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Hypopigmented mycosis fungoides mimicking leprosy successfully treated with oral and topical corticosteroids: a new great imitator?

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Abstract

Hypopigmented mycosis fungoides (HMF) is a rare variant of patch stage MF, which is often misdiagnosed. A 35-year-old male presented with non-pruritic white patches on his chest that had been present for 10 years. The patient had previously been treated for leprosy without any improvement. Physical examination showed well-defined multiple hypopigmented patches and macules on the chest, posterior trunk, and gluteus, with some lesions exhibiting anhidrosis and central erythema. The result of sensibility examination was unclear. Slit-skin-smear examination for acid-fast bacilli and anti-phenolic-glycolipid-1 examination were negative. Histopathological examination showed Pautrier microabscesses. The patient was diagnosed with HMF and was treated with 16 mg methylprednisolone b.i.d., topical application of desoximetasone, and 1% methoxsalen lotion followed by sun exposure. A significant improvement was observed during the following 6 months. This case shows that HMF needs to be considered in patients presenting with chronic unexplained hypopigmented patches to avoid unnecessary treatment and progression to more advanced stages.

Keywords: hypopigmented mycosis fungoides, leprosy, mimicking

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Introduction

Mycosis fungoides (MF) is the most common type of cutaneous T-cell lymphoma (CTCL), which is characterized by malignant monoclonal T-cell infiltration of the skin (1). The clinical course of MF can be divided into three stages: patch stage, plaque stage, and tumor stage (2). The patch stage typically presents with asymptomatic to extremely pruritic erythematous patches that may last for decades before progressing to the plaque stage, although some stages can simultaneously be found (3).

Hypopigmented MF (HMF) is a rare variant of patch stage MF, which presents as hypopigmented instead of erythematous patches (4). Unlike classic MF, which commonly occurs in the 5th and 6th decades, HMF tends to affect a younger population and has been reported in children (2). Due to its atypical presentation, it is often overlooked and mistreated as other hypopigmentation disorders.

This case presents a young adult male with HMF who presented with multiple longstanding asymptomatic hypopigmented patches, mistreated for leprosy.

Case report

A 35-year-old male presented with a chief complaint of non-pruritic white patches on his chest that had persisted for 10 years. The condition initially appeared on the chest, and the patches gradually increased in size and number. The patient could not confidently state whether the patches were numb. The patient had previously been treated for leprosy using rifampicin, ofloxacin, and minocycline for 1 year without any significant improvement. No household members experienced the same complaint. The patient reported no history of leprosy or malignancy.

Physical examination showed that the patient was in good general condition with normal vital signs. Dermatological examination revealed well-defined multiple hypopigmented patches and macules on the chest, posterior trunk, and gluteus; some lesions exhibited anhidrosis and central erythema (Fig. 1). Neurological examination did not show nerve enlargement or motor deficit; however, sensation impairment was difficult to assess due to the inconsistent result reported by the patient.

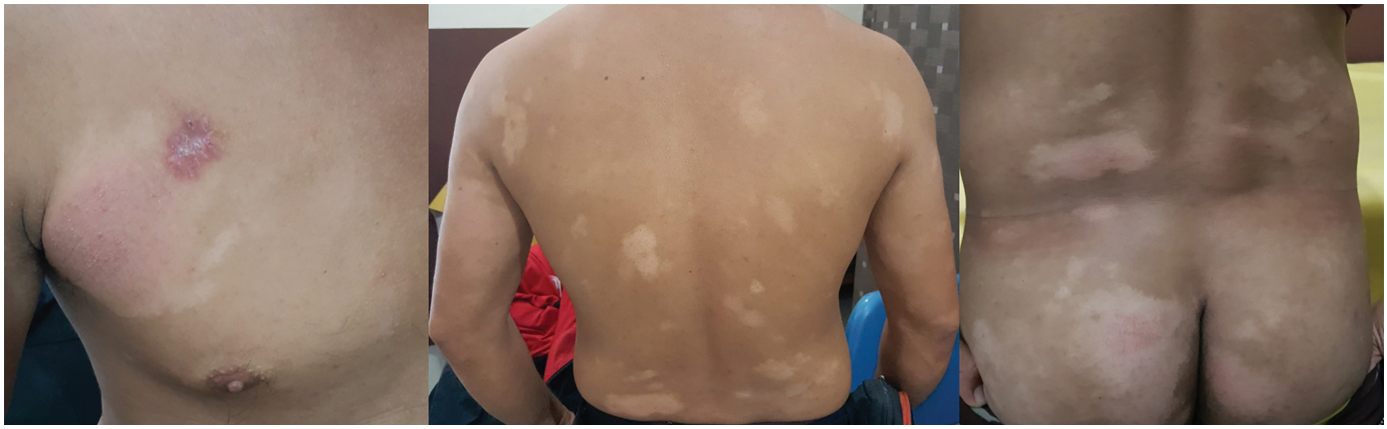

Figure 1 | Pre-treatment.

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The results of routine hematological examination, blood glucose profile, liver function, and renal function were within normal limits. Slit-skin-smear examination for acid-fast bacilli and anti-phenolic-glycolipid-1 (PGL-1) examination were negative. Skin scraping examination using a potassium hydroxide examination revealed no fungal elements. Histopathological examination showed lymphocytic infiltration into the epidermis without any signs of epidermal damage (Fig. 2A). Focal parakeratosis was seen in the stratum corneum (Fig. 2B) and some lymphocytes coalesced and formed Pautrier microabscesses (Fig. 2C). Parakeratosis and lymphocytic infiltration of the epidermis and dermis were evident (Fig. 2D), while granuloma was not observed.

From the history taking, physical findings, and results of supporting examinations, the patient was diagnosed with HMF. The patient was treated with 16 mg methylprednisolone b.i.d., topical application of desoximetasone, and topical application of 1% methoxsalen lotion followed by sun exposure. Significant improvement was observed after 1 month of treatment. The dose was maintained for 3 months before being tapered to 8 mg b.i.d. Significant clinical improvement with no development of new lesions was maintained until 6 months of follow-up (Fig. 3).

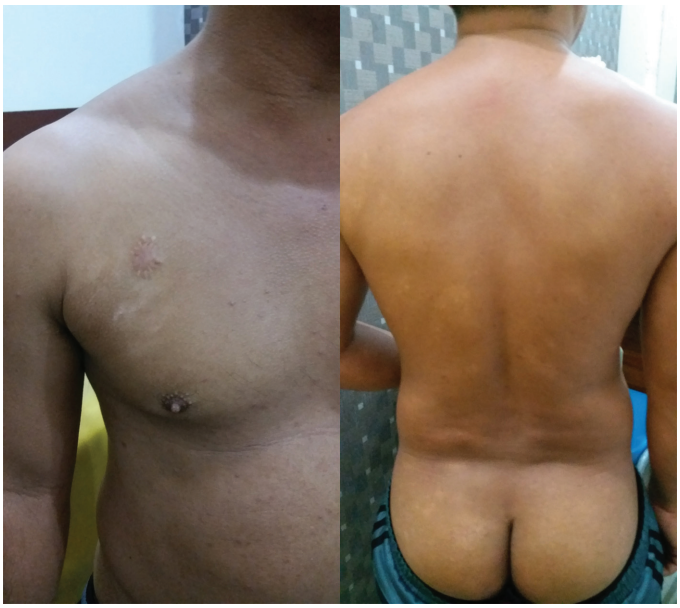

Figure 3 | Six months after treatment.

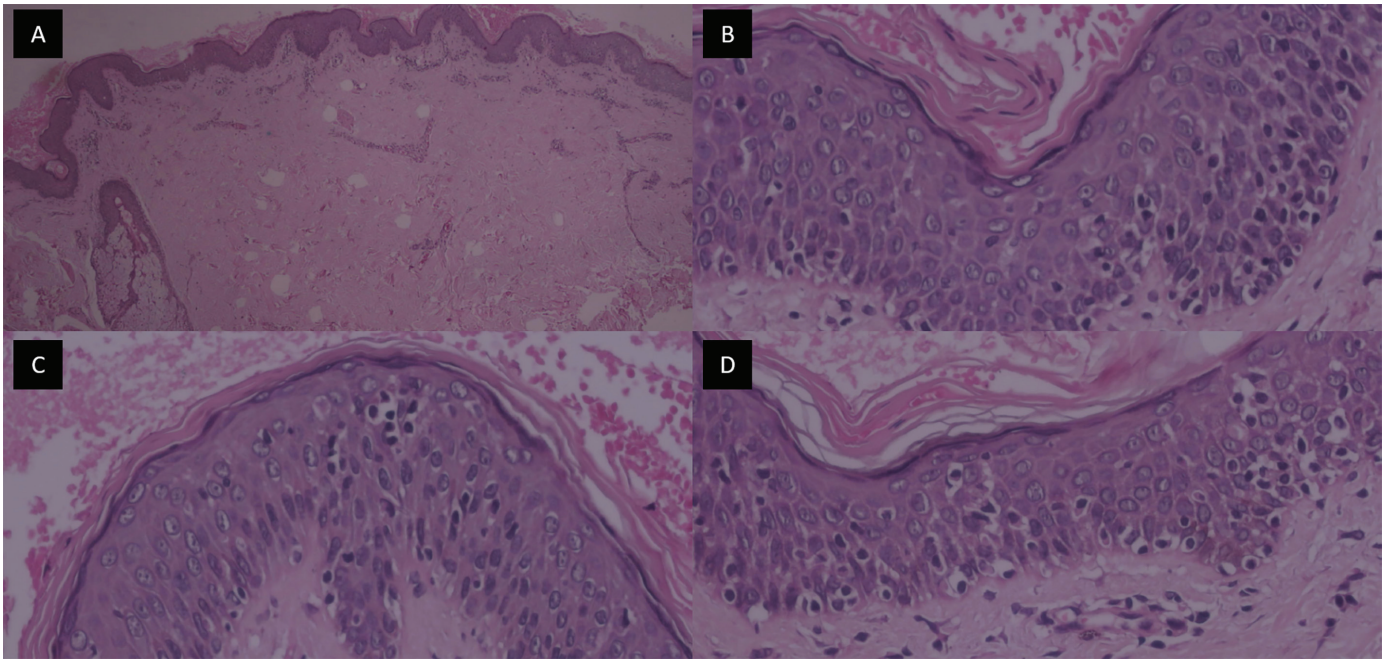

Figure 2 | A) Lymphocytic infiltration into the epidermis without any signs of epidermal damage (4×); B) focal parakeratosis was seen in the stratum corneum (10×); C) some lymphocytes coalesced and formed Pautrier microabscesses (10×); D) parakeratosis and lymphocytic infiltration of the epidermis and dermis were evident (10×), granuloma was not observed.

Discussion

Our patient had an non-specific clinical presentation and thus prompted the exclusion of a vast array of differential diagnoses. Differential diagnoses in this case included leprosy, progressive macular hypomelanosis, vitiligo, and pityriasis versicolor.

Clinically, leprosy was the initial and main differential diagnosis due to the anhidrotic appearance of some of the hypopigmented patches. Anhidrosis is one of the typical clinical presentations of leprosy, which is caused by denervation of the skin appendages and neural sheath thickening (5). This suspicion was further confounded by the unclear sensitivity examination. Although an anesthetic hypopigmented patch is one of the cardinal symptoms of leprosy, in some types of leprosy, particularly those with a high bacterial index (BI), impaired sensation could be unclear due to low cell-mediated immunity (6). However, the 10-year clinical course our patient presented prompted the consideration of other diagnoses because, given such a duration, leprosy with a high BI would have typically more likely resulted in apparent clinical sensory and motor manifestations (6). Leprosy was eventually excluded through the absence of granuloma on histopathological examination, which is a characteristic histopathological feature in leprosy (7).

Progressive macular hypomelanosis is characterized by ill-defined nummular hypopigmented macules (8), and thus the size and border in this case did not favor this diagnosis. Typical vitiligo presents with depigmented and well-defined hypopigmented lesions, although new evolving lesions may present with hypopigmentation and an unclear border (9). Finally, hypopigmented macules in pityriasis versicolor are usually covered with fine scales, especially upon stretching (10), which were absent in this case. It was possible to exclude progressive macular hypomela-

nosis and vitiligo using histopathological examination, and pityriasis versicolor could be excluded due to the absence of fungal elements under microscopic examination.

Clinical histopathological correlation is the cornerstone for establishing HMF. The most widely reported and accepted key histopathological difference between HMF and classic MF is the prominent epidermotropism of CD8+ T-cells in HMF instead of CD4+ T-cells, which are observed in classic MF (2). This phenomenon is thought to induce non-specific melanocyte injury and hence causes hypopigmented lesions (2). The high number of CD8+ T-cells, which favors Th-1 response compared to Th-2 response, is also linked to the better prognosis of HMF because they are postulated to play a role in preventing MF progression to more advanced stages (4). Pautrier microabscess, which is reported to be a rare finding in HMF (2), was observed in our case and acted as an important clue for diagnosing HMF. Other histopathological features in our case, such as focal parakeratosis and dermal lymphocytic infiltrate, also support MF (2).

Corticosteroids, in both topical and oral preparations, are effective in patch stage HMF and MF (1). Although topical corticosteroids are the more commonly prescribed agent, we also decided to administer oral corticosteroid due to the large body surface area affected. In addition to corticosteroids, phototherapy, especially ultraviolet B, is a commonly used and effective treatment. However, due to access limitation, topical 1% methoxsalen lotion application followed by sun exposure 1 to 2 hours afterward was used instead. Methoxsalen is a photosensitizer agent that is used to accentuate the effect of ultraviolet A treatment (11).

Although the prognosis of HMF is favorable, recurrence is frequent, and thus regular follow-up and avoiding excessively aggressive treatment are warranted (2, 12).

Conclusions

HMF needs to be considered in patients presenting with chronic unexplained hypopigmented patches. Although the prognosis is favorable, prompt diagnosis and treatment are important to avoid unnecessary treatment and progression to more advanced stages.

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