International Journal of Medical Science in Clinical Research and Review
Online ISSN: 2581-8945
Available Online at https://ijmscrr.in/ Volume 8|Issue 01 (January-February) |2025 Page: 42-44

Case Report

Hyperpigmented Mycosis Fungoides: A Rare Variant with Diagnostic Challenges

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Article Received: 28-December-2024, Revised: 18-January-2025, Accepted: 08-February-2025

ABSTRACT:
Hyperpigmented mycosis fungoides (MF) is a rare variant of cutaneous T-cell lymphoma, often misdiagnosed due to its resemblance to inflammatory dermatoses. We report a 77-year-old lady with a 12-year history of pruritic, hyperpigmented rashes initially misdiagnosed as chronic eczema. Diagnosis was confirmed after multiple biopsies, leading to treatment with methotrexate and PUVA therapy, resulting in remission. This case highlights the challenges in diagnosing atypical MF variants, emphasizing the need for early clinico-pathological correlation. Increased awareness of hyperpigmented MF can aid in reducing diagnostic delays and improving patient outcomes.

Key words: Mycosis Fungoides, Cutaneous T-Cell Lymphoma, Hyperpigmentation, Skin Neoplasms

INTRODUCTION:

Mycosis fungoides (MF) is the most common cutaneous T-cell lymphoma, often misdiagnosed due to its indolent nature and resemblance to inflammatory dermatoses. It progresses through patch, plaque, and tumor stages over several years. Hyperpigmented MF is a rare variant, presenting as pigmented macules and patches, often lacking erythema and scaling. Unlike classic MF, which typically has a CD4+ phenotype, hyperpigmented MF is often CD8+ and associated with a better prognosis. It is more frequently reported in younger individuals and those with darker skin tones.

We present a 77-year-old Middle Eastern lady with a 12-year history of progressive, pruritic hyperpigmented lesions, initially misdiagnosed as chronic eczema. Diagnostic delay due to inconclusive biopsies and prolonged treatment with topical steroids led to a late MF diagnosis. This case highlights the need for early clinico-pathological correlation and increased awareness of atypical MF variants to prevent misdiagnosis.

Case Description:

We report the case of a 77-year-old Middle Eastern lady with diabetes, hypertension, and knee osteoarthritis, who presented with a 12-year history of pruritic, hyperpigmented skin rashes. Initially misdiagnosed as chronic eczema, she was treated with antihistamines and topical steroids, which provided only transient improvement. Over time, her condition progressed, and a skin biopsy in 2021 led to the diagnosis of mycosis fungoides. She was then started on definitive treatment, resulting in remission.

The patient reported a 12-year history of intensely pruritic skin rashes affecting multiple areas, including the face, neck, forearms, anterior abdomen, back, thighs, and distal lower extremities. The condition severely impacted her quality of life, confining her to her home for the past five years. The lesions were characterized by persistent hyperpigmentation and itching but were notably absent of pain, ulceration, sinus formation, or discharge.

On examination, multiple hyperpigmented, annular, lichenoid, and confluent macular lesions were observed. There was no evidence of lymphadenopathy or hepatosplenomegaly. Routine laboratory investigations, including complete blood count, renal function, and liver function tests, were within normal limits.

Initially, the patient was clinically diagnosed with chronic eczema and managed with topical steroids, depigmenting creams, and antihistamines. Although she showed some initial improvement, her response to treatment waned over time. A skin biopsy performed five years after symptom onset was inconclusive. Despite multiple consultations, the diagnosis remained elusive, and she continued self-medicating with topical steroids and antihistamines.

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Four years later, she experienced worsening hyperpigmentation, involving more than 10% of the total body surface area (BSA), along with intense pruritus that became refractory to conventional treatments. Repeat biopsies in 2021 and 2022 at two different centers confirmed the diagnosis of mycosis fungoides.

She was staged as T2A N0 M0 B0 and initiated on weekly intramuscular methotrexate (25 mg). Within the first month of therapy, she showed significant symptom resolution. Treatment was continued for nearly a year, after which she was transitioned to psoralen plus ultraviolet A (PUVA) phototherapy. She remains in remission with ongoing PUVA therapy.

DISCUSSION:

Mycosis fungoides (MF) is the most common form of cutaneous T-cell lymphoma and the second most common extranodal non-Hodgkin's lymphoma. It is a rare lymphoma of T- and B-cell origin, with the skin being the primary site of involvement. MF is characterized by malignant T-cell infiltration of the skin and other organs, leading to progressive cutaneous and systemic involvement. The disease progresses through three clinical phases patch, plaque, and tumor stages over years or decades. It primarily affects middle-aged and elderly adults of all races, presenting with persistent or slowly progressive skin lesions of varying sizes and shapes.

Early diagnosis of MF can be challenging. The time from symptom onset to diagnosis varies between 2 and 4.2 years (1,2), with the PROCLIPI STAGE MS study reporting a median diagnostic delay of 36 months in early-stage MF (2). The diagnostic challenge arises from inconsistencies between clinical presentation and pathological features, as well as overlap with reactive and inflammatory dermatoses (3). Early-stage MF often presents with pruritus and erythematous scaly patches, which can resemble other inflammatory conditions, increasing the likelihood of misdiagnosis. Pruritus, affecting 80% of MF patients (4), is the most common symptom and significantly impacts their quality of life. Clinically, MF can be mimicked by other biopsy-proven inflammatory diseases such as eczema, psoriasis, nonspecific dermatitis, lichen planus, lupus, and parapsoriasis (5,6). Diagnosing MF based on clinical or pathological criteria alone is insufficient. Therefore, a combination of medical history, clinical evaluation, and repeat histopathological assessment is essential to avoid misdiagnosis (7). Additionally, frequent self-medication can alter lesion appearance, further complicating diagnosis, and the use of topical steroids and systemic immunosuppressants may modify histological findings (3).

Besides the classic MF (Alibert-Bazin type) and the three commonly described variants (folliculotropic MF, pagetoid reticulosis, and granulomatous slack skin), other clinicopathological subtypes include hypopigmented, poikilodermatous, erythrodermic, granulomatous, hyperpigmented, ichthyosiform, syringotropic, papular, purpuric, interstitial, pustular, bullous, verrucous, and psoriasiform MF(4,5,8). Over the past 30 years, several atypical variants have been described, sometimes with distinct prognostic and therapeutic implications(6). These variants can mimic benign inflammatory skin disorders, making multiple biopsies from various lesions and detailed clinical-pathological correlation essential for diagnosis (5,9).

Hyperpigmented MF is an extremely rare subtype, presenting as multiple pigmented macules and patches without poikilodermatous changes. It is characterized by a CD8+ phenotype on immunohistochemistry, whereas classic MF typically shows a CD4+ phenotype. This subtype mainly affects individuals with darker skin tones and is more common in younger patients (<35 years old) than classic MF (10,11,12). Hyperpigmented MF generally has a better prognosis than classic MF (13). Lee JS et al. reported a 42-year-old patient with steadily progressing hyperpigmentation that evolved into a purpuric plaque over 17 years (6). Similarly, Ying Yi Lu et al. described a 62-year-old patient with a seven-year history of hyperpigmented MF that remained non-progressive (13).

Our patient, presented at the age of 65 with an atypical variant of MF, characterized by 12 years of progressive hyperpigmentation and pruritus, without advancing to plaque or tumor stage. Initially misdiagnosed as chronic eczema, she was treated with antihistamines and topical steroids without early biopsy, leading to a delayed diagnosis. The first biopsy, conducted five years later, was inconclusive, and subsequent treatment with stronger steroids and antihistamines did not resolve the issue. This case highlights the importance of clinico-pathological correlation in early MF diagnosis, as delayed biopsy and prolonged treatment contributed to the diagnostic delay.

Additionally, this case underscores the need for long-term follow-up in hyperpigmented MF, as it may eventually progress. While hyperpigmented MF is more commonly reported in younger individuals, it can occur at any age. The patient's steady progression of hyperpigmentation and pruritus over 12 years, without progression to plaque or tumor stage, suggests a potentially non-aggressive course. We conclude that marked hyperpigmentation of prolonged duration in such cases may represent a distinct variant of MF rather than simply being a characteristic of the disease.

Mycosis fungoides (MF) remains a diagnostic challenge, particularly in its early stages, due to its clinical overlap with inflammatory dermatoses and non-specific histopathological findings. This case of a 77-year-old

woman with a 12-year history of pruritic, hyperpigmented skin lesions, initially misdiagnosed as chronic eczema, highlights the risk of delayed diagnosis in atypical MF variants. Despite initial symptomatic relief with topical steroids and antihistamines, her condition gradually worsened, and an early biopsy was inconclusive. A definitive diagnosis of hyperpigmented MF was made only after repeat biopsies, emphasizing the need for serial histopathological assessment in suspected cases.

Hyperpigmented MF is a rare subtype, often associated with CD8+ immunophenotype and a potentially indolent course, yet long-term progression remains uncertain. The patient responded well to methotrexate and PUVA therapy, achieving remission. This case underscores the importance of early clinical suspicion, repeated biopsies, and long-term monitoring in chronic, treatment-resistant skin conditions to improve diagnostic accuracy and patient outcomes.

Financial support: Nil

Conflict of Interest: Nil

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