Title

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Authors

Ojeaburu, Leslie
Larsen, Tyler

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Mycosis Fungoides

Leslie Ojeaburu[1], Tyler Larsen[1,2]

[1] Medicine, University of California, Los Angeles,
[2] Medicine, VA Greater Los Angeles Healthcare System

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Abstract

Mycosis fungoides, also known as cutaneous T-Cell lymphoma, is a rare hematologic malignancy characterized by cutaneous involvement. There is significant variability in dermatologic presentation in early stages and often atypical pathology findings on biopsy that often make early diagnosis quite challenging. Here we present a case of an elderly patient who presented with chronic pruritis and cutaneous T-Cell Lymphoma involving the entire body, currently managed with a combination of phototherapy and anti-pruritic topical medications.

A man in his 70s with prostate cancer, post-traumatic stress disorder, and bipolar disorder type I presented to the emergency department with one week of suicidal ideation for which he was admitted to the psychiatry service. The Internal Medicine service was consulted for intractable pruritis which contributed to his psychological distress. Dermatologic examination revealed diffuse thick, waxy, hyperpigmented patches and scaling plaques involving over eighty percent of his body surface area with scaling on his palms and soles.

On interview the patient reported that the rash initially started on his legs almost a decade prior to this presentation and was diagnosed as atopic dermatitis. However, the rash failed to improve on various therapies including dupilumab and quickly spread to cover over fifty percent of his body surface area. A repeat biopsy six months prior to his current presentation subsequently showed "atypical epidermotrophic small to medium lymphocytes with irregular nuclear contours" consistent with mycosis fungoides plaque type. The patient trialed several treatments including methotrexate, mycophenolate, and bexarotene with only minimal improvement and symptomatic relief. He reported that ultraviolet phototherapy was the most effective treatment and gave some modest improvements in symptoms. Dermatology consultants during this admission recommended topical emollients to manage his pruritis as well as continued systemic treatments and scheduled UV phototherapy. Following his discharge, the patient had follow up with oncology where peripheral flow cytometry failed to identify any clonal T-cell population. Given the refractory nature of his symptoms and progressive disease the patient underwent a trial of mogamulizumab and then brentuximab vedotin with only modest response.

Mycosis fungoides is the most common manifestation of cutaneous T cell lymphoma.[1,2] With a global incidence greater than five million people, mycosis fungoides is most common in patients over the age of fifty and in males. The disease typically presents with pruritis and non-specific dermatologic findings like plaques, hyperpigmented lesions, or alopecia, and it can progress to overt tumors or systemic disease in the later stages. The cutaneous findings of mycosis fungoides may be widespread throughout the body, including the palms or soles in about ten percent of cases, which may lead to it being confused for other dermatologic pathologies including atopic dermatitis, plaque psoriasis, and cutaneous infections.[2] Diagnosis relies on histopathology findings on skin biopsy such as monoclonal expansion of atypical CD4+ cerebriform T-Cells, epidermotropism, and pautrier abscesses.[1] A high suspicion of a malignant process (i.e., presentation in older patient, resistant to therapy) and integration of the clinical presentation is needed in cases with non-characteristic pathology findings.[1]

Treatment anchors on a combination of topical agents (corticosteroids, nitrogen mustard, bexarotene), UV phototherapy, and systemic chemotherapies and biologics.[1,3] Patients with widespread disease refractory to topical or localized treatments and patients with leukemic involvement of their cutaneous T cell lymphoma (Sezary syndrome) may benefit from newer biologic therapies targeting T cell receptors such as mogamulizumab, a humanized monoclonal antibody which targets CC chemokine receptor 4 (CCR4), and brentuximab vedotin which targets the cell membrane protein CD30.[3] Unfortunately, treatment resistant mycosis fungoides is common and recurrence is a normal phenomenon in the majority of cases.

Figure 1. Diffuse hyperpigmented patches and hyperkeratotic plagues in Mycosis Fungoides.

Author Contributions

All authors have reviewed the final manuscript prior to submission. All the authors have contributed significantly to the manuscript, per the International Committee of Medical Journal Editors criteria of authorship.

- Substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data for the work; AND
- Drafting the work or revising it critically for important intellectual content; AND
- Final approval of the version to be published; AND
- Agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Disclosures/Conflicts of Interest

The authors declare they have no conflicts of interest

Corresponding Author

Tyler Larsen, MD
11301 Wilshire Blvd
Building 500, Suite 3214-B
Los Angeles, CA 90073
tlarsen@mednet.ucla.edu
310-478-3711 x47031

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