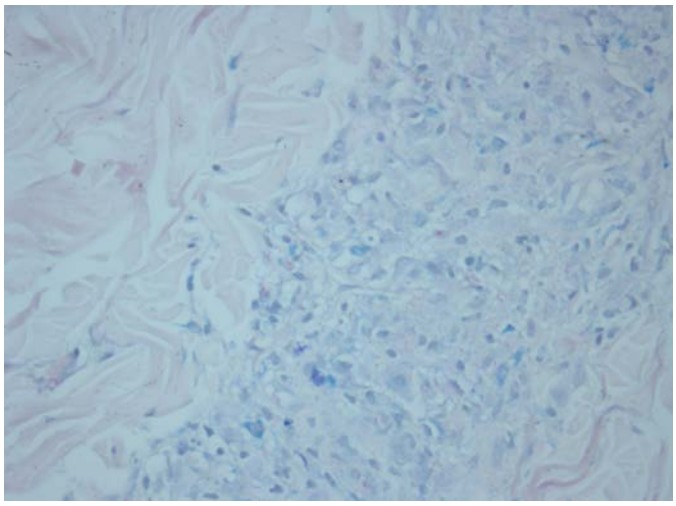
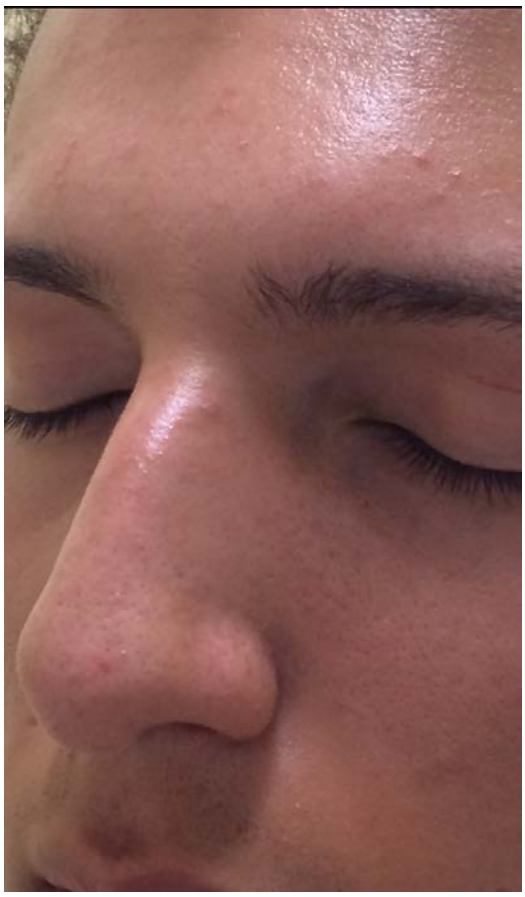


Figure 12. Ziehl-Neelsen stain from the patch biopsy specimen with less pronounced infiltrate of acid-fast bacilli (x200)

The nasal and slit-skin smears confirmed a multibacillary form of leprosy. Blood analysis was unremarkable, except for a slight increase in erythrocyte sedimentation rate.

Treatment with multidrug therapy (MDT), as recommended by the World Health Organization, was started. The patient was treated for 12 months with rifampicin 600 mg once-monthly; dapsone 100 mg daily; clofazimine 300 mg once-monthly, and 50 mg daily. Rapid and sustained clinical improvement was seen after one month of treatment. Post-inflammatory and clofazimine-associated hyperpigmentation were observed. No other drug-related adverse effects were reported.



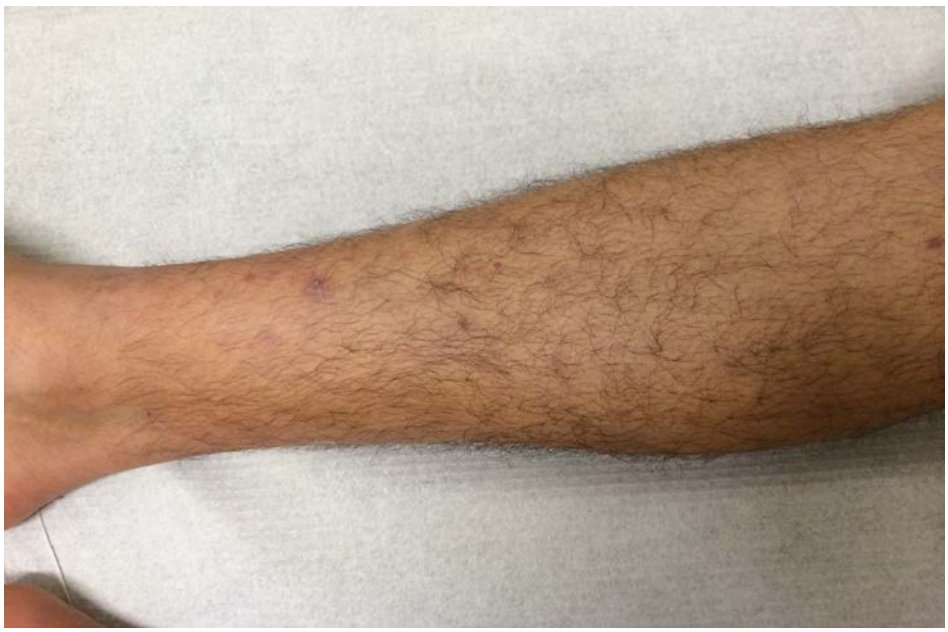


Figure 13. Rapid clinical response after one month of therapy; clofazimine-related hyperpigmentation was also observed. **Figure 14.** After one month of therapy, lower limb lesions showed dramatic improvement.

Smear negativity was also achieved. All cohabitants were observed. None were suspicious for the disease.

Discussion

In our case, the diagnosis of HL was supported by the finding of several typical features: cutaneous nodules, hypoesthesia, and peripheral nerve enlargement. The dermal histiocytic infiltrate exhibited acid-fast bacilli with a high bacillary index.

Wade HL is a rare manifestation of lepromatous leprosy. The first description of this variant, also known as “histoid leproma,” was made by Wade in 1960 [1]. In the era of long-term dapsone monotherapy, HL was considered the result of drug resistance and a manifestation of disease relapse [2]. After the introduction of MDT in 1981, HL became even less frequently observed. Most of the cases were reported from India, a high-prevalence country for the disease [3]. *De novo* HL, not related to previous anti-leprosy therapy, has been described only in few cases. We reported a rare case of *de novo* HL in a Brazilian patient now living in Portugal.

The pathogenesis of this unusual variant is not well defined. According to Kaur, its expression is probably related to the interplay between genetic and immunological factors [2].

Aside from the typical clinical presentation, as seen in our patient, nodule ulceration can be a less common feature [2]. Ulceration has been associated with transepidermal elimination of *Mycobacterium leprae*, with a postulated role in disease transmission [4]. HL favors the buttocks, arms, back, and face, with a special tropism for bony prominences, like elbows and knees.

Clinical and histopathological features similar to dermatofibroma explain the “histoid” designation [5]. Clinical differential diagnoses also include xanthoma, neurofibroma, molluscum contagiosum, and cutaneous metastasis.

Owing to its rarity, there are few studies regarding HL. A retrospective study from India revealed an incidence of 1.8%, among 2150 patients diagnosed in a 15 years interval [2]. There was a male predominance with a male/female ratio of 5.7:1. *De novo* histoid leprosy occurred in only 12,5% of the patients.

MDT is also the recommended therapeutic scheme in HL. Rifampicin, dapsone and clofazimine are given for a 12 month period or until smear negativity is achieved [6].

Unlike other forms of leprosy, numerous long and well-preserved acid-fast bacilli are seen, explaining the consistency of high bacterial indexes in slit-skin and nasal smears (5+/6+) observed in this variant [7, 8]. Therefore, early recognition and treatment of HL are of utmost importance to achieve spread control of the infection [9]. In non-endemic areas this can be particularly problematic, as clinicians are less aware of this entity. Dynamic migration flows from endemic areas makes leprosy a diagnosis to consider in the appropriate clinical and epidemiological settings.

Conclusion

HL is a rare variant of lepromatous leprosy associated with a high bacillary index, thus representing a potential reservoir for the infection. Hence, its early recognition and treatment are imperative even in non-endemic countries.

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