Mycosis fungoides: A Chameleon of Dermatology

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ABSTRACT

Mycosis fungoides (MF) is the most common type of cutaneous T-cell lymphoma, characterized by skin-homing of clonal, mature malignant T-lymphocytes. MF develops slowly over several years and may have a variety of clinical presentations. In addition to the classical clinical lesions-itchy patches, plaques or tumors, it may also present with atypical hypopigmented lesions, dermatophytic lesions or psoriasiform lesions that may be confused with common benign conditions such as eczema and psoriasis.

Clinical picture may pose a significant challenge and a diagnostic dilemma to the dermatologist. Longterm follow up and serial biopsies help to establish definitive diagnosis.

Keywords : Mycosis fungoides (MF), Cutaneous lymphoma, Psoriasis, Vitiligo.

INTRODUCTION

Mycosis fungoides (MF) is the commonest variant of primary cutaneous T cell lymphoma, accounting for almost 50% of all primary cutaneous lymphomas.[1, 2] It most commonly affects middle-aged and elderly adults of all races[3]. Typically, neoplastic T cells localize to the skin and produce patches, plaques, tumors or erythroderma[4]. It is characterized by a relatively consistent constellation of clinical, histologic, immunophenotypic and molecular aberrations[5].

MF typically manifests as an indolent cutaneous eruption with erythematous scaly patches or plaques and may progress to generalized erythroderma, cutaneous tumors or extracutaneous involvement. The initial skin lesions are often confined to sun protected areas. The rate of progression is unpredictable. Tumors, however, develop only in a minority of patients from patches, plaques, or de novo. They more commonly arise on the face and body folds. Leonine facies results from malignant T cell infiltration leading to extensive thickening and skin fold accentuation. The palms and soles can develop hyperkeratosis and skin fissuring. These phases may be distinct or overlapping at diagnosis[6,7].

Several atypical forms resembling other dermatoses like vitiligo, dermatophytosis and psoriasis may pose a diagnostic challenge to the dermatologist. Clinical diagnosis, follow up and serial biopsies may unravel the mystery and establish the diagnosis of this great mimicker. We have encountered similar situation in our study of 3 cases of MF.

CASE REPORT

Case 1 :

A young adult male aged 28 years old presenting with itchy, erythematous, scaly patches over the trunk (Figure 1a, 1b) visited several dermatologists and a diagnosis of dermatophytosis was made. Topical and systemic anti-fungals were prescribed repeatedly with no response. In View of chronicity and inadequate response to the above treatment patient was subjected to thorough clinical examination which revealed erythematous patches and plaques with mild scaling. No induration was seen. Few patches were atrophic. No lymphadenopathy was seen. Other systemic examination was within normal limits . KOH examination of scales from the lesions did not show any fungal hyphae. All biochemical investigations were within normal limits. Histopathological examination revealed mild spongiosis in epidermis and perivascular lymphocytic infiltrate. Few lymphocytes showed nuclear irregularity with halo around them.

Case 2 :

A 52yr old male referred to the DVL department with six months history of scaly plaques with superficial erosions and crusted nodulo-ulcerative lesions all over the body. With these symptoms he was misdiagnosed as plaque type psoriasis and advised various mild and high potency topical corticosteroids which did not offer any relief. Thorough skin examination revealed reddish brown ulcerated lesions and indurated erythematous plaques with superficial erosions on trunk and extremities (Figure 2a, 2b). His nails were intact without pitting or dystrophy. He had cervical and inguinal lymphadenopathy. Auscultation of chest did not reveal any abnormality. No organomegaly was seen. VDRL and HIV serology were negative. Lesional biopsy revealed: atypical lymphocytes with cerebriform nuclei. Pautrier's abscess was present in the epidermis (Figure 3).

Case 3 :

A middle aged male came to DVL department with multiple asymptomatic hypopigmented patches over the trunk and proximal extremities. These hypopigmented patches first appeared on the trunk and later on proximal extremities too. The initial hypopigmented lesions gradually turned into erythematous patches and increased in size as well. There was no history of systemic symptoms such as fever, lassitude or weight loss.

On examination there were asymptomatic, hypopigmented and few vitiligo-like depigmented irregular atrophic macules and patches on the trunk and extremities. These patches showed a finely wrinkled slightly scaly surface (Figure 4). They were not indurated and there was no telangiectasia. No lymphadenopathy or organomegaly seen. All other investigations were within normal limits.

Histological examination of a vitiligo like lesion showed an infiltrate of mononuclear cells at the interface with prominent epidermotrophism. No atypical cells were seen.

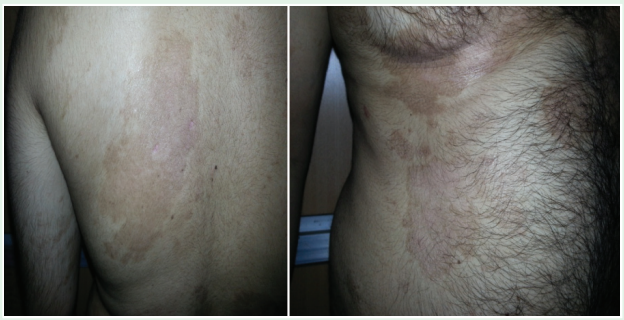

Figure 1: Photographs showing erythematous, scaly patches over the trunk.

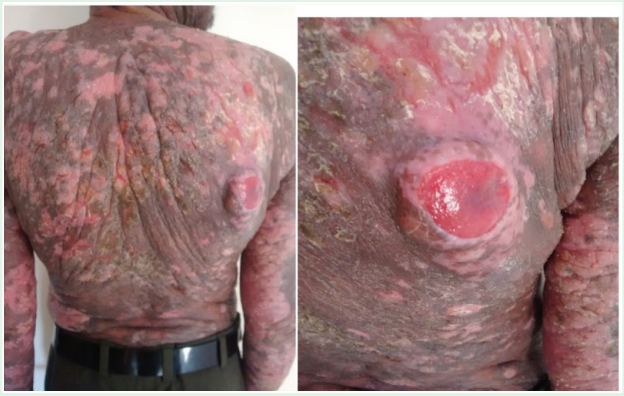

Figure 2: Photographs showing reddish brown ulcerated lesions and indurated erythematous plaques with superficial erosions on trunk and extremities.

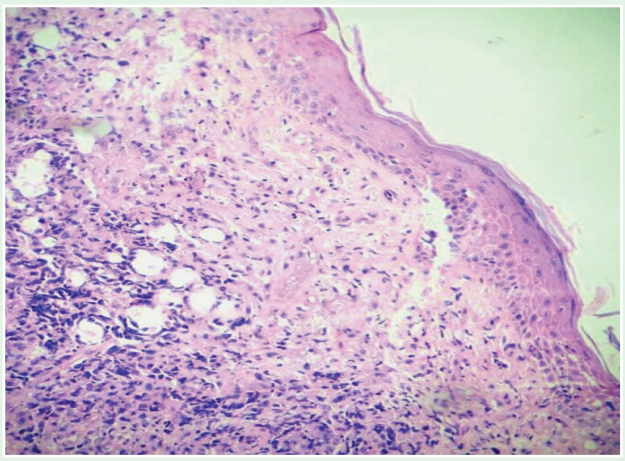

Figure 3: Section showing atypical lymphocytes with cerebriform nuclei. Pautrier's abscess was present in the epidermis.

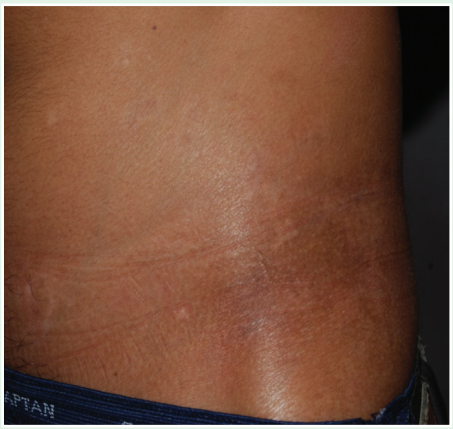

Figure 4: Photograph showing hypopigmented and few vitiligo-like depigmented irregular atrophic macules and patches on the trunk and extremities.

DISCUSSION

Mycosis fungoides is the most common type of cutaneous T-Cell lymphoma accounting for about fifty percent of all cases. It is an epidermotropic cutaneous T-Cell lymphoma caused by proliferation of small to medium sized lymphocytes with cerebriform nuclei. Clinically MF is divided into four main sub types:

Patch stage, plaque stage, tumor stage & erythroderma. As the disease progresses, patches may evolve over a variable period of time, into infiltrated plaques with a more generalized distribution. Plaques may be followed by ulcerated and exophytic tumors. Tumors develop only in a minority of patients, although it is common to have patch, plaque and tumor lesions simultaneously on different parts of the body.

Some patients with patch stage MF never progress to other forms of the disease. Tumors are the presenting sign in about 10% of cases. Lymphadenopathy is usually a late occurrence. Visceral dissemination (lungs, spleen, liver, gastrointestinal tract) may develop subsequently. Extracutaneous dissemination is directly correlated to the extent of cutaneous disease[7].

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. It mimics several dermatoses. In addition to the classical presentation, several atypical forms hypopigmented lesions, psoriasiform lesions, palmo plantar form and dermotophyte like lesions have been reported. Early lesions of MF may pose a significant diagnostic challenge to clinicians and dermoto-pathologists. Many such patients need long term follow up and serial biopsies to make definitive diagnosis.

The descriptive term "Mycosis fungoides" chosen in 1806 by Alibert, suggests that first differential diagnosis of tinea corporis for a typical MF lesion.Here the patient presented with scaly annular plaques all over the body. He was diagnosed to be a case of dermatophytosis and was given several antifungals but without any clinical relief. Then a biopsy was done and it revealed atrophy of the epidermis; a superficial, perivascular, and interstitial lymphocytic infiltrate with numerous atypical lymphocytes; and exocytosis of atypical lymphocytes into the epidermis with formation of microabscesses-findings consistent with the diagnosis of mycosis fungoides. Treatment with Nb UVB led to long-term remission of the mycosis fungoides.

Recalcitrant tinea corporis has been reported as the presenting manifestation of patch-stage MF in several studies. This may be due to the immunosupression by the underlying condition.

In early stages of MF the characteristic lesion consists of erythematous macules or papules. Often some degree of scaling is observed, similar to psoriasis. The edges of the lesions may exhibit increased scaling, corresponding to a growing infiltrate but the histopathological picture differs from that of psoriasis.

The term hypopigmented MF was mentioned for the first time by Ryan et al in 1973[8]. This variant of MF is more commonly seen in children and dark skinned individuals; especially Asians and occurs at a much earlier age as compared to the classical MF. Hypopigmented MF lesions were considered to be the earliest and benign lesions and often clinically mistaken for other hypopigmented lesions like leprosy, vitiligo etc.

Histopathology and immunohistochemistry are helpful to make accurate diagnosis. The histopathological lesions shows scattered lymphocytes in basal epidermis with or without Pautrier's microabscesses. The main purpose of presenting these case reports is to show the clinical similarity of MF with other dermatoses and to ensure a proper diagnosis for better treatment. Thus the clinician must be vigilant and be prepared to take a holistic view of the clinical, histological, immunphenotypic and molecular genetic evidence to make a proper diagnosis, accurate staging and classification.

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Please cite this article as: Swetha D,Pratap D V S. Mycosis fungoides: A Chameleon of Dermatology.Perspectives in medical research 2015;3:2:18-20.

Sources of Support: Nil,Conflict of interest:None declared