International Journal of Advances in Medicine
*Singh T et al. Int J Adv Med. 2015 Feb;2(1):60-63*
http://www.ijmedicine.com
pISSN 2349-3925 | eISSN 2349-3933
DOI: 10.5455/2349-3933.ijam20150215

**Case Report**

# Mycosis fungoides: cutaneous T-cell lymphoma

**Tarunveer Singh[1], Kanwarjit S. Dhillon[1]*, Deepak Sharma[1], Mohd Sadiq Umar[1], Uroos Fatima[2], K. R. Varshney[3], Alankret Dhillon[4]**

[1]Department of Dermatology, Era's Lucknow Medical College, Lucknow, Uttar Pradesh, India
[2]Department of Pathology, Era's Lucknow Medical College, Lucknow, Uttar Pradesh, India
[3]Department of Microbiology, Era's Lucknow Medical College, Lucknow, Uttar Pradesh, India
[4]III MBBS-Part B, Era's Lucknow Medical College, Lucknow, Uttar Pradesh, India

**Received:** 17 December 2014
**Accepted:** 18 January 2015

***Correspondence:**
Dr. Kanwarjit S. Dhillon,
E-mail: kanwarjit29@yahoo.co.in



## ABSTRACT

Mycosis fungoides (MF) is the most common group of cutaneous T-cell lymphomas. It is a rare non-Hodgkin's lymphoma of mature, skin-homing, clonal, malignant T lymphocytes, usually observed in mid to late adulthood, that initially presents in the skin as patches, plaques, tumors, or generalized erythema (erythroderma) and can involve the lymph nodes and peripheral blood. In this review, we survey the MF literature of the last decade and highlight the major trends.

**Keywords:** Mycosis fungoides, Cutaneous T-cell lymphoma, Epidermotropism

## INTRODUCTION

Mycosis fungoides was first described in 1806 by French dermatologist Jean-Louis-Marc Alibert.[1,2]

The name mycosis fungoides is very misleading - it loosely means "mushroom-like fungal disease". The disease, however, is not a fungal infection but rather a type of non-Hodgkin's lymphoma. It was so named because Alibert described the skin tumors of a severe case as having a mushroom-like appearance.[3]

Mycosis Fungoides (MF) is the most common group of cutaneous T-cell lymphomas. Much progress has been made in recent years in understanding the origin of the malignant T cell in MF and the pathophysiology and immunology of the disease.

This recent work has made a great impact on diagnosis, prognostication, and treatment.

## CASE REPORT

A 48 year old male, teacher by occupation came to our OPD with chief complaints of multiple red coloured round lesions (Figure 1 & 2) over his entire body and generalized itching for last 2 years.

As stated by the patient, he was alright 2 years back when he noticed small erythematous lesions over his chest. The lesions started to increase in size and number within a few months. The patient also complaint of generalized itching over the whole body. Patient went to various medical practitioners for the same but was not relieved. His Hb and RBC counts were normal but his WBC counts were raised to 32900/mm$^3$. Skin biopsy revealed dermal dense infiltrates of lymphoid tissue with focal epidermotropism (Figure 3). He was diagnosed stage 1B Mycosis fungoides ($T_2$ $N_0$ $M_0$ $B_0$) according to TNMB classification. Patient was started on NBUVB therapy and Interferon-α.

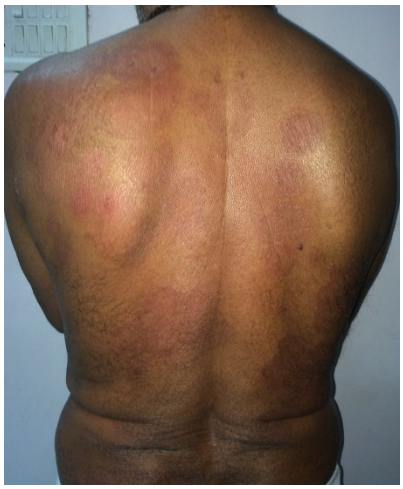

**Figure 1: Erythematous macules on the back.**

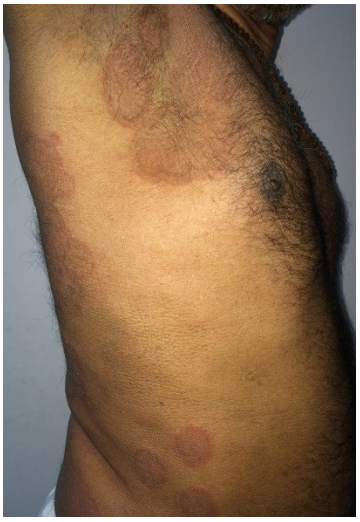

**Figure 2: Red coloured macules.**

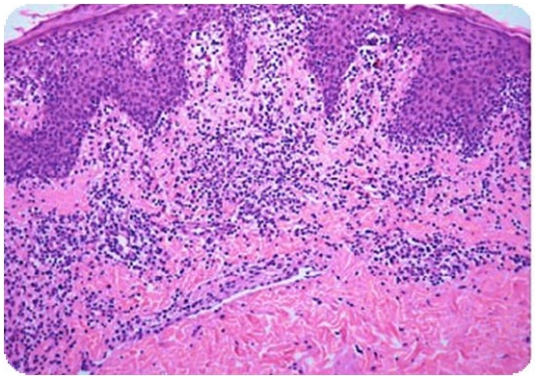

**Figure 3: Dermal Infiltrate with epidermotropism.**

## DISCUSSION

Mycosis Fungoides (MF) may be defined as a rare indolent Non-Hodgkin's lymphoma of a mature, skin-homing, clonal, malignant T lymphocytes [predominantly helper (CD4+) cells], usually observed in mid to late adulthood, that initially presents in the skin as patches, plaques, tumors or generalized erythema (erythroderma) and can involve the lymph nodes and peripheral blood and other extracutaneous sites.[4-6]

**Table 1: TNMB classification.**

| TNMB | Description |
|---|---|
| **Skin** | |
| T1 | Limited patches, papules, and/or plaques covering, <10% of the skin surface |
| T2 | Patches, papules, or plaques covering ≥10% of the skin surface |
| T3 | One or more tumors (≥1 cm diameter) |
| | Confluence of erythema covering ≥80% body surface area |
| **Node** | |
| N0 | No clinically abnormal lymph nodes |
| N1 | Clinically abnormal lymph nodes; histopathology Dutch grade 1 or NCI LN0–LN2 |
| N2 | Clinically abnormal lymph nodes; histopathology Dutch grade 2 or NCI LN3 |
| N3 | Clinically abnormal lymph nodes; histopathology Dutch grade 3 or NCI LN4 |
| NX | Clinically abnormal lymph nodes without histologic confirmation or inability to fully characterize the histologic subcategories |
| **Visceral** | |
| M0 | No visceral organ involvement |
| M1 | Visceral involvement (must have pathology confirmation and organ involved should be specified) |
| **Blood** | |
| B0 | Absence of significant blood involvement: <5% of peripheral blood Sézary cells. |
| B1 | Low blood tumor burden: > 5% of peripheral blood Sézary cells |
| B2 | High blood tumor burden: ≥1000/μL Sézary cells with positive clone |
| | One of the following can be substituted for Sézary cells: |
| | ▪ CD4/CD8 cells ≥10% |
| | ▪ CD4+/CD7− cells ≥40% |
| | ▪ CD4+/CD26− cells ≥30% |
| **Stage** | **TNMB** |
| IA | T1, N0, M0, B0-B1 |
| IB | T2, N0, M0, B0-B1 |
| IIA | T1-T2, N1-N2-NX, M0, B0-B1 |
| IIB | T3, N0-N1-N2-NX, M0, B0 |
| IIIA | T4, N0-N1-N2-NX, M0, B0 |
| IIIB | T4, N0-N1-N2-NX, M0, B1 |
| IVA1 | T1-T4, N0-N1-N2-NX, M0, B2 |
| IVA2 | T1-T4, N3, M0, B0-B2 |
| IVB | T1-T4, N1-N2-N3-NX, M1, B0-B2 |

About 75% of cutaneous lymphomas belong to the group of T-cell lymphomas; Cutaneous T-Cell Lymphomas (CTCL) account for two-thirds of cases of primary cutaneous lymphoma and, MF, which afflicts more than 50% of patients with CTCL, happens to be the most common of skin lymphomas. Median age at diagnosis is 55-60 years, but MF may occur in children and adolescents as well.[7-11] Men are more commonly affected than women. TNMB classification[12-14] of mycosis fungoides is given as above (Table 1):

Modified international society for cutaneous lymphomas/European organization for research and treatment of cancer revisions to TNMB classification of MF/Sezary syndrome.

Early lesions of MF may pose a significant diagnostic challenge to clinicians and dermato-pathologists. Many such patients need long-term follow-up and serial biopsies to make a definitive diagnosis. One of the reasons is that MF is one of the most difficult diagnoses to make in dermatopathology. Epidermotropism is one of the main feature seen in pathology. The other reason is that mycosis fungoides is a great imitator and clinical diagnosis, particularly in the early stages, is extremely difficult.

Variability in the clinical presentation and progression of MF makes multiple therapeutic options available, although its management is complex and there are no simple treatment algorithms;[15,16] skin-directed therapies that include topical corticosteroids, nitrogen mustard, carmustine, local or total body radiation therapy, topical bexarotene, and phototherapy have been shown to give good response in early stage MF.[17] However, systemic chemotherapy or targeted therapy with a monoclonal antibody, oral retinoids, recombinant interferon alpha, and fusion toxins are used in more advanced stages.[18,19]

*Funding: No funding sources*
*Conflict of interest: None declared*
*Ethical approval: Not required*

## REFERENCES

1. Rapini Ronald P, Bolognia Jean L, Jorizzo Joseph L. Mycosis fungoides. In: 1. Rapini Ronald P, Bolognia Jean L, Jorizzo Joseph L, eds. Dermatology. 2nd ed. St. Louis: Mosby; 2007: 1867.
2. Alibert JLM. Descriptions des maladies de la peau observées a l'Hôpital Saint-Louis, et exposition des meilleures méthodes suivies pour leur traitement (in French). Paris: Barrois l'ainé. 1806: 286.
3. Cerroni Lorenzo, Kevin Gatter, Helmut Kerl. An illustrated guide to skin lymphomas. Cerroni Lorenzo, Kevin Gatter, Helmut Kerl, eds. A Guide. Malden, Massachusetts: Blackwell Publishing; 2005: 13.
4. Kim EJ, Junkins-Hopkins JM, Vittorio CC, Rook AH. Mycosis Fungoides and Sezary syndrome: an update. Curr Oncol Rep. 2006;8:376-86.
5. Hess Schmid M, Dummer R, Kempf FW, Hilty N, Burg G. Mycosis fungoides with mucinosis follicularis in childhood. Dermatology. 1999;198:284-7.
6. Elmer KB, George RM. Cutaneous T-cell lymphoma presenting as benign dermatoses. Am Fam Physician. 1999;59:2809-13.
7. Neuhaus IM, Ramos-Caro FA, Hassanein AM. Hypopigmented mycosis fungoides in childhood and adolescence. Pediatr Dermatol. 2000;17:403-6.
8. Rodriguez-Vazquez M, Garcia-Arpa M, Gonzalez-Garcia J. Juvenile mycosis fungoides treated with bexarotene and PUVA. Int J Dermatol. 2007;46:99-102.
9. Tan E, Tay YK, Giam YC. Profile and outcome of childhood mycosis fungoides in Singapore. Pediatr Dermatol. 2000;17:352-6.
10. Annessi G, Paradisi M, Angelo C, Perez M, Puddu P, Girolomoni G. Annular lichenoid dermatitis of youth. J Am Acad Dermatol. 2003;49:1029-36.
11. Pabsch H, Rutten A, Van Stemm A, Meigel W, Sander CA, Schaller J. Treatment of childhood mycosis fungoides with topical PUVA. J Am Acad Dermatol. 2002;47:557-61.
12. Willemze R, Jaffe ES, Burg G, Cerroni L, Berti E, Swerdlow SH, et al. WHO-EORTC classification for cutaneous lymphomas. Blood. 2005;105(10):3768-85.
13. Olsen E, Vonderheid E, Pimpinelli N, Willemze R, Kim Y, Knobler R, et al. Revisions to the staging and classification of mycosis fungoides and Sezary syndrome: a proposal of the International Society for Cutaneous Lymphomas (ISCL) and the cutaneous task force of the European Organization for Research and Treatment of Cancer (EORTC). Blood. 2007;110(6):1713-22.
14. Olsen EA, Whittaker S, Kim YH, Duvic M, Miles Prince H, Lessin SR, et al. Clinical end points and response criteria in mycosis fungoides and Sezary syndrome: a consensus statement of the international society for cutaneous lymphomas, the United Stated Cutaneous Lymphoma Consortium, and the cutaneous lymphoma task force of the European Organisation for Research and Treatment of Cancer. J Clin Oncol. 2011;29(18):2598-607.
15. Hortwitz SM, Olsen EA, Duvic M, Porcu P, Kim YH. Review of the treatment of mycosis fungoides and Sezary syndrome: a stage-based approach. J Natl Compr Canc Netw. 2008;6(4):436-42.
16. Dummer R, Dreyling M. Primary cutaneous lymphoma: ESMO clinical recommendations for diagnosis, treatment and follow-up. Ann Oncol. 2008;19(Suppl 2):72-6.
17. Prince HM, Whittaker S, Hoppe RT. How I treat mycosis fungoides and Sézary syndrome. Blood. 2009;114(20):4337-53.

18. Knobler E. Current management strategies for cutaneous T-cell lymphoma. Clin Dermatol. 2004;22(3):197-208.
19. Lundin J, Hagberg H, Repp R, Cavallin-Ståhl E, Fredén S, Juliusson G, et al. Phase 2 study of alemtuzumab (anti-CD52 monoclonal antibody) in patients with advanced mycosis fungoides/Sezary syndrome. Blood. 2003;101(11):4267-72.

DOI: 10.5455/2349-3933.ijam20150215
**Cite this article as:** Singh T, Dhillon KS, Sharma D, Umar MS, Fatima U, Varshney KR, Dhillon A. Mycosis fungoides: cutaneous T-cell lymphoma. Int J Adv Med 2015;2:60-3.