

Case Report

Wade's histoid leprosy in a 14-year-old teenage boy

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Abstract

Wade's histoid leprosy (HL) is a rare variant of multibacillary leprosy, with characteristic clinical, immunologic, histopathologic, and bacteriologic features. It is associated with resistance to sulfa drugs or polychemotherapy and is rarely observed in patients who have not undergone prior treatment. Clinically, HL resembles keloid or dermatofibroma. Furthermore, HL is rare in children and is difficult to diagnose even by experts. This report describes a case of HL in a 14-year-old Brazilian boy, who presented with multiple nodular and tumor-like lesions, simulating keloids. He had not undergone prior treatment with anti-leprosy drugs, which accentuates the relevance of this case report.

Keywords: Histoid leprosy. Keloid. Multibacillary leprosy.

INTRODUCTION

Histoid leprosy is a very rare clinicopathological variant of leprosy, which is very difficult to diagnose due to different clinical and histopathological findings that mimic a fibromatous disorder. Histoid leprosy is a very rare clinicopathological variant of leprosy in childhood. It is important that health professionals recognize atypical leprosy presentations in order to perform appropriate treatment and to prevent physical damage and psychological suffering in these children.

CASE REPORT

A 14-year-old male patient, resident in Planaltina de Goiás/GO, presented with multiple keloid-simulating nodules, distributed over the left inner forearm (**Figure 1**), of 2-years duration. Subsequently, the face and lower limbs were involved. There was no history of prior treatment with anti-leprosy drugs and he denied any family history of leprosy.

Upon neurological examination, a thickening of the ulnar nerves without signs of neuritis and bilateral plantar hypoesthesia with a disability degree of 1 were noted. The auricular, radial, median, posterior tibial, and fibular nerves did not exhibit signs of thickening or pain, while the muscular strength of the upper and lower limbs was preserved, with no change in palmar sensitivity.

A histopathology examination revealed a lesion compatible with a dermatofibroma. The patient refused taking any drugs or surgical treatment at any health center or public or private hospitals where he had sought prior assistance, until he was referred to our dermatology department. Following examination at our department, the patient was diagnosed with multibacillary leprosy, Wade's histoid variant.

The differential diagnosis included neurofibroma, leishmaniasis, and dermatofibroma. Bacteriologic examination of slit skin smear revealed a bacteriologic index of 4,25+. Histopathology of the nodules showed a nodular and well-circumscribed proliferation of spindle-shaped histiocytes, arranged in a storiform pattern in the upper dermis (**Figure 2**). Fite-Faraco stain demonstrated cells packed with numerous acid-fast bacilli (**Figure 3**). Based on these features, the diagnosis of histoid leprosy (HL) was confirmed, and the patient was treated with multidrug therapy (MDT). At the end of the 12-month treatment period, a partial reduction was noted in the nodules and in the bacilloscopy result indicators; treatment was deemed unsatisfactory. Thus, the decision was taken to prolong treatment with MDT for up to 24 months. The patient recently completed 16 months of treatment, showing progressive regression of the nodules.

DISCUSSION

Histoid leprosy is a rare type of multibacillary leprosy (MB), with specific clinical, histopathologic, bacteriologic, and immunological features¹. It was described by Wade in 1963 and it is associated with resistance to sulfa drugs or polychemotherapy. It is rarely observed in patients who have not undergone prior treatment²⁻⁴, which reinforces the importance of this case report.

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The etiopathogenesis of HL is not well understood⁵. However, it has been suggested to occur as a result of the action of drug-resistant mutant bacilli, or due to a decrease in cellular systemic immunity, with corresponding increase in local cellular and humoral immunity⁶. Efforts to restrict the disease or its spread are based on these findings⁵.

The average age of infection is between 21 and 40 years⁷; HL rarely occurs in childhood⁷.

Clinically it is characterized by cutaneous and/or subcutaneous nodules and papules, which are firm, skin-colored to yellowish-brown, emanating from an apparently normal skin^{6,7}. These nodules resemble keloid or dermatofibroma. The lesions are usually located on the posterior and lateral areas of the arms, buttocks, thighs, dorsum of the hands, lower part of the back, and over the elbows and knees⁶⁻⁹. The palms and soles are usually not affected¹⁰. The keloid-simulating lesions in this patient highlight the clinically compatible nature of this disease with the histoid form.

Histoid leprosy, especially presenting as keloids and dermatofibroma, clinically simulates neurofibroma, xanthoma, sarcoidosis, cutaneous metastasis, diffuse cutaneous leishmaniasis, lobomycosis⁵, molluscum contagiosum⁷ and a papulonodular variant of secondary syphilis¹⁰.

The circumscribed nature of the lesion, the predominance of spindle-shaped cells, and the morphology of *histoid* bacilli are distinctive features in the histopathology of HL^{8,9}. The lesion consists of fusiform histiocytes arranged in a whorled, crisscross, or storiform pattern⁹. These histiocytes resemble fibroblasts and the morphology of the lesions mimics neoplasms⁸, like dermatofibroma and neurofibroma. The acid-fast bacilli are longer than ordinary lepra bacilli and they are not found in globi formation^{8,9}. Furthermore, the Wade variant is essentially underpinned by the demonstration of alcohol-acid-resistant bacilli that are isolated and grouped together.

The histopathologic similarity of a dermatofibroma-like lesion explains why the uncritical observation of some practicing pathologists or their unfamiliarity with leprosy biopsies may result in misdiagnoses, such as in the present case. This is common in both private and public pathology laboratories.

Once the clinical appearance of a histoid leproma resembles a dermatofibroma or keloid, many specialist physicians fail to suspect HL. This phenomenon has increasingly been observed recently, since leprosy studies have been neglected by several dermatologists.

While specialists encounter difficulties in diagnosing leprosy, it is a more serious challenge for professionals who operate within the primary care system. This underlines the important role of renowned centers, in training and enhancing the awareness of professionals in the health system, thereby, equipping primary care providers with the ability to suspect unusual clinical cases like this one.

Once microorganisms reach a high bacillary load, the host acts as a source of reservoir^{9,11,12} and spreads the disease. Moreover, children with delayed diagnosis and appropriate treatment are faced with various degrees of incapacity, which compromises their future working capacity.



FIGURE 1 - Keloid nodules over the left forearm.

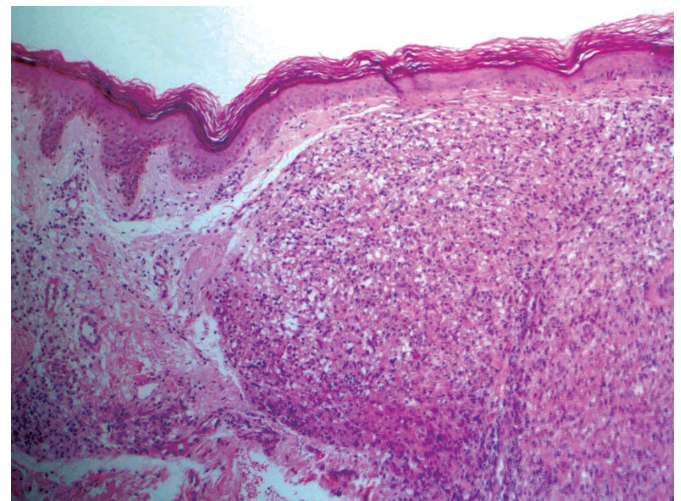


FIGURE 2 - Skin biopsy: nodular proliferation of spindle-shaped histiocytes in the dermis.

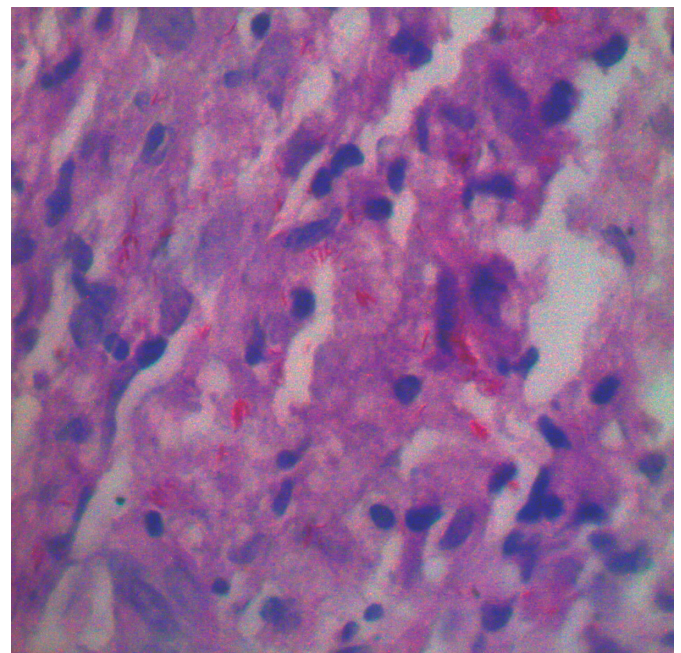


FIGURE 3 - Fite-Faraco staining: numerous alcohol-acid-resistant bacilli isolated and grouped together.

In addition, the impact of leprosy on children goes beyond physical damage, as it also entails psychological suffering due to the prejudice and stigma associated with the disease. The presence of lesions in visible areas, including the face and upper limbs, led to this adolescent avoiding social contact, demonstrating how the disease, though curable today, is still associated with widespread rejection and discrimination.

Conflict of interest

The authors declare that there is no conflict of interest.

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