

Mycosis fungoides: The great imitator

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ABSTRACT

Introduction: Mycosis fungoides although a cutaneous T-Cell lymphoma can immitate many dermatological disorders. A 60 year old man presented to our hospital with generalized annular plaques, few with verrucous surface. The annular lesions imitated Psoriasis, Tinea, Syphilis and the diagnosis was in dilemma. Histopathology gave the light and path to diagnosis. **Case report:** A 60 year old man presented with complains of erythematous scaly lesions since 20 year, Lesions were initially flat and erythematous which later became ulcerated, crusted and painful. H/O exacerbation and remission was present. **Conclusions:** Mycosis Fungoides is a great imitator both clinically and histopathologically. We are presenting a case report of patient with Mycosis Fungoides, Stage IIA.

Key words: Cutaneous T cell lymphoma; lymphoma; NB-UVB

INTRODUCTION

In the ocean of similar dermatological manifestations it is very important to keep an eye of eagle to diagnose a case. A patient presenting as chronic plaques psoriasis, which on investigation turned out to be Mycosis Fungoides.

CASE REPORT

A 60 year old man presented with multiple erythematous scaly plaques with ulcerations all over the body since 20 years. There was history of exacerbation and remission. Patient was treated by various doctors on the lines of Psoriasis and Tinea corporis without any improvement in patient condition. No history of recurrent fever or weight loss.

Mucocutaneous examination

Multiple erythematous scaly plaques with circinate margins present over extremities and trunk with ulcerations (Figs 1A and B). Hyperpigmented patches were present over the trunk in bathing trunk pattern (Fig. 2).

Multiple verrucous plaques present over the palms with

ulceration and over dorsal aspect of feet (Figs 3A and B). Auspitz sign was negative. Lymph nodes.

Left inguinal lymph nodes enlarged approx 1 × 1 cm, soft and mobile with no dermopathic changes.

The patient's informed consent was obtained.

INVESTIGATIONS

Complete blood count, Liver function test, Renal function test-were within normal limits. Peripheral Smear - Normal blood picture, no evidence of Sezary cells. KOH MOUNT - Negative for fungal filaments; V.D.R.L - Negative; H.I.V-I AND II - Negative; CT and M.R.I was advised but was deferred as the patient was not affordable. FNAC - Negative for atypical lymphoid cells.

SKIN BIOPSY - Showed moderately dense superficial perivascular patchy lichenoid infiltrate of lymphocytes with mild spongiosis within the epidermis. The epidermis shows slight psoriasiform hyperplasia and infiltration by numerous large lymphocytes without much spongiosis. The stratum corneum shows elongated mounds of parakeratosis. Several lymphocytes are aligned along the basal layer in toy soldier pattern. The papillary dermis shows thickening of collagen bundles that appear wiry.

How to cite this article: Sajjan V, Chandela M, Pandit AM, Manjunathswamy B. Mycosis fungoides: The great imitator. Our Dermatol Online. 2015;6(1):76-79.

Submission: 07.09.2014; **Acceptance:** 29.10.2014

DOI: 10.7241/ourd.20151.18



Figure 1: (A and B) Multiple erythematous annular plaques with hyperpigmented patches and ulcerations



Figure 2: Hyperpigmented patches were present over the trunk in bathing trunk pattern

The above features are suggestive of plaque type of mycosis fungoides (Figs 4A and B).

Prior to the study, every patient gave written consent to the examination and biopsy after having been informed about the procedure.

Differential diagnosis

1. Tinea Corporis
2. Chronic Plaques Psoriasis
3. Secondary Syphilis
4. Mycosis fungoides

Treatment

Patient was started on radiation therapy (NB-UVB), with topical steroids.

Outcome and follow-up

The lesion stopped spreading and improved but the patient was lost for the follow up.

DISCUSSION

Mycosis fungoides (MF) and the Sezary syndrome are a group of extranodal non-Hodgkin's lymphomas



Figure 3: (A and B) Multiple verrucous plaques were present over the palms and dorsal aspect of feet

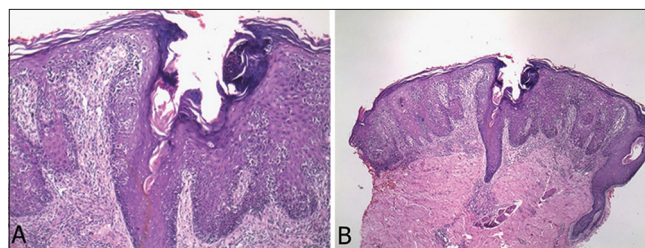


Figure 4: (A, B) Histopathological pictures showing moderately dense superficial perivascular patchy lichenoid infiltrate of lymphocytes with mild spongiosis within the epidermis. The epidermis shows slight psoriasiform hyperplasia and infiltration by numerous large lymphocytes without much spongiosis. The stratum corneum shows elongated mounds of parakeratosis. Several lymphocytes are aligned along the basal layer in toy soldier pattern. The papillary dermis shows thickening of collagen bundles that appear wavy

of T-cell origin with primary cutaneous involvement. The group differs from other primary cutaneous T-cell lymphomas (CTCL) by its unique clinical features and histopathology [1].

In its early stages, it mimics many common benign dermatoses, and hence a definitive diagnosis can be delayed. The affected T cells are of CD4+ phenotype with loss of CD7 (pan-T-cell antigen) and demonstrate T-cell receptor (TCR) rearrangement. The prognosis of patients with MF is highly dependent on the extent and type of skin involvement. The initial cutaneous presentation of MF can be patches, plaques, tumors, or erythroderma. Patients who present with limited patch/

plaque disease have good prognosis with an overall long-term survival that is similar to the expected survival of a matched control population whereas Patients with hypopigmented mycosis fungoides (HMF) present at a younger age in contrast to those with classic MF [2].

Patients presenting with limited or generalized patch/plaque type disease without peripheral lymphadenopathy can rarely exhibit extracutaneous involvement. Therefore, the staging evaluation differs for patients with MF compared to patients with other non-Hodgkin's lymphomas. Patients with tumorous or erythrodermic type of skin involvement have a less favorable prognosis, while those presenting with extracutaneous disease have a poor prognosis. Myriad of therapeutic options are available for patients with MF and the Sezary syndrome.

Selection of a specific treatment plan is based primarily on the clinical stage of the disease.

Variants and subtypes

Folliculotropic mycosis fungoides - occurs in adults and men are most commonly affected. Presents as grouped erythematous follicular papules, acneiform lesions with severe pruritis, indurated plaques and rarely it is associated with non-scarring alopecia. Characteristic lesions are hairless infiltrated plaques.

Pagetoid Reticulosis-Localized types is characterized by presence of a single asymptomatic, psoriasiform, hyperkeratotic or circinate plaques on the extremities. Granulomatous Slack. Skin-occurs mostly in adult men. The flexure shows area of pendulous lax skin associated with Hogkins Lymphoma has been reported [3].

Staging

Various types of staging systems are done for Mycosis Fungoides. Two important once are TNM staging (Table 1) and Bunn and lambert system (Table 2) which involves clinical findings like body surface area involved, lymph node status and erythroderma. Staging of Mycosis Fungoides makes it easier to prognosticate and to make a treatment plan.

Differential diagnosis

The differential diagnosis with Tinea corporis was ruled out on the basis of KOH mount as it was negative. Secondarily syphilis was ruled out by V.D.R.L negativity. There was no histopathological correlation with Chronic plaque

psoriasis and there is no evidence that early aggressive systemic therapy is preferable to conservative therapy in the management of limited disease. Anticonvulsants such as carbamazepine, phenytoin, phenobarbital and valproic acid are the major offenders [4]. Out of these Carbamazepine induced pseudo-MF is the common one [5,6]. Drugs like Gleevec, protein tyrosine kinase inhibitor used in the treatment of chronic myelogenous leukemia, melanoma and gastrointestinal stromal tumor are also implicated [7].

Treatment

Topical therapy is the main stay of treatment for patch/plaque type without extracutaneous involvement, whereas chemotherapy and other aggressive systemic regimens are reserved for recalcitrant disease or extracutaneous involvement [8]. Various treatment modalities (Table 3) are available for treatment of

Table 1: Stages of Mycosis Fungoides. TNMB Classification of Mycosis Fungoides and Sezary Syndrome

TNMB Classification of Mycosis Fungoides and Sezary Syndrome	
T (skin)	
T1	Limited patch/plaque (involving<10% of total skin surface)
T2	Generalized patch/plaque (involving>10% of total skin surface)
T3	Tumor(s)
T4	Erythroderma
N (lymph node)	
N0	No enlarged lymph nodes
N1	Enlarged lymph nodes, histologically uninvolved
N2	Enlarged lymph nodes, histologically involved (nodal architecture unaffected)
N3	Enlarged lymph nodes, histologically involved (nodal architecture effaced)
M (viscera)	
M0	No visceral involvement
M1	Visceral involvement
B (blood)	
B0	No circulating atypical (Sezary) cells or <5% of lymphocytes
B1	Low blood tumor burden (>5% of lymphocytes are Sezary cells, but not B2)
B2	High blood tumor burden (1000/mL Sezary cells+positive clone)

Table 2: Stages of Mycosis Fungoides. Bunn and Lambert system

Bunn and lambert system	Clinical findings	Tumor-node-metastasis stage
IA	<10% BSA patch/plaque	T1 N0
IB	>10% BSA patch/plaque	T2 N0
IIA	Patch or plaque with palpable nodes without histological involvement	T1/2 N1
IIB	Cutaneous tumours with or without palpable nodes	T3 N0/1
III	Erythroderma with or without palpable nodes	T4 N0/1
IVA	Non-palpable or palpable nodes with histological involvement	T-any N2/3
IVB	Visceral involvement	T-any N-any M1

Table 3: Treatment of mycosis fungoides/Sezary syndrome

Stage	First line	Second line	Experimental	Not suitable
IA	Skin-directed therapy or no therapy	Skin-directed therapy or no therapy	Bexarotene gel	Chemotherapy
IB	Skin-directed therapy	α -Interferon+PUVA, Total skin electron beam	Denileukin Diftitox, Bexarotene	Chemotherapy
IIA	Skin-directed therapy	α -Interferon+PUVA, Total skin electron beam	Denileukin Diftitox, Bexarotene	Chemotherapy
IIB	Radiotherapy/total skin electron beam, chemotherapy	α -Interferon, Denileukin Diftitox, Bexarotene	Autologous PBSCT, mini-allograft	Cyclosporin
III	PUVA + α interferon, Extracorporeal photopheresis + α Interferon, methotrexate	Total skin electron beam, Bexarotene, Denileukin, Diftitox, chemotherapy, Alemtuzumab	Autologous PBSCT, mini-allograft	Cyclosporin
IV A	Radiotherapy/Total skin electron beam, chemotherapy	α -Interferon, Denileukin Diftitox, Alemtuzumab, Bexarotene	Autologous PBSCT, mini-allograft	Cyclosporin
IV B	Radiotherapy, chemotherapy	Palliative therapy	Mini-allograft	-

Abbreviations: PBSCT - Peripheral blood stem cell transplant; PUVA - Psoralen ultraviolet A; Skin-directed therapy including topical Emollients, Steroids, Mechlorethamine, Carmustine, Bexarotene gel, UVB/PUVA, Superficial Radiotherapy. Stage III includes Sezary syndrome, although some cases are seen in stage IVA. ECP is ideal for those patients with peripheral blood involvement. (From the joint BAD and UKCLG guidelines for Primary CTCL)

Mycosis Fungoides. Our patient was treated with topical steroids and Radiation therapy in the form of NB-UVB which led to improvement of lesions.

CONSENT

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. The examination of patients is conducted according to the Declaration of Helsinki principles.

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Source of Support: Nil, **Conflict of Interest:** None declared.