

Mycosis Fungoides presented with De Novo Ulceration: A Case Report

Dr. Jayanti Singh¹, Dr. Vijay Paliwal², Dr. Puneet Bhargava³, Dr. Deepak Kumar Mathur⁴, Dr. C.M. Kuldeep⁵

¹MBBS, Junior Resident, Department of Dermatology, SMS Medical College & Hospital, Jaipur (Rajasthan) India

²MD, Associate Professor, Department of Dermatology, SMS Medical College & Hospital, Jaipur (Rajasthan) India

³MD, Professor, Department of Dermatology, SMS Medical College & Hospital, Jaipur (Rajasthan) India

⁴MD, Senior Professor & Unit Head, Department of Dermatology, SMS Medical College & Hospital, Jaipur (Rajasthan) India

⁵MD, Senior Professor, Department of Dermatology, SMS Medical College & Hospital, Jaipur (Rajasthan) India

Abstract—*Mycosis fungoides is the commonest cutaneous T cell lymphoma. It may be difficult to distinguish it from other dermatoses. Non responsive ulcerated plaques may be a presenting feature of mycosis fungoides. A definitive diagnosis can be made only on careful correlation of the clinical with the histopathological, immunophenotypical and molecular features. A case of mycosis fungoides who presented with de novo ulceration in plaque stage of mycosis fungoides was observed and presented which is a rare presentation.*

Key words: *Mycosis Fungoides, Ulceration, Plaque*

I. INTRODUCTION

Most common variant of primary cutaneous T cell lymphoma is mycosis fungoides (MF), generally associated with an indolent clinical course and is characterized by well-defined clinico-pathological features.¹ It typically begins as slowly progressive dermatitis like patches and plaques and when untreated evolves to nodules and eventual systemic dissemination.²

We report a case of MF who presented to us with de novo ulceration in plaque stage of MF

II. METHODOLOGY

A case report of a rare presentation of MF with multiple erythematous painful, slightly itchy, well-defined, round to oval plaques of size ranging from 6-7 mm to 10-11 cm on trunk and limbs some of them associated with ulceration since 12-13 months has come to Charak Bhawan, a Skin OPD attached to SMS Medical College, Jaipur (Rajasthan) India. This presentation itself is very rare and when it was explored it was found to be a case of MF.

III. CASE REPORT

A 68 year old male presented to us with multiple erythematous painful, slightly itchy, well-defined, round to oval plaques of size ranging from 6-7 mm to 10-11 cm on trunk and limbs some of them associated with ulceration since 12-13 months.

It seems to have started as 2-3 mm erythematous macules on the legs, gradually involved the trunk and upper limbs. Over a period of 4-5 months, they gradually increased in size up to 10-11cm, started to ulcerate in centre and developed whitish scales in periphery. Later, he had fever, loss of appetite, loss of weight.

On examination, generalized multiple scaly plaques of variable size ranging from 3×5 cm to 10×15 cm were seen. One of the plaque was covering almost entire abdomen and chest, most of them were

ulcerated and associated with exudation and crusting. The other adenexa, mucosa and systemic examinations were normal.

On laboratory investigations peripheral blood film showed few atypical cells. The bone marrow biopsy revealed mild erythroid hyperplasia. The electrolytes and renal function tests were deranged. Ultrasound of abdomen showed moderate ascites; other investigations including X ray chest, CT thorax and abdomen were unremarkable.

Skin biopsy revealed moderately dense superficial perivascular patchy lichenoid infiltrate of lymphocytes with focal spongiosis and psoriasisiform hyperplasia. The epidermis infiltrated by numerous large lymphocytes, aligned along basal layer in toy soldier pattern. Immunohistochemistry revealed strong positivity with CD3, CD4, CD45RO and was negative for CD8.

On basis of clinico-pathological findings and ISCL/EORTC revision to the staging diagnosis of MF stage 1B with ulceration was made.

Figure 1



Figure 2

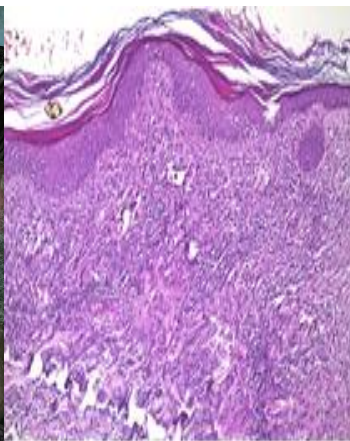
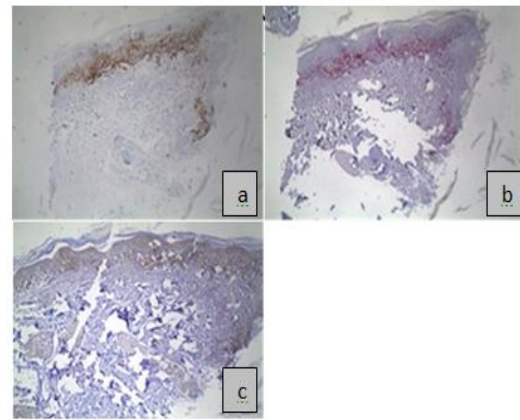


Figure 3



Legends:

- Figure 1** showing multiple erythematous scaly and ulcerated large plaques covering almost completely the chest and abdomen.
- Figure 2** showing superficial perivascular patchy lichenoid infiltrate of lymphocytes in dermis along with infiltration of epidermis by large lymphocytes aligned in toy soldier pattern.
- Figure 3**
 - showing uniformly positivity for CD3 in lichenoid lymphocytic infiltrate in upper dermis and epidermis.
 - showing CD45(RO) strongly positive in the lichenoid infiltrate in dermis in most of the lymphocytes and also in those within the epidermis
 - showing CD4 strongly positive (red colour) in more than 50% of cells in the dermal infiltrate and the epidermis.

IV. DISCUSSION

This study describes a rare case of MF. Classically, MF has 4 stages: patch, plaque, tumoral and erythroderma or sezary syndrome. The most common presentation being patch/plaque MF.^{3,4} The onset of the disease is often insidious. Its initial cutaneous symptoms may be difficult to distinguish from various other inflammatory dermatoses such as chronic eczema, psoriasis, atopic dermatitis, etc.

MF with de novo ulceration is a rare presentation. Ulceration in MF is usually seen in tumors and is attributed to rapid tumor growth and necrosis and has been associated with poor quality of life and high morbidity.^{1,5} Ulceration in this case was seen over patches and plaques. Ulceration in these stages of MF has been described in few cases in association with methotrexate toxicity and in coexistence with CD30+ cutaneous T-cell lymphoma simulating pyoderma gangrenosum in a patient with ulcerative colitis.^{6,7} The index case presented denovo with ulceration without any history of ulcerative colitis or taking methotrexate.. He did not have ulcerative colitis and never took methotrexate.

The histopathology of the early lesions of MF shows patchy lichenoid or band like lymphocytic infiltrate in the papillary dermis. Epidermotropic T lymphocytes with a clear halo around the nuclei are aligned along basal layer of epidermis. With the progression of disease the infiltrate becomes more intense, monomorphic and also more epidermotropic. Clustering of epidermotropic atypical lymphocytes occurs, also known Pautrier microabscesses. The malignant T cells seen in MF are small to medium sized lymphocytes which have nuclei with dense heterochromatin, cerebriform nuclear contours and perinuclear halo.^{1,2} However, epidermotropism may not be seen in early stages of MF and Pautrier microabscesses are also rare in early stages.⁸

On immuno-histochemistry the tumor cells are CD2+, CD3+, CD4+, CD45RO+ , usually CD7- and rarely CD8+.^{1,2}

The index case presented to us with erythematous scaly ulcerated plaques which were earlier being treated as psoriasiform dermatitis with topical steroids but were not responding to treatment. The time interval between onset of skin changes and definite pathologic diagnosis of MF is 4–6 years. In the present case this time interval was reduced to 12 months. He was diagnosed early because of atypical ulcerations and plaques.

In cases of non-responding ulcerative plaques, careful correlation of clinical and pathological findings, repeated biopsy and extended immunohistochemical analysis may be required to avoid missed and delayed diagnosis of MF.

V. CONCLUSIONS

As this type of presentation of MF cases is very rare but should not be ignored in clinical settings.

CONFLICT

None declared till date.

REFERENCES

- [1] S.J. Whittaker. Cutaneous lymphoma and lymphocytic infiltrates. In: Rook's Textbook of Dermatology (Burns T, Breathnach S, Cox N, Griffiths C, eds), 8th edn. West Sussex: Wiley Blackwell Publishing, 2010: 57.3-13.
- [2] Murphy GF, Schwarting R. Cutaneous lymphomas and leukemias. In: Lever's histopathology of the skin (Elder DE, Elenitsas R, Murphy GF, Johnson BL, Xu X, eds), 10th edn. Philadelphia: Lippincot Williams and Wilkins, 2009: 936–944.
- [3] Willemze R, Jaffe ES, Burg G, et al. WHOEORTC classification for cutaneous lymphomas. Blood 2005; 105: 3768–3785.

-
- [4] Kim YH, Liu HL, Mraz-Gernhard S, et al. Long-term outcome of 525 patients with mycosis fungoides and Sezary syndrome: clinical prognostic risk factors for disease progression. *Arch Dermatol* 2003; 139: 857–866.
- [5] Blasik LG, Newkirk RE, Dimond RL, Clendenning WE. Mycosis fungoides d'emblée: a rare presentation of cutaneous T-cell lymphoma. *Cancer* 1982; 49: 742–747.
- [6] Salameh F, Barzilai A, Baum S, et al. Mycosis fungoides and CD30+ cutaneous T-cell lymphoma simulating pyoderma gangrenosum in a patient with ulcerative colitis. *J Dermatol Case Rep* Aug 2009; 3(2): 30–33.
- [7] Breneman DL, Storer TJ, Breneman JC, et al. Methotrexate-induced cutaneous ulceration in patients with erythrodermic mycosis fungoides. *Ther Clin Risk Manag* Oct 2008; 4(5): 1135–1141.
- [8] Massone C, Kodama K, Kerl H, et al. Histopathologic features of early (patch) lesions of mycosis fungoides. A morphologic study on 745 biopsy specimens from 427 patients. *Am J Surg Pathol* 2005; 29: 550–60.