Pediatric and Adult Hypopigmented Mycosis Fungoides: A Case Report

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Abstract: Hypopigmented mycosis fungoides (HMF) is a rare variant of cutaneous T-cell lymphoma that predominantly affects children and individuals with darker skin. We report two Filipino patients, a 4-year-old girl and a 34-year-old man, who presented with progressive, non-pruritic hypopigmented patches. Histopathology and immunohistochemistry confirmed HMF, revealing CD8+ predominance in the pediatric case and CD3+ epidermotropic cells in the adult. The child demonstrated a favorable response to narrowband UVB phototherapy, while the adult showed early improvement with topical corticosteroids. These cases underscore the importance of considering HMF in persistent hypopigmented dermatoses and highlight the value of clinicopathologic correlation for timely diagnosis and management.

Keywords: Hypopigmented mycosis fungoides, Cutaneous lymphoma, Epidermotropism, Phototherapy

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Introduction

Hypopigmented skin lesions are a common concern among patients attending dermatologic outpatient clinics. One of the underlying conditions is mycosis fungoides (MF), the most prevalent form of primary cutaneous T-cell lymphoma (CTCL). MF accounts for approximately 62% of all CTCL cases and is classified as an extranodal non-Hodgkin lymphoma [1].

Hypopigmented MF (HMF) is a clinical variant of MF [2]. HMF is more common in individuals with darker skin phototypes, including those of African, South Asian, Middle Eastern, and Hispanic descent, where the lesions are more clinically apparent [3,4]. Most studies report an approximately 1:1 female-to-male ratio [5]. HMF tends to occur at a younger age, with cases described in children, adolescents, and young adults [6], including infants as young as six months [7].

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Clinically, HMF usually presents as circular or irregular hypopigmented patches or thin plaques with fine scaling. These lesions are often asymptomatic or only mildly pruritic and are most commonly located on the torso, buttocks, and extremities. Diagnosis can be challenging and is often delayed due to the disease's slow progression and resemblance to other hypopigmented conditions, such as vitiligo, tinea corporis, pityriasis versicolor, pityriasis alba, post-inflammatory hypopigmentation, progressive macular hypomelanosis, and leprosy [8]. Consequently, HMF is frequently overlooked, leading to delays in treatment.

This case report aims to highlight the essential role of clinicopathologic correlation in the timely diagnosis and management of HMF. Additionally, it seeks to contribute to the limited literature on this rare condition by detailing its clinical presentation, diagnostic approach, and management.

Case Presentation

Case 1: A 4-year-old Filipino girl presented with a 3-year history of hypopigmented macules and patches involving the face, trunk, buttocks, and bilateral upper and lower extremities. The lesions initially appeared as erythematous plaques on the trunk. Dermatologic examination revealed multiple hypopigmented macules and patches distributed over the face, trunk, back, gluteal region, and extremities **(Figure 1A).**

Routine investigations, including complete blood count, peripheral blood smear, liver and renal function tests, lactate dehydrogenase, and chest radiograph, were within normal limits. A 4-mm skin punch biopsy was performed twice. The repeat biopsy showed basket-weave orthokeratosis, a few lymphocytes along the dermoepidermal junction with focal epidermal collections, and a moderately dense superficial perivascular infiltrate composed of lymphocytes and melanophages. Immuno-histochemistry (IHC) revealed CD4 and CD8 highlighting epidermotropic and atypical cells in the epidermis, with CD8 predominance over CD4 and focal loss of CD7—findings consistent with HMF **(Figure 1B)**.

The patient was treated with narrowband UVB phototherapy, initiated at 300 mJ/cm² with a 10% incremental increase per session three times weekly, resulting in areas of repigmentation after three months **(Figure 1C)**.

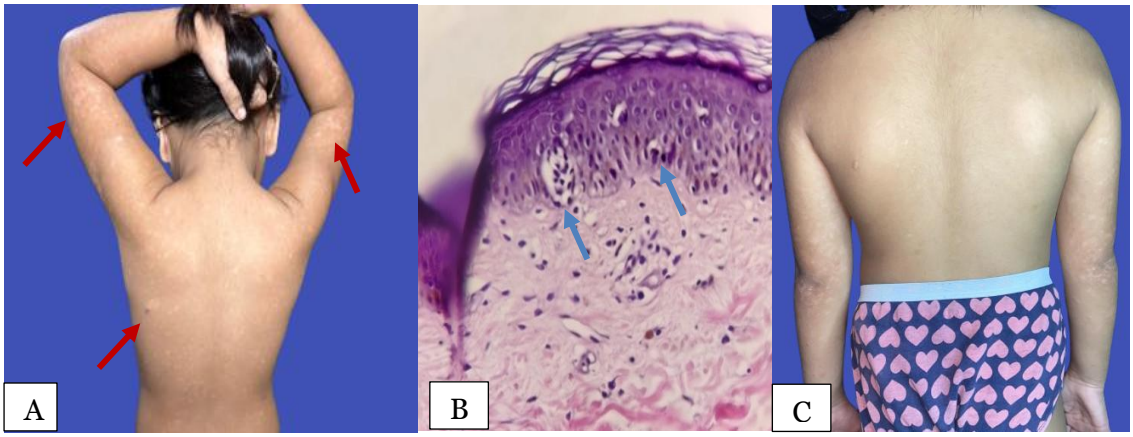

Figure 1. (A) Dermatologic examination showing multiple hypopigmented macules and patches on the trunk of the 4-year-old female (red arrows); **(B)** Histopathologic sections (H&E) demonstrating a focal collection of vacuolated cells at the dermoepidermal junction, with a few cells exhibiting epidermotropism (blue arrow); **(C)** Reduction in the number of hypopigmented lesions following narrowband UVB therapy.

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Case 2: A 34-year-old Filipino man presented with a 5-year history of non-pruritic hypopigmented patches, which initially appeared on the bilateral upper extremities and gradually increased in size and number, eventually spreading to the trunk and bilateral lower extremities. Dermatologic examination revealed multiple hypopigmented macules and patches on the trunk and extremities **(Figure 2A).**

Routine investigations, including complete blood count, liver and renal function tests, and chest radiograph, were normal. A 4 mm skin punch biopsy was performed three times, with the third biopsy demonstrating superficial perivascular dermatitis. Histopathologic examination revealed basket-weave orthokeratosis, basal layer hyperpigmentation, subtle epidermotropism, and a focal collection of vacuolated cells at the dermoepidermal junction. A mild superficial perivascular infiltrate composed of lymphocytes, histiocytes, and melanophages was also noted. Immunohistochemistry highlighted epidermotropic and atypical epidermal cells with CD3 staining, findings consistent with HMF **(Figure 2B)**. Due to financial constraints, CD4 and CD8 staining were not performed, and CD3 staining was carried out to support the diagnosis.

Phototherapy was not feasible due to the patient's work schedule and distance from the hospital. Consequently, he was started on clobetasol propionate 0.05% lotion applied twice daily, with minimal repigmentation observed on the trunk and extremities after one month of treatment. The patient was, however, lost to follow-up.

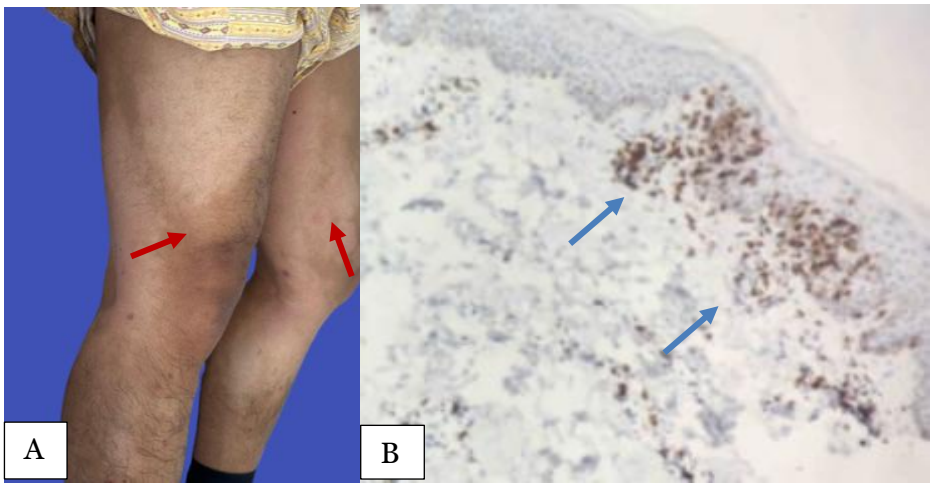

Figure 2. (A) Dermatologic examination showing multiple hypopigmented patches on the lower extremities of the 34-year-old male (red arrows); **(B)** Immunohistochemistry with CD3 demonstrating epidermotropic and atypical cells in the epidermis (arrow).

Discussion

HMF is a distinct variant of mycosis fungoides characterized by infiltration of cytotoxic CD8+ T lymphocytes in the skin, which target melanocytes and lead to pigment loss [9]. This immunopathogenic mechanism underpins the hallmark hypopigmented lesions observed predominantly in children, adolescents, and young adults, as seen in the 4-year-old girl and the 34-year-old man presented. HMF poses significant diagnostic and therapeutic challenges. The chronicity of lesions, their distribution, and subtle symptoms such as pruritus or atrophy may mimic benign dermatologic conditions, often delaying diagnosis, as evident in both cases where a prolonged history preceded diagnosis.

Diagnosis of HMF relies on clinicopathologic correlation [9], with histopathology and immunohistochemistry being

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equally important for confirmation. Immunohistochemical analysis is particularly valuable because neoplastic cells in HMF often express CD8, a hallmark feature of this variant [5]. Unlike classical MF, which is characterized by epidermotropic CD4+ T cells, hypopigmented MF typically shows a predominance of CD8+ cells [8].

In a retrospective review of 67 patients with CD8+ MF, 20.9% presented with hypopigmented lesions [10]. Similarly, Koikkara et al. found CD8+ predominance in 56% of cases, and Rodney et al. observed a CD8+ predominant infiltrate in 58.3% of patients [4,11], supporting the notion that HMF is a CD8+-predominant variant of MF. Partial loss of CD7 has also been reported in some cases [12]. This was evident in our first case, which demonstrated CD8 predominance and partial CD7 loss, consistent with literature linking CD8+ cytotoxic T cells to melanocyte destruction and hypopigmentation. In the second case, immunohistochemistry revealed CD3+ epidermotropic cells, in line with findings by Nazareth, who reported that neoplastic cells in HMF express CD3, a pan–T-cell marker [13]. Koikkara also found CD3 positivity in all patients evaluated [11].

First-line management for HMF includes phototherapy, such as narrowband ultraviolet B, and photochemotherapy (psoralen and ultraviolet A [PUVA]), often combined with topical agents including corticosteroids, retinoids, imiquimod, or nitrogen mustard [6]. Successful outcomes with NB-UVB combined with topical corticosteroids have been reported [8]. Early-stage HMF generally responds well to skin-directed therapies. In the pediatric case, narrowband ultraviolet B led to notable repigmentation within three months, affirming its efficacy. Adults may require alternative or adjunctive therapies, particularly when phototherapy is impractical, as demonstrated by the second patient, who showed minimal improvement with topical clobetasol.

Prognostically, stratification and clinical staging remain the most reliable indicators for MF. HMF carries an excellent prognosis, especially in pediatric populations, with low rates of progression to advanced disease. Although relapse of hypopigmented lesions during treatment is relatively common, disease progression remains uncommon [14]. Regular follow-up is essential to monitor treatment response and detect any early signs of progression.

Conclusion

This report highlights two rare cases of HMF in Filipino patients, one pediatric and one adult, underscoring the clinical variability and diagnostic challenges of this condition. Early-stage HMF may present with hypopigmented patches that mimic benign skin disorders, often delaying diagnosis and treatment. Histopathologic and immunohistochemical studies, particularly the presence of CD8+ epidermotropic lymphocytes, remain essential for accurate identification. Both cases demonstrated favorable early responses to treatment, reinforcing that HMF generally carries an excellent prognosis. Therefore, a high index of suspicion is necessary in patients with persistent hypopigmented patches, especially in covered areas. Timely recognition, appropriate diagnostic work-up, and individualized management strategies are crucial for improving outcomes in patients with HMF.

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Potential Conflict of Interest

The authors declare no potential conflicts of interest.

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Contribution of Authors

All authors critically reviewed and revised the manuscript and approved the final version.

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