ISSN: 2584-0355
DOI: doi.org/10.51219/MCCRJ/Ángeles-Manteca/81

URF PUBLISHERS
connect with research world

Medical & Clinical Case Reports Journal

https://urfpublishers.com/journal/case-reports

Vol: 2 & Iss: 2

Case Report

Case Report, Erythroderma and Dactylitis in Mycosis Fungoides

Ángeles Manteca[1*], Daniela Caicedo[2], Verónica Posso[3], Ramírez Lisseth[1], Suárez Efraín[1], Vergara Eduardo[1], Riofrío Ivonne[1], Echeverria Julian[1], Bastidas Jacqueline[1], Cueva Andrea[1] and Augusta Basantes[1]

[1]Dermatology Resident, Universidad Tecnológica Equinoccial. Quito, Ecuador

[2]Department of Dermatology, Hospital Carlos Andrade Marín, Universidad Central del Ecuador, Quito, Ecuador

[3]Department of Dermatopathology, Hospital Carlos Andrade Marín, Universidad UTE, Quito, Ecuador

Citation: Manteca A, Caicedo D, Posso V, et al. Case Report, Erythroderma and Dactylitis in Mycosis Fungoides. *Medi Clin Case Rep J* 2024;2(2):305-307. DOI: doi.org/10.51219/MCCRJ/Ángeles-Manteca/81

Received: 23 May, 2024; **Accepted:** 29 May, 2024; **Published:** 03 June, 2024

***Corresponding author:** Ángeles Manteca, Carlos Andrade Marín Hospital, University UTE, Quito, Ecuador, Tel: +593 987549106; E-mail: angelitos89@hotmail.com

ABSTRACT

MF is a type of mature T cell non-Hodgkin lymphoma that typically appears in the skin, but it can also affect the blood, viscera, and nodes. Tumors, erythroderma, patches and plaques that might be localized or widespread are examples of skin lesions. Although the exact origin of MF is unknown. Common characteristics include epigenetic changes, aberrant RNA splicing, altered JAK-STAT signaling, and T cell receptor (TCR)/T cell activation.

We present the case of a 65-year-old male with many episodes of Erythroderma in addition to moderate pruritus and dactylitis, in the last 2 years. At Hospital Carlos Andrade Marin in Quito, Ecuador.

Keywords: Erythroderma; Mycosis fungoides; Dactylitis

Abbreviations: CTCL: Cutaneous T cell lymphoma; MF: Mycosis Fungoides; SS: Sézary Syndrome

Introduction

Erythroderma, also known as exfoliative dermatitis, is a severe and sometimes fatal disorder that manifests as diffuse erythema and scaling over 90% of the skin's surface area. Erythroderma can be the clinical manifestation of many different systemic and cutaneous disorders (such as atopic dermatitis and psoriasis), medication hypersensitivity reactions, and, less frequently, Sézary syndrome, a leukemic subtype of cutaneous T cell lymphoma[1-3]. Within cutaneous T cell lymphoma (CTCL), Mycosis fungoides (MF) and Sézary syndrome (SS) are the most prevalent subtypes[3].

MF is a type of mature T cell non-Hodgkin lymphoma that typically appears in the skin, but it can also affect the blood, viscera, and nodes. Tumors, erythroderma, and patches or plaques that might be localized or widespread are examples of skin lesions.(3) Although the exact origin of MF is unknown, common characteristics include epigenetic changes, aberrant RNA splicing, altered JAK-STAT signaling, and T cell receptor (TCR)/T cell activation[3,4].

Patients with poikilodermatous skin abnormalities, generalized erythroderma, or chronic nonspecific dermatitis may be suspected of having MF. A skin biposy hast he highest yield to help the physician diagnose MF; several skin biopsies are frequently needed. The lesional skin with the highest degree of induration should be included in the single biopsy, if one is taken. A 4 mm punch biopsy at the very least is advised. Biopsies can usually be preserved in formalin before being stained

with hematoxylin and eosin or subjected to further pathological testing. Individuals from endemic areas should have serologic testing for HTLV-I[3,5,6].

The European Organization for Research and Treatment of Cancer (EORTC) cutaneous lymphoma task force and the International Society for Cutaneous Lymphoma (ISCL) have proposed an algorithm for diagnosing and staging early MF based on clinical, histopathologic, molecular, and immunopathologic criteria. It is advised that the ISCL/EORTC algorithm be used to confirm the diagnosis of every patient included in MF trials or databases[3,7,8] **(Table 1)**.

Case Presentation

We present a 65-year-old male with medical history of mixed anxiety disorder, gastritis, chronic deep vein thrombosis. He had presented progressive pruritus since approximately 63 years of age, had had two skin biopsies, for which he had been diagnosed of psoriasis. 6 months ago, our patient presented erythroderma that required hospitalization at other hospitals. The differential diagnoses included MF, psoriasis, atopic dermatitis, adverse drug reaction. He had been managed with: topical corticosteroids, methotrexate, or cyclosporine, with partial improvement.

In February 2024, he presented to the emergency room at Hospital Carlos Andrade Marín with generalized, intense pruritus, erythroderma and eczematous plaques covered with thin crust or scales. The nails were thick, as were the palms and soles and presented painful f. The 3rd left finger presented indurated edema. The eyes and ears discharged yellowish fluid **(Figure 1 and Figure 2)**. Laboratory exams were within normal range, aside from a mildly elevated eosinophilia of 1500 per cc^3. Infectious diseases were ruled out (HIV, CMV and Anti-HTLV antibodies 1 and 2 were negative). Auto antibodies including ANA, Anti-DNA, Anti-Histone, Anti-Ro/La were negative). Paraneoplastic syndromes were ruled out and included full-body CT scan, upper endoscopy, colonoscopy, occult blood in stool; and they were all negative.

Figure 1: **A-C**. Generalized erythroderma and desquamation, affecting more than 90% of the body surface.

A, B. Infiltration of the entire facial mass, auricular pavilions.

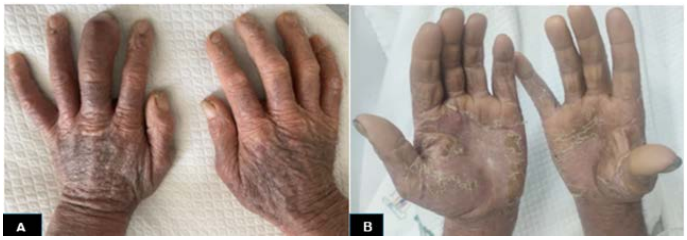

Figure 2: **A**. Marked dactylitis of the 3rd finger of the left hand. **B**. Bilateral palmar hyperkeratosis with significant scaling.

Two skin biopsies were taken, and previous biopsies were requested for comparison. The previous biopsies revealed psoriasiform and spongiotic dermatitis, and a diagnosis of psoriasis was precluded. However, immunohistochemistry was performed, and showed: epidermis with foci of parakeratosis, spongiosis and irregular hyperplasia of the ridge network, the dermis with lymphocytic infiltrate with nuclear atypia, hyperchromasia, located in the superficial dermis, some extending into the epidermis to form occasional pautrier's microabscesses."

Immnohistochemistry Results:

LCA CD3, CD4, CD5 positive in dermal and intraepidermal lymphocytes.

CD8: positive in dermal t-lymphocytes.

CD4/CD8 ratio: 4:2

CD2, CD7: scantly positive in some dermal lymphocytes.

CD30: positive greater than 25%, in this sample, 60%.

KI67: 20%.

CD20 and Granzyme B: negative.

With the results of cutaneous Atypical Lymphoid proliferation, a diagnosis of Mycosis Fungoides with CD30 greater than 25% (suggests transformation) was made.

Upon his hospitalization and due to previous treatment failure with methotrexate cyclosporine was started at a dose of 3mg/kg daily, as well as loratadine 10mg QID, hydroxyzine 10mg HS, and topical steroids and emollients. The patient presented marked improvement of his clinical manifestations, including his dactylitis at one-month review **(Figure 3)**. At the two month review, the patient started gaining weight and the skin continued to improve **(Figure 4)**.

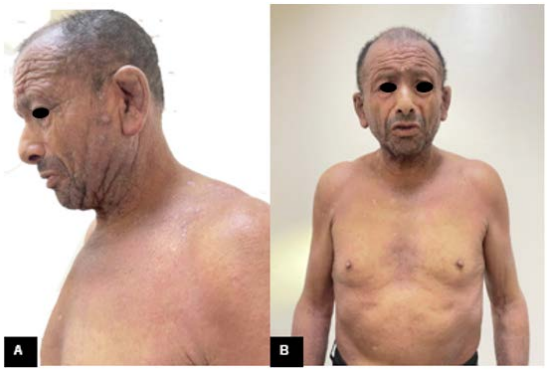

Figure 3: **A, B**. Patient one month after the last hospitalization, with improvement of lesions.

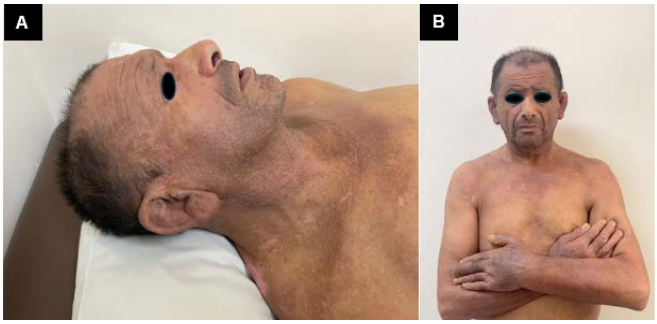

Figure 4: **A, B**. Patient at present, two months after the last hospitalization, with improvement of lesions and even weight gain.

Discussion

Patients with MF commonly present with persistent and/or slowly progressive skin lesions of varying size and shape. Skin lesions may be localized or widespread patches or plaques, tumors, and/or generalized erythroderma. The skin is often pruritic and the patient‘s quality of life can be profoundly affected[9]. Other clinical manifestations include opportunistic infections, alopecia, and, less commonly, involvement of other organs. Some of these clinical features are used in the point-based diagnostic algorithm **(Table 1)**.

Table 1: Diagnosis of early mycosis fungoides[7].

Criteria	Scoring system
Clinical	
Basic	2 points for basic criteria and two additional criteria
Persistent and/or progressive patches/thin plaques	1 point for basic criteria and one additional criterion
Additional:	
1. Non-sun-exposed location	
2. Size/shape variation	
3. Poikiloderma	
Histopathologic	
Basic	2 points for basic criteria and two additional criteria
Superficial lymphoid infiltrate	1 point for basic criteria and one additional criterion
Additional:	
1. Epidermotropism without spongiosis	
2. Lymphoid atypia*	
Molecular biologic	
1. Clonal T cell receptor gene rearrangement	1 point for clonality
Immunopathologic	
1. <50% CD2+, CD3+, and/or CD5+ T cells	1 point for one or more criteria
2. <10% CD7+ T cells	
3. Epidermal/dermal discordance of CD2, CD3, CD5, or CD7¶	

A total of 4 points is required for the diagnosis of mycosis fungoides based on any combination of points from the clinical, histopathologic, molecular biologic, and immunopathologic criteria.

* Lymphoid atypia is defined as cells with enlarged hyperchromatic nuclei and irregular or cerebriform nuclear contours.

¶ T cell antigen deficiency confined to the epidermis.

A definitive diagnosis of MF is often preceded by a „premycotic“ period ranging from months to decades, during which the patient may have pruritus and nonspecific, slightly scaling skin lesions and nondiagnostic biopsies for months to years. These lesions may wax and wane over years, and a diagnosis of parapsoriasis en plaque or nonspecific dermatitis is often made[3-8].

Evaluation

The diagnosis of MF is suspected in patients who present with chronic nonspecific dermatitis, poikilodermatous skin findings, or generalized erythroderma. Skin biopsy with routine histology is the single most important laboratory tool that will assist the clinician in establishing the diagnosis of MF(5). Often, multiple skin biopsies are required. If only one biopsy is performed, it should include the lesional skin with the most induration[3,10].

As we present this case report, it is important to emphasize the negativity of HTVL-1, reported in the clinical history, because the histologic differential diagnosis is leukemia/lymphoma and HTLV-1 positive adult T-cell Leukemia/Lymphoma[3,11].

Conclusions

The diagnosis of this case of Mycosis Fungoides is based on clinical, histopathologic, immunopathologic findings. The patient had had two years of dermatologic manifestations, as we know we must do a good anamnesis and the necessary studies for a good diagnosis and treatment. Also, we can’t forget the psychological and social impact for a holistic management[12,5].

Conflict of Interest: The authors declare no conflicts of interest.

References

1. Campo E, Jaffe ES, Cook JR, et al. The international consensus classification of mature lymphoid neoplasms: a report from the clinical advisory committee. Blood 2022;140(11):1229-1253.
2. Alaggio R, Amador C, Anagnostopoulos I, et al. The 5th edition of The world health organization classification of haematolymphoid tumours: Lymphoid neoplasms. Leukemia 2022;36(7):1720-1748.
3. Hoppe RT, Kim YH. Clinical manifestations, pathologic features, and diagnosis of mycosis fungoides. UpToDate 2024.
4. Bolognia JL, Schaffer JV, Cerroni L. Dermatology 5th edition Vols 2. Elsevier - Saunders, Mosby, Churchill 2024.
5. Fung MA, Murphy MJ, Hoss DM, Grant-Kels JM. Practical evaluation and management of cutaneous lymphoma. J Am Acad Dermatol 2002;46(3):325-357.
6. Cuellar-Barboza A, Ocampo-Candiani J, Herz-Ruelas ME. A Practical Approach to the Diagnosis and Treatment of Adult Erythroderma. Actas Dermosifiliogr (English Edition) 2018;109(9):777-790.
7. Pimpinelli N, Olsen EA, Santucci M, et al. Defining early mycosis fungoides. J Am Acad Dermatol 2005;53(6):1053-1063.
8. Ryu HJ, Kim S Il, Jang HO, et al. Evaluation of the international society for cutaneous lymphoma algorithm for the diagnosis of early mycosis fungoides. Cells 2021;10(10):2758.
9. Demierre MF, Gan S, Jones J, Miller DR. Significant impact of cutaneous T-cell lymphoma on patients’ quality of life: Results of a 2005 National Cutaneous Lymphoma Foundation Survey. Cancer 2006;107(10):2504-2511.
10. Lee H. Mycosis fungoides and Sezary syndrome. Blood Res 2023;58.
11. Karube K, Takatori M, Sakihama S, et al. Clinicopathological features of adult T-cell leukemia/lymphoma with HTLV-1-infected Hodgkin and Reed-Sternberg-like cells. Blood Adv 2021;5(1):198-206.
12. Walia R, Yeung CCS. An Update on Molecular Biology of Cutaneous T Cell Lymphoma. Front Oncol 2020;9:1558.