Case Report

Mycosis fungoides: cutaneous T-cell lymphoma

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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/mm3. Skin biopsy revealed dermal dense infiltrates of lymphoid tissue with focal epidermotropism (Figure 3). He was diagnosed stage 1B Mycosis fungoides (T2 N0 M0 B0) according to TNMB classification. Patient was started on NBUVB therapy and Interferon-α.
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.

<table>
<thead>
<tr>
<th>Stage</th>
<th>TNMB</th>
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<tbody>
<tr>
<td>IA</td>
<td>T1, N0, M0, B0-B1</td>
</tr>
<tr>
<td>IB</td>
<td>T2, N0, M0, B0-B1</td>
</tr>
<tr>
<td>IIA</td>
<td>T1-T2, N1-N2-NX, M0, B0-B1</td>
</tr>
<tr>
<td>IIB</td>
<td>T3, N0-N1-N2-NX, M0, B0</td>
</tr>
<tr>
<td>IIA</td>
<td>T4, N0-N1-N2-NX, M0, B0</td>
</tr>
<tr>
<td>IIB</td>
<td>T4, N0-N1-N2-NX, M0, B1</td>
</tr>
<tr>
<td>IVA1</td>
<td>T1-T4, N0-N1-N2-NX, M0, B2</td>
</tr>
<tr>
<td>IVA2</td>
<td>T1-T4, N3, M0, B0-B2</td>
</tr>
<tr>
<td>IVB</td>
<td>T1-T4, N1-N2-N3-NX, M1, B0-B2</td>
</tr>
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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. Men are more commonly affected than women. TNMB classification 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/Sézary 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; 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. However, systemic chemotherapy or targeted therapy with a monoclonal antibody, oral retinoids, recombinant interferon alpha, and fusion toxins are used in more advanced stages.

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REFERENCES


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