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Case Report **OPEN ACCESS**

Mycosis fungoides in childhood: A diagnostic challenge for primary care physicians: A case report

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ABSTRACT

Mycosis fungoides (MF) is the most common form of cutaneous T-cell lymphoma in adult male, though it is rare in children. Pediatric MF often presents with hypopigmented patches, which can be mistaken for common skin conditions such as pityriasis alba, tinea versicolor, vitiligo, leprosy, or post-inflammatory hypopigmentation. This difficulty can lead to delayed diagnosis and misdiagnoses by healthcare workers. We report a case of hypopigmented mycosis fungoides (HMF) in a 4-year-old boy. The diagnosis was confirmed by skin biopsy and immunohistochemistry, which demonstrated features consistent with MF. The patient was initially treated with a topical steroid, however, owing to an inadequate clinical response, treatment was subsequently changed to a topical calcineurin inhibitor. This case highlights the importance of considering HMF in children with persistent or progressive hypopigmented lesions that do not respond to conventional treatment. Early skin biopsy and immunohistochemical correlation.

Keywords: mycosis fungoides, hypopigmentation, cutaneous T-cell lymphoma, pediatric dermatology

INTRODUCTION

Mycosis fungoides (MF) is the most common form of cutaneous T-cell lymphoma (CTCL) that typically presents in middle age with a 2:1 male to female predominance [1]. The annual incidence of MF ranges from 2 to 4 cases per 1,000,000 individuals [2]. To our knowledge, the incidence of MF in the pediatric population is rarely reported in the literature [3]. There are several common presentations of MF among the pediatric age group, such as hypopigmented mycosis fungoides (HMF), classical MF, folliculotropic MF, and poikilodermatous MF. HMF is the most common clinical variant reported in the children as compared with others [4]. HMF commonly presents on the trunk and extremities, particularly in sun-protected areas. Frequently, the lesions may resemble other skin conditions, and the differential diagnoses include pityriasis alba, tinea versicolor, vitiligo, leprosy, and post-inflammatory hypopigmentation, which can lead to misdiagnosis among primary care doctors and delayed referral to the dermatology department for skin biopsy [5].

We report a case of HMF in a 4-year-old boy, with the diagnosis confirmed by histopathological examination of a skin biopsy. This case highlights the importance of a systematic approach to childhood hypopigmented lesions and maintaining a high index of suspicion for HMF when lesions are persistent, progressive, or unresponsive to initial treatment. Early referral to a dermatology service for further evaluation and skin biopsy may facilitate timely diagnosis and management. Early-stage HMF generally has a favorable prognosis, although long-term clinical surveillance remains important.

CASE PRESENTATION

A 4-year-old boy, who was previously well, presented with a 1-year history of progressive generalized hypopigmented lesions. The lesion initially started as erythematous papules, followed by scaling, and then became a hypopigmented patch. It was not associated with itchiness, and it was painless in nature. He was brought to the primary care clinic and treated for a few diagnoses such as pityriasis rosea, post-inflammatory hypopigmented lesions, and eczema. He was treated with a mild-potency of topical steroid and emollient; however, the lesions did not improve. His parents denied any family history of blood disorders or malignancy. It was not associated with constitutional symptoms, and there was no fever. Due to the unresolved lesion, the patient was then referred to the dermatology department. The patient was first seen by the dermatology team approximately 11 months after the onset of the lesions. Upon review by the dermatology team, the child appeared cheerful. There was a generalized distribution of variably sized and partly-ill-defined hypopigmented macules and patches involving both upper and lower limbs, the anterior trunk, and posterior trunk as illustrated in **Figure 1**.

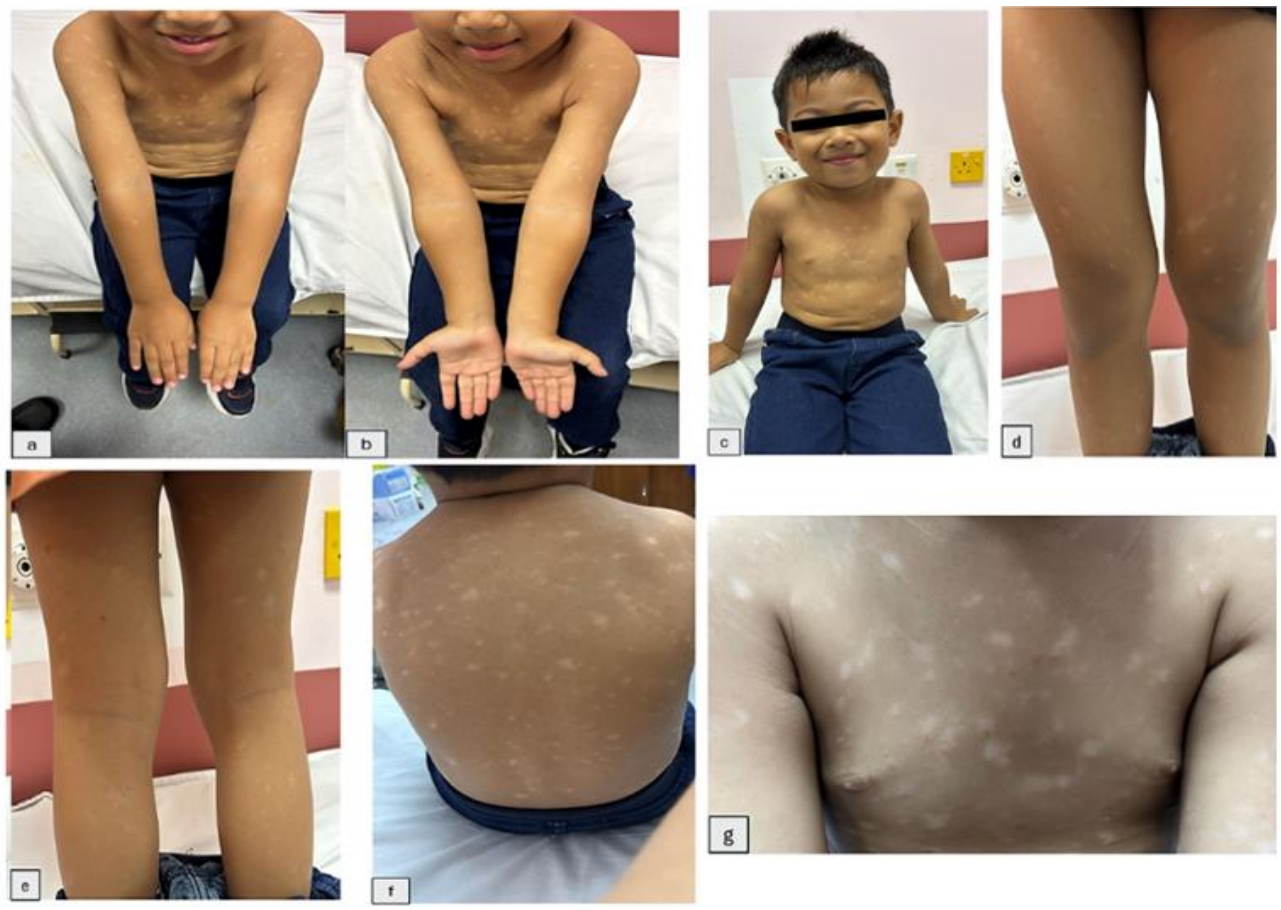

Figure 1. Clinical distribution of hypopigmented mycosis fungoides: (a) Mmultiple hypopigmented macules and patches over the dorsal aspects of both upper limbs; (b) similar lesions over the flexor forearms, with sparing of the palms; (c) widespread lesions over the anterior trunk and upper limbs; (d) scattered lesions over the anterior aspects of both lower limbs; (e) lesions over the posterior aspects of both lower limbs; (f) numerous hypopigmented lesions over the posterior trunk; & (g) a closer view of variably sized, partly ill-defined hypopigmented patches over the upper anterior trunk (the face, neck, palms, and soles were spared, with no mucosal or nail involvement) (reprinted with permission of the parents)

Table 1. Blood investigation summary

Blood parameter	Result	Reference range
WCC	9.3	5.0-15.0 × 10^9
Hb	12.0	11.0-14.0 g/dL
MCV	85.2	85.0-87.0 fL
MCH	25.0	24.0-30.0 pg
PLT	354	200-490 10^9/L
Urea	3.1	2.8-7.2 mmol/L
Creatinine	24	23-68 µmol/L
K	4.8	3.5-5.1 mmol/L
Na	136	135-145 mmol/L
ALT	17	< 45 U/L
AST	37	< 75 U/L
Alb	45	35-52 g/L
TBil	4.4	5.0-21.0 µmol/L
DBil	0.8	< 3.4 µmol/L
VDRL	Non-reactive	
Anti-HCV	Non-reactive	
HBsAg	Non-reactive	
HIV Ag/Ab	Non-reactive	

Note. MCV: Mean corpuscular volume; MCH: mean corpuscular hemoglobin; K: Potassium; Na: Sodium, ALT: Alanine transaminase; AST: Aspartate aminotransferase; Alb: Albumin, TBil: Total bilirubin; DBil: Direct bilirubin; ESR: Erythrocyte sedimentation rate; VDRL: Venereal Disease Research Laboratory; Anti-HCV: Hepatitis C virus antibody; HBsAg: Hepatitis B surface antigen; & HIV Ab/Ag: Human immunodeficiency virus antibody/antigen

However, there was sparing of face, necks, palms and soles, with no mucosal or nail involvement. There were no palpable cervical, axillary, or inguinal lymph nodes.

Laboratory investigations were performed to assess hematological status, hepatic and renal function, electrolyte balance, systemic inflammation, and possible infectious mimics or comorbid infections as summarized in **Table 1**.

The full blood count was within the age-appropriate laboratory reference ranges, with a white cell count (WCC) of 9.3 × 10^9/L, hemoglobin (Hb) of 12.0 g/dL, and platelet (PLT) count of 354 × 10^9/L, indicating no leukocytosis, anemia, or thrombocytopenia. Renal function and electrolytes were normal (urea 3.1 mmol/L, creatinine 24 µmol/L, potassium 4.8 mmol/L, and sodium 136 mmol/L). Liver enzymes and albumin were also within the reference ranges (AST 37 U/L, ALT 17 U/L, and albumin 45 g/L), with no biochemical evidence of hepatic dysfunction. Total bilirubin was mildly below the laboratory reference range at 4.4 µmol/L; in isolation, this was not considered clinically significant. The erythrocyte sedimentation rate was 19 mm/hour, within the reference range, providing no evidence of a marked systemic inflammatory response. Screening for syphilis (VDRL), hepatitis C (anti-HCV), hepatitis B (HBsAg), and HIV (HIV-antigen/antibody) was non-reactive.

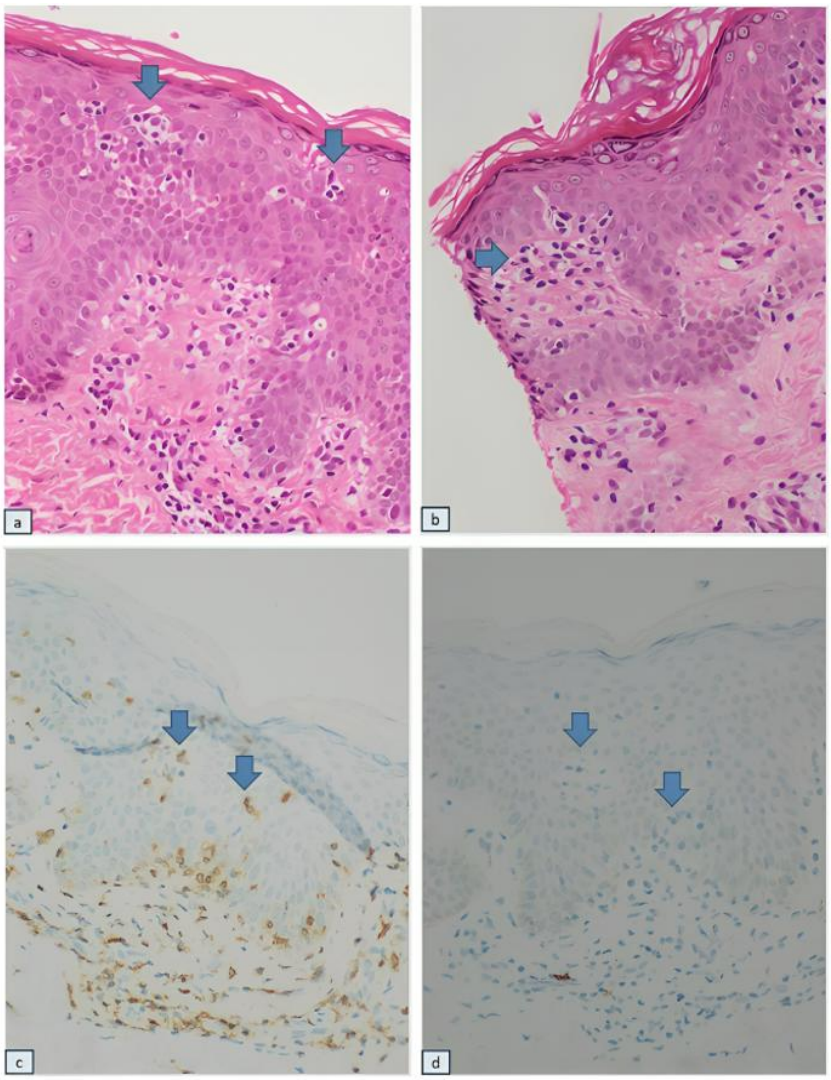

Figure 2. A skin biopsy showed (a) singly distributed intraepidermal atypical lymphocytes and small clusters (arrow) of intraepidermal atypical lymphocytes (H&E × 400); (b) basilar tagging (arrow) by the lymphocytes were also appreciated (H&E × 400); (c) the atypical lymphocytes were positive for CD3 (arrow), & (d) the atypical cells were negative for CD20 (arrow) (reprinted with permission of the parents)

A skin biopsy was arranged; the histopathological and immunohistochemistry findings are shown in **Figure 2**. The sections demonstrated singly distributed atypical lymphocytes and small intraepidermal clusters (part a in **Figure 2**), together with alignment of atypical lymphocytes along the basal epidermal layer (basilar tagging) (part b in **Figure 2**). In normal skin, lymphocytes are sparse and do not form atypical intraepidermal clusters or conspicuous basilar tagging. Immunohistochemistry showed that the atypical lymphocytes expressed CD3 (part c in **Figure 2**) but not CD20 (part d in **Figure 2**), supporting a T-cell rather than B-cell lineage. In conjunction with the clinical presentation, these findings were consistent with HMF.

The patient started on a mid-potency topical steroid, betamethasone valerate cream 0.05% twice daily over the trunk and limbs, pimecrolimus cream twice daily over the face, and an emollient. After 6 months of treatment, the patient still developed a new hypopigmented lesion. Subsequently, treatment was maintained with tacrolimus 0.03%, applied twice daily over the trunk, limbs, and face. The patient was given a subsequent dermatology follow-up.

DISCUSSION

HMF is a rare variant of CTCL that clinically presents with hypopigmented macules and patches, typically occurring over the extremities. The lesions are usually mistaken for other forms of hypopigmented lesion, leading to misdiagnosis and delayed diagnosis, especially in a primary care setting where the condition is rare and not widely recognized [6].

The diagnostic evaluation of a hypopigmented lesion begins with a thorough clinical assessment. The demographic background of the patient and the clinical history of the lesion are crucial in distinguishing various conditions that cause hypopigmented lesions. The approach is well described in [7], who mentioned that the patients' and family's background, courses of the lesion, overall health history, and any trauma or chemical exposure prior to the onset of the lesion are important in narrowing down the possible diagnosis. Next, the lesions need to be differentiated according to whether they are congenital or acquired and further classified as localized or diffuse.

Most patients with hypopigmented lesions will attend a primary care clinic, and they are often misdiagnosed with other conditions, such as pityriasis alba, tinea versicolor, vitiligo, and post-inflammatory hypopigmented lesions [8]. In most cases, initial investigations in primary care focus on excluding common benign conditions, often through empirical treatment with topical corticosteroids, antifungals, or emollients. However, a lack of clinical improvement after weeks to months of appropriate therapy should prompt referral for dermatological assessment as the patient may have a serious condition and a high index of suspicion of HMF needs to be raised [9].

As illustrated in this case, the patient sought medical attention on multiple occasions for his skin lesions, which progressively increased in both numbers and size over the course of 1 year. Despite repeated empirical treatment given in the primary care setting, the lesions worsened and only showed partial improvement; subsequently the patient was referred to the dermatology department for further evaluation. A skin biopsy was performed, which revealed histopathological and immunohistochemical features consistent with HMF, thereby establishing the definitive diagnosis.

MF staging typically incorporates skin involvement (patch/plaque distribution), lymph node status, blood involvement (circulating Sézary cells), and visceral organ involvement and it has to be aligned with the TNMB staging system that has been proposed by the International Society for Cutaneous Lymphoma (ISCL) and European Organization for Research and Treatment of Cancer (EORTC) [10].

The treatment options for MF vary according to the stage. For early-stage disease with no extracutaneous involvement, skin-directed therapies may be suitable options for patients. These ranges from topical steroids and topical tacrolimus to phototherapy with narrow-band UVB and PUVA. For systemic treatment, which is used mainly in advanced-stage disease, the most commonly used therapeutic modalities include steroids and biological agents. The prognosis of early-stage MF is good, with a 95% of 5-year of survival rate [11].

In relation to the case presentation, he was diagnosed with an early-stage of MF, as examination showed no extracutaneous involvement; hence, the patient was treated with a topical steroid at initial diagnosis. However, due to the partial response to the topical steroid treatment was maintained with a topical calcineurin inhibitor.

Compared with the published case in [12] involving a 7-years-old boy, our patient presented at a younger age; however, the case shared a similar clinical course marked by a diagnostic delay of approximately one year, despite multiple healthcare encounters. This observation suggests that delayed diagnosis in HMF reflects the subtle and nonspecific nature of its clinical appearance, which closely resembles common benign dermatoses. The disease initially manifested as erythematous lesions before gradually evolving into hypopigmented patches. This early transition, well described in HMF, may represent a crucial but easily overlooked warning sign and highlights the importance of maintaining clinical vigilance when skin lesions persist or evolve over time.

CONCLUSION

HMF is a rare but important pediatric variant of CTCL. It can resemble common hypopigmented conditions, which often leads to delays in diagnosis. In this case, a 4-year-old boy with progressively spreading hypopigmented lesions ultimately had HMF confirmed by skin biopsy. This case highlights several practical takeaways, especially for primary care doctors, as they are commonly the first to encounter the patient. These include being vigilant for MF in children with persistent, non-responsive hypopigmented lesions; early dermatologist input, and timely skin biopsy are crucial for an accurate and timely diagnosis. Once the diagnosis of HMF has been made, it must be aligned with the ISCL/EORTC TNMB criteria to guide treatment decisions. Importantly, when MF is detected at an early stage, it generally has a favorable prognosis with skin-directed therapies such as topical steroids, calcineurin inhibitors, or phototherapy.

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AI statement: The authors stated that generative AI tools were used only for language refinement and grammar correction during manuscript preparation. The authors reviewed, edited and took full responsibility for the scientific content, interpretation, and accuracy of the manuscript. No AI tools were used for data.

Declaration of interest: No conflict of interest is declared by the authors.

Data sharing statement: Data supporting the findings and conclusions are available upon request from the corresponding author.

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