

Hyperpigmented macules surrounded by hypopigmented rim in a case of lepromatous leprosy: an atypical presentation

Máculas hiperpigmentadas delimitadas por borda hipopigmentada em um caso de hanseníase virchowiana: uma apresentação atípica

Máculas hiperpigmentadas delimitadas por un borde hipopigmentado en un caso de lepra lepromatosa: una presentación atípica

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ABSTRACT

Introduction: leprosy is a chronic infectious mycobacterial disease caused by *M. leprae*. It primarily involves the skin and nerves, presenting with skin lesions and nerve enlargement. The skin lesions vary from hypopigmented, anesthetic macules/plaques to erythematous papules, plaques, and nodules, depending upon the type of leprosy. Hyperpigmented lesions in leprosy are rarely reported. Although hypopigmented lesions are commonly seen, perilesional hypopigmentation is a rare occurrence in leprosy. Perilesional hypopigmentation is seen as Woronoff's ring around resolving lesions of psoriasis. A similar phenomenon called the halo phenomenon is associated with various skin tumors. **Case**

Presentation: we describe a rare presentation of a leprosy case presenting with hyperpigmented lesions surrounded by a perilesional hypopigmented rim.

Keywords: *Leprosy. Multidrug therapy. Woronoff's ring. Hypopigmented halo. Mycobacterium leprae.*

RESUMO

Introdução: a hanseníase é uma doença infecciosa crônica, causada pelo *M. leprae*. Acomete principalmente a pele e os nervos periféricos, apresentando lesões cutâneas e espessamento de nervos. As lesões cutâneas variam de máculas/placas hipopigmentadas anestésicas, a pápulas, placas e nódulos eritematosos, a depender do tipo de hanseníase. Lesões hiperpigmentadas raramente são relatadas na hanseníase. Por outro lado, lesões hipopigmentadas são comumente observadas. Entretanto, a hipopigmentação perilesional é uma ocorrência rara na hanseníase. A hipopigmentação perilesional, denominada de anel de Woronoff, é encontrada como um halo ao redor das lesões de psoríase em resolução. Um fenômeno semelhante denominado de fenômeno do halo é observado em associação com vários tumores de pele. **Descrição do caso:** descrevemos um caso raro de hanseníase, apresentando lesões hiperpigmentadas delimitada por uma borda perilesional hipopigmentada.

Palavras-chave: *Hanseníase. Poliquimioterapia. Anel de Woronoff's. Halo hipopigmentado. Mycobacterium leprae.*

RESUMEN

Introducción: la lepra es una enfermedad infecciosa crónica causada por *M. leprae*. Afecta principalmente a la piel y a los nervios periféricos, con lesiones cutáneas y engrosamiento de los nervios. Las lesiones cutáneas varían de máculas/placas hipopigmentadas anestésicas a pápulas, placas y nódulos eritematosos, según el tipo de lepra. Las lesiones hiperpigmentadas son poco frecuentes en la lepra. En cuanto a las lesiones hipopigmentadas, son frecuentes. Sin embargo, la hipopigmentación perilesional es rara en la lepra. La hipopigmentación perilesional, llamada anillo de Woronoff, se encuentra como un halo alrededor de las lesiones de psoriasis que se resuelven. Un fenómeno similar llamado fenómeno de halo se observa en asociación con diversos tumores cutáneos. **Descripción del caso:** describimos un caso poco frecuente de lepra, que presenta lesiones hiperpigmentadas delimitadas por un borde perilesional hipopigmentado.

Palabras clave: *Lepra. Terapia Multimedicamentosa. Anillo de Woronoff. Halo Hipopigmentado. Mycobacterium leprae.*

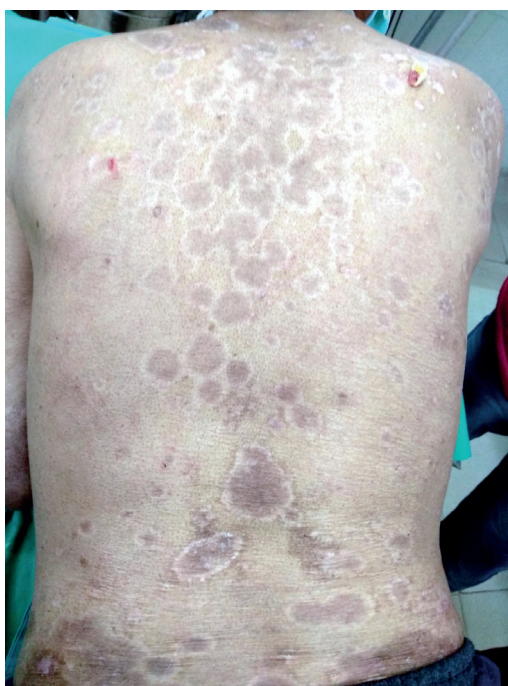
INTRODUCTION

Leprosy, or Hansen's disease, can present with various skin lesions commonly described as hypopigmented, coppery, or erythematous in color. The morphology and distribution of the lesions vary depending on the type of leprosy^{1,2}. Both hyperpigmented lesions and perilesional hypopigmentation have rarely been reported in leprosy. We describe a rare case with both of these findings.

CASE PRESENTATION

A 66-year-old male presented with chief complaints of skin lesions over the trunk and limbs along with loss of sensations over hands and feet for nine months. There was no history of similar complaints in the family. On examination, multiple hyperpigmented macules, tending to be confluent, were seen over the trunk and limbs (Figure 1). The lesions were symmetrically distributed, and few had partial loss of temperature and pain sensations. The skin over the back was shiny and infiltrated. Bilateral ulnar, radial cutaneous, and lateral popliteal nerves were found to be enlarged and non-tender. Sensory examination revealed 'glove and stocking' anesthesia over the hands and feet. Motor examination was normal. A slit skin smear was positive for acid-fast bacilli. A diagnosis of lepromatous leprosy was made, a skin biopsy taken from the hyperpigmented center, and the hypopigmented peripheral rim was sent for histopathological examination.

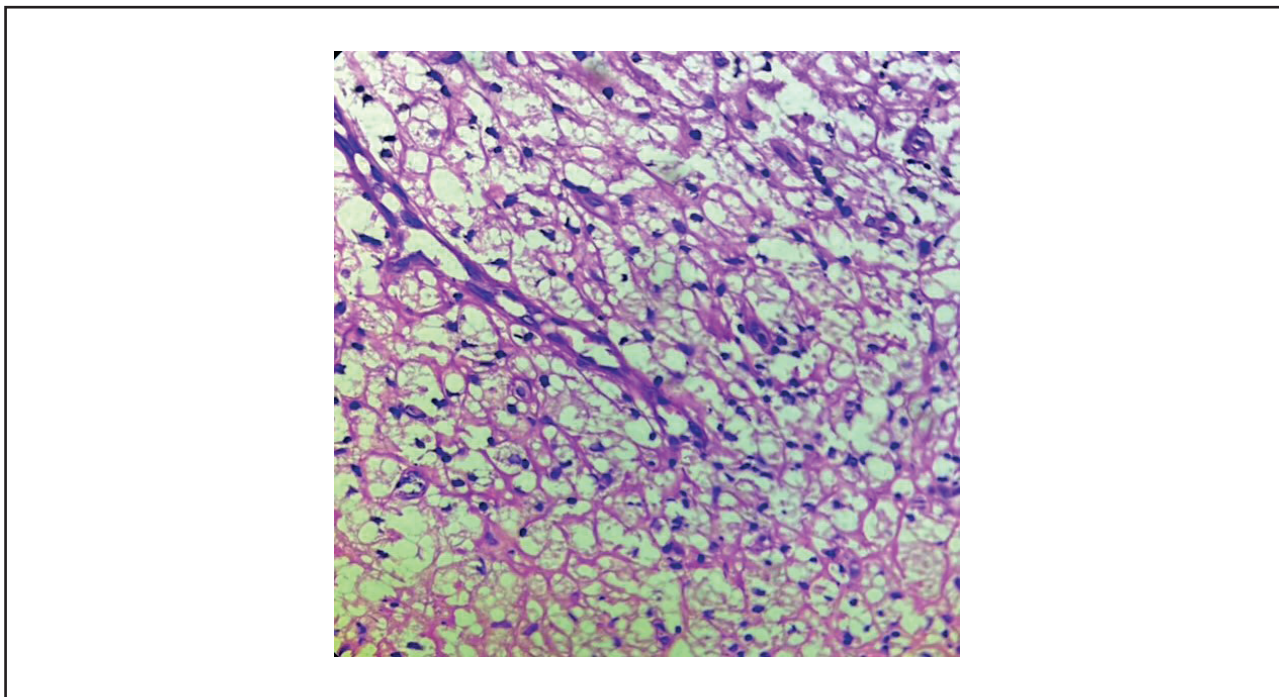
Figure 1 – Hyperpigmented macules surrounded by perilesional hypopigmented rim over the back.



Source: Created by the author.

Both showed a subepidermal Grenz zone, foamy macrophages, and scattered lymphocytes consistent with a diagnosis of lepromatous leprosy (Figure 2). Special stains for melanin could not be performed due to a lack of availability at our center. The patient was started on WHO multibacillary multidrug therapy, comprising monthly supervised doses of rifampicin 600 mg, clofazimine 300 mg, daily dapsone 100 mg, and clofazimine 50 mg.

Figure 2 – Photomicrograph showing foamy macrophages, and scattered lymphocytes (H and E, 100x).



Source: Created by the author.

DISCUSSION

Skin lesions in Hansen's disease can have varied colors, commonly described as hypopigmented, coppery, or erythematous^{1,2}. Hyperpigmentation in leprosy can occur but is usually post-inflammatory associated with leprosy reactions or secondary to treatment with drugs like clofazimine or minocycline^{3,4}. Primary lesions in leprosy are not usually hyperpigmented, and only a few such cases have been reported in the literature³⁻⁹. The majority of these reports are of borderline-tuberculoid leprosy presenting with hyperpigmented macules and patches⁴⁻¹⁰. One of the patients in the report by Singh et al. also showed perilesional hypopigmentation besides the hyperpigmented macules⁵. A literature search revealed only a single case of borderline-lepromatous leprosy reported with such lesions by Abideen et al.³. In their case, the patient had both hypopigmented and hyperpigmented macules. The hyperpigmented macules also had a rim of perilesional hypopigmentation around them. Histopathology of hyperpigmented and

hypopigmented lesions using special stains in their case revealed melanocytes with a mild increase and a mild decrease in melanin granules, respectively³. The cause of hyperpigmentation in primary leprosy lesions has not been elucidated. Some proposed hypotheses include melanocyte activation due to the upregulation of stimulating factors like hepatocyte growth factor, stem cell factor, and fibroblast growth factor⁴. The primarily hypopigmented lesions in leprosy are believed to be due to defective transfer of melanin from melanocytes to keratinocytes¹¹, melanocyte destruction and inhibition due to bacillary multiplication, infiltration, and nerve damage¹², and release of neurochemical mediators due to nerve damage¹³. The cause of perilesional hypopigmentation is not exactly known. In the only case of borderline-lepromatous leprosy reported by Abideen et al., defective melanin transfer to keratinocytes was proposed as the cause of perilesional hypopigmentation based on immunohistochemistry (IHC) and special stains on histopathology³. In our case, we could not perform IHC or special staining due to lack of availability. In the literature search, no case of lepromatous leprosy with a similar presentation could be found.

CONCLUSION

Primary lesions in leprosy are characteristically described as hypopigmented, erythematous, or coppery. Hyperpigmented lesions have rarely been reported, and most of these cases are of borderline tuberculoid leprosy. We report a rare presentation of lepromatous leprosy presenting with hyperpigmented macules with perilesional hypopigmentation.

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