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Indian Journal of Postgraduate Dermatology

Case Report

Hypopigmented Mycosis Fungoides Presenting as a Single Large Patch in a Middle-Aged Woman – An Unusual Presentation

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Received: 06 January 2024
Accepted: 09 July 2024
Published: 23 August 2024

DOI
10.25259/IJPGD_3_2024

Quick Response Code:

ABSTRACT

Hypopigmented mycosis fungoides (HMF) is a rare variant of mycosis fungoides which presents as achromic lesions commonly affecting adolescents. A 50-year-old female presented with a single large atrophic hypopigmented patch of size 30 cm × 20 cm over inner aspect of the right upper thigh of 2 years duration. Histopathology showed prominent basilar epidermotropism with lymphoid cells invading the spinous layer; immunohistochemistry staining showed CD3, CD5 and CD8 positivity. The case was diagnosed as HMF and treated with narrowband ultraviolet B phototherapy, methotrexate 10 mg/week and betamethasone dipropionate 0.05% cream resulting in moderate clinical improvement. Assessment at 24 weeks showed no progression of disease. The present case highlights atypical HMF presenting with a single diffuse hypopigmented patch in a 50-year-old female. Although the prognosis for HMF is good, it has malignant potential and should always be treated optimally with long-term follow-up.

Keywords: Hypopigmented mycosis fungoides, Cutaneous T-cell Lymphoma, Single large patch, Atypical presentation

INTRODUCTION

Hypopigmented mycosis fungoides (HMF) is a rare variant of mycosis fungoides which presents as hypopigmented-to-achromic lesions mainly distributed on the trunk and proximal extremities usually in adolescents and younger adults unlike conventional mycosis fungoides (MF), which is mostly seen in the fifth to sixth decades of life.[1] We report an atypical case of hypopigmented MF in a 50-year-old woman presenting with a single large hypopigmented atrophic patch of 2 years duration, who responded favourably to treatment with arrest in the progression of disease.

CASE REPORT

A 50-year-old female presented with a single asymptomatic whitish patch over the inner aspect of upper right thigh since 2 years. Initially, the lesion started as a small macule of size 1cm diameter which gradually enlarged over 2 years with thinning of the skin over the central part of the lesion since past 8 months. General examination and systemic examination were within normal limits.

On dermatological examination, a single diffuse large hypopigmented patch of size 30 cm × 20 cm was seen over the inner aspect of the right upper thigh, just below the inguinal fold. The margins of the patch showed ill-defined borders and prominent hypopigmentation compared to the central part of the patch where severe atrophy, dry wrinkled appearance and irregular hyperpigmentation are shown [Figure 1]. Sensation to touch, temperature and pain were preserved over the patch.

A differential diagnosis of Vitiligo, lichen sclerosis et atrophicus, borderline tuberculoid leprosy and HMF were considered. Complete laboratory workup showed normal hemogram, peripheral smear, serum biochemistry, viral screening, chest X-ray and ultrasonography of abdomen. Histopathological examination revealed band-like monomorphic lymphoid infiltrates within the upper dermis showing prominent epidermotropism without any spongiosis [Figure 2a]. Immunohistochemistry (IHC) staining showed that most of the lymphoid cells expressed CD3, CD5 and CD8 [Figure 2b] and were negative for CD4, CD7 and CD20.

Based on the clinical presentation of a hypopigmented atrophic patch, absence of frank depigmentation and aggressive clinical behaviour along with monomorphic lymphoid cells with epidermotropism showing positivity for mature T-cell markers (CD3, CD5), a diagnosis of CD4−/CD8+ HMF was made. The patient was treated with narrowband ultraviolet B phototherapy, weekly methotrexate 10 mg along with betamethasone diproprionate 0.05% cream at night for 12 weeks which resulted in moderate clinical improvement [Figure 1]. Assessment after 24 weeks showed no progression of disease with absence of lymphadenopathy, atypical cells in peripheral smear or evidence of systemic involvement.

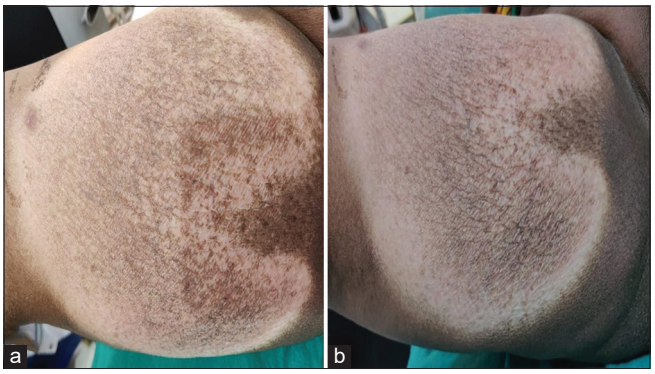

Figure 1: (a) Single large hypopigmented patch of size 30 cm × 20 cm seen over the inner aspect of the right upper thigh with prominent hypopigmentation over the margins and central atrophy. (b) Moderate improvement in hypopigmentation and atrophy noticed after 12 weeks of treatment.

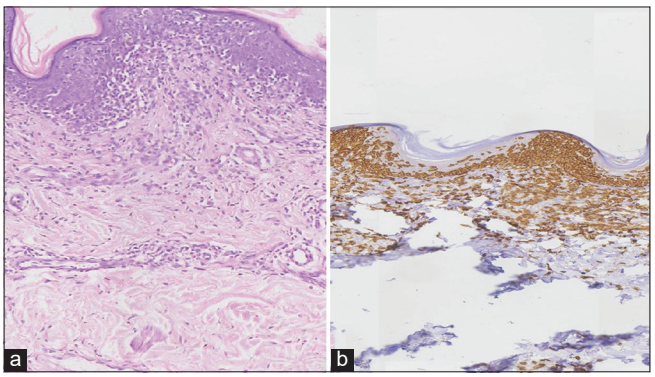

Figure 2: (a) Epidermal thinning, prominent basilar epidermotropism and lymphoid cells invading the spinous layer (haematoxylin and eosin, ×10) (b) Immunohistochemistry staining shows strong CD-8 positivity of lymphocytes (×4).

DISCUSSION

HMF has a significantly lower incidence than the classical mycosis fungoides and usually presents with multiple hypopigmented lesions of varying sizes. It may account for 17–59% of all childhood MF cases and is usually reported in patients with Fitzpatrick skin types IV to VI.[2] The location of patch in the present case over upper thigh was in agreement with the typical features of HMF, but clinical presentation with a single large hypopigmented atrophic patch and age (50 years) was atypical features.

Salient histopathological features seen in HMF include prominent epidermal and dermal lymphocytic infiltrate; epidermotropism is predominantly by neoplastic CD8+ T-cells in contrast to classical MF where neoplastic CD4+T-cells predominant.[3] Hypopigmentation represents a Th1 protective immune response with neoplastic or reactive CD8+ T-cells in cellular infiltrate which favours a benign disease with good prognosis. In our case, prominent basilar epidermotropism, lymphoid cells invading the spinous layer and CD3, CD5 and CD8 positivity on IHC staining favoured HMF. Mechanisms explaining the loss of pigmentation in HMF include degeneration of melanocytes, defective transfer of melanosomes and abnormal melanogenesis.[4]

Most HMF patients get diagnosed during early patch stage which lasts for many years and does not progress beyond stage IB, that is skin lesions covering more than 10% of body surface area (BSA) without involvement of lymph node or viscera. Despite favourable prognosis, awareness regarding potential lethality of HMF is vital. Thorough clinical evaluation with complete staging is necessary and should include physical examination of peripheral lymph nodes, peripheral blood examination, quantification of Sezary cells, T-lymphocytes using flow cytometry and imaging studies to exclude visceral involvement. Treatment modalities with

good efficacy in HMF include phototherapy, topical nitrogen mustard and total skin electron beam therapy.[5] In the present case, the patient was treated with methotrexate along with phototherapy based on practical considerations resulting in clinical improvement and arrest in disease progression.

CONCLUSION

The present case highlights atypical HMF presenting with a single diffuse hypopigmented patch in a 50-year-old female. Although the prognosis for HMF is good, it has malignant potential and should always be treated optimally with meticulous follow-up over long-term considering its high recurrence rate.

Ethical approval

Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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How to cite this article: Tarra R, Gopal KVT, Babu KS, Bulla RR. Hypopigmented Mycosis Fungoides Presenting as a Single Large Patch in a Middle-Aged Woman – An Unusual Presentation. Indian J Postgrad Dermatol. 2024;2:132-4. doi: 10.25259/IJPGD_3_2024