

## Cutaneous T-Cell Lymphoma (Mucosal Fungoid) as First Manifestation of Metastatic Lymphoma

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### ABSTRACT

Primary cutaneous T-cell lymphomas are a diverse group of extranodal non-Hodgkin lymphomas characterized by the infiltration of malignant T-cells within the skin. Mycosis fungoides (MF), the most common subtype, typically presents with skin-limited manifestations. However, in a subset of cases, it can herald the onset of systemic lymphoma, representing a diagnostic challenge for clinicians.

This article explores a compelling clinical case of primary cutaneous T-cell lymphoma, specifically mycosis fungoides, manifesting as an initial sign of disseminated or metastatic lymphoma. We dissect the complex diagnostic and management considerations associated with this unique clinical presentation. Through a review of the current literature and clinical insights, we delineate the clinical, histopathological, and immunophenotypic features that facilitate the diagnosis and differentiation of MF as an isolated cutaneous entity from MF with secondary extracutaneous involvement.

The article highlights the critical role of multidisciplinary collaboration among dermatologists, oncologists, and pathologists in achieving timely and accurate diagnosis and guiding therapeutic decisions. It further underscores the evolving landscape of treatment options for this challenging subset of patients, ranging from skin-directed therapies to systemic agents. Additionally, this case provides a stark reminder of the importance of ongoing surveillance, as the clinical course of MF may unpredictably transition to a more aggressive form.

In conclusion, this case serves as a vivid illustration of mycosis fungoides with mucosis fungoides as its initial presentation, challenging the conventional understanding of primary cutaneous T-cell lymphomas. The intricate interplay of clinical, histological, and immunophenotypic data informs the diagnosis and informs the broader clinical community about the potential metamorphosis of cutaneous lymphomas into systemic lymphomas. Understanding such cases is crucial for early intervention and improved patient outcomes.

**KEYWORDS:** Cutaneous, T-cell, lymphomas, extranodal.

### ARTICLE DETAILS

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### INTRODUCTION

Mucosal fungoid, a rare variant of cutaneous T-cell lymphoma, usually presents chronically in the skin, but its initial manifestation as a metastatic lymphoma is exceptionally rare and poses unique diagnostic and therapeutic challenges. This article presents a case report that illustrates the relevance of early clinical suspicion, accurate

diagnosis and therapeutic implications in this clinical setting.<sup>1,2</sup>

In the context of cutaneous lymphomas, which constitute 3.9% of non-Hodgkin's lymphomas, mycosis fungoides is considered the most common type of cutaneous T-cell lymphoma. It mainly affects male patients aged 55 to 60 years, and most of them do not progress to a tumor stage.<sup>3,4</sup>

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### OBJETIVE

The fundamental purpose of this clinical case article is to comprehensively address and analyze a rare and challenging scenario in medical practice: cutaneous T-cell lymphoma, specifically mycosis fungoides, presented as the first manifestation of metastatic lymphoma. Our goal is to present a detailed analysis of this exceptional case, highlighting the clinical, histopathological and molecular features that define it, with the aim of enriching the understanding of this rare entity and, at the same time, providing a valuable contribution to the medical literature.

### PRESENTATION OF THE CASE

Male, 60 years old, from Chihuahua, Mexico, occupation: electrical technician, married, with no relevant hereditary or personal history. He started in December 2022 with erythematous, pruritic skin lesions on the trunk and lower extremities and brownish plaques with fine scales, accompanied by icteric staining of skin and integuments.(Figure 1) He also reported unintentional weight loss of 12 kg in 4 months and progressive left hemiparesis.

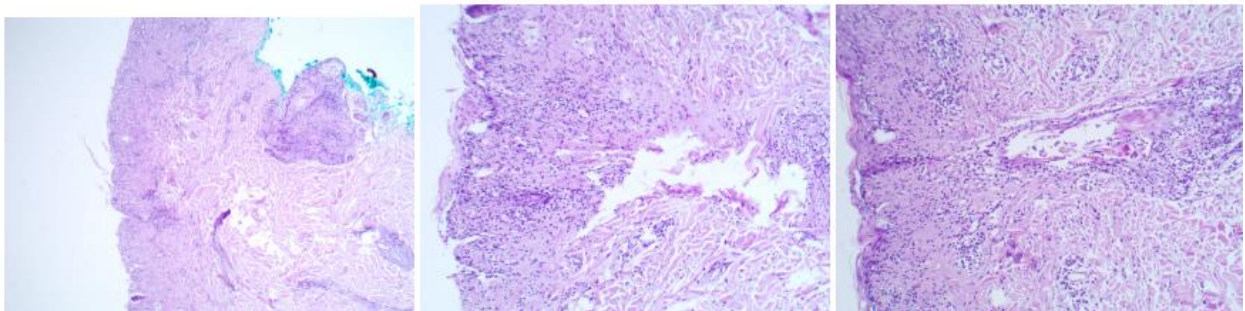


**Figure 1. Cutaneous lesions on the trunk, legs and arms.**

Laboratory tests: Elevation in liver function tests, brain MRI where demyelinating lesion of right pyramidal pathway is documented.

A skin lesion biopsy was performed: The histologic sections performed show a skin spindle involved by an atypical lymphocytic infiltrate, localized in the dermoepidermal

junction, with few lymphocytes migrating to the epidermis the dermoepidermal junction, with few lymphocytes migrating into the epidermis, with areas of vacuolization of the basal layer and evidence of pigment incontinence.(Figure 2)

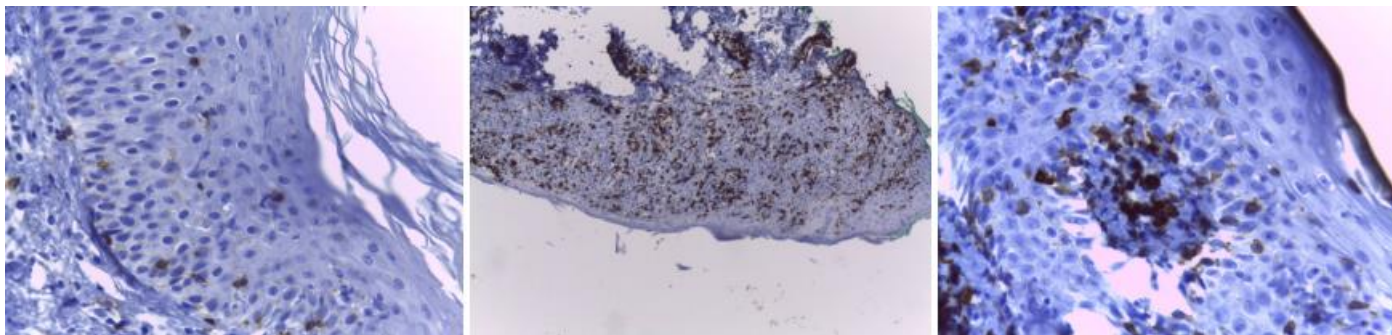


**Figure 2. Skin biopsy images.**

With immunohistochemical report CD-4:Positive 3 + in 70% of lymphocytes, CD -8: Positive 3 + in 10% of lymphocytes, CD -5 : Positive 3 +, CD- 7 Positive 3 + , With these data we

conclude that it is an atypical infiltrate with immunophenotype of mycosis fungoides in plaque phase.(Figure 3)

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**Figure 3. Immunohistochemical images.**

Patient with T-cell neoplasm with CNS, liver and skin infiltration. It was decided to manage CHOP chemotherapy, Cyclophosphamide 1150 mg, Doxorubicin 80 mg, Vincristine 2 mg, Prednisone 50 mm, also receiving holocranial palliative radiotherapy 3DCRT technique, dose 20 Gy in 5 fractions.

### CONCLUSIONS

The patient in the present case has central nervous system involvement, characterized by gait disturbance, left hemibody weakness, evidenced by brain MRI lesion of the pyramidal pathway. Case of mycosis fungoides with visceral involvement TNM (T2/N0/M1) Clinical stage IV B due to CNS infiltration.

This clinical case raises fundamental questions about mucosal fungoid, a T-cell lymphoma that usually affects the skin. How can it evolve into a metastatic form in lymph nodes or other tissues? This question suggests the possibility of phenotypic reprogramming of neoplastic T cells in mucosa fungoides, which could allow their spread to other tissues. Further genomic and molecular studies are required to elucidate this phenomenon. 4,5

The presentation of mucosa fungoides as a metastatic lymphoma presents significant diagnostic challenges, as it is uncommon to initially consider this condition in the diagnosis of skin lesions with unusual manifestations. This emphasizes the importance of maintaining high clinical suspicion and performing repeated biopsies and immunohistochemical testing to confirm the diagnosis. 6

The therapeutic management of metastatic fungoid mucosis is particularly complex and varies according to the extent of disease and organ involved. Radiotherapy, chemotherapy, and biologic agents may be options in select cases, but the lack of standardized therapeutic guidelines underscores the need for an individualized approach and interdisciplinary discussion in the management of these patients.7

This case underscores the importance of considering fungoid mucosis in the differential diagnosis of atypical skin lesions

and highlights the need for greater awareness and understanding of this rare presentation of metastatic lymphoma in the medical community. Furthermore, it emphasizes the relevance of collaboration between dermatologists, oncologists and pathologists in the evaluation and effective management of patients with metastatic fungoid mucosis, which can significantly influence the prognosis and quality of life of these individuals.7,8

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