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Research Article

MYCOSIS FUNGOIDES MIMICKING CHRONIC ACTINIC DERMATOSES-A DIAGNOSTIC CHALLENGE !!!

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ABSTRACT

Mycosis fungoides (MF) or cutaneous T cell lymphoma(CTCL) is a neoplasm with an indolent course having varied clinical and histologic manifestations. Various entities like Spongiotic dermatitis, allergic contact dermatitis, seborrhoeic dermatitis and other benign conditions like eczema and psoriasis may histologically and clinically mimick with mycosis fungoides and represent a potential trap for unsuspecting pathologists. An early diagnosis with vigilant follow-up and multiple biopsies can halt the further progression of the disease. Patients with only cutaneous involvement has a better prognosis compared to those with systemic involvement.

INTRODUCTION

Mycosis fungoides, an epidermotropic primary cutaneous T cell lymphoma affecting the T-helper (CD4+) cells. Its Incidence is about 0.4/100000. It is the most common type accounts for 50% of primary cutaneous lymphoma.[1] Clinical presentation of cutaneous lymphoma is varied and deceptive in its early stage as it may mimic many benign processes, such as eczema, psoriasis and contact dermatitis. Early in the course of the disease, the clinical and histologic diagnosis of mycosis fungoides is difficult and pose a diagnostic challenge. Here we present a case of Mycosis fungoides who was earlier misdiagnosed as chronic actinic dermatitis.

Case Report

A 62 yr male with c/o itching all over the body since 4 yrs was clinically diagnosed as a case of Chronic Actinic Dermatitis and treated for the same with remission and relapse. On examination there was marked thickening and lichenification of skin all over the face, ear with few excoriations and also thickened plaque like growths over thigh and trunk. [Fig 1 a,b,c,d] Few old lesions showed hypopigmentation. There was mild pallor, with no lymphadenopathy and hepatosplenomegaly. Peripheral blood smear was normal with no atypical cells. Chest X-ray was normal & USG of abdomen and pelvis showed mild hepatomegaly. LFT was normal. Punch biopsy of skin showed irregular acanthosis, elongation of rete pegs, Fig [2 a,b,c]dense diffuse infiltration of small to medium sized atypical lymphocytes in upper dermis and around dermal appendages showing features of epidermotropism. IHC came out to be positive for CD3 only[Fig 3a,b,c] CD 20 being negative Thorough screening of thorax and abdomen by CT scan didn't reveal an deep seated lymphadenopathy or any mass lesion. Bone marrow aspiration was normal with no infiltration of tumour cells to rule out sezary syndrome. With this histomorphology a diagnosis of Primary cutaneous T cell lymphoma -(Mycosis Fungoides-follicular variant) was given.

DISCUSSION

Mycosis fungoides (MF)is an indolent tumor limited to the skin with a widespread distribution. The malignancy begins in the skin but may progress over time to involve the entire lymphoreticular system, including lymph nodes and internal organs. Most cases of CTCL are diagnosed in patients 50 to 60 years of age. It is twice as common in black persons as in white persons, but it may affect persons in any age or ethnic group [3].

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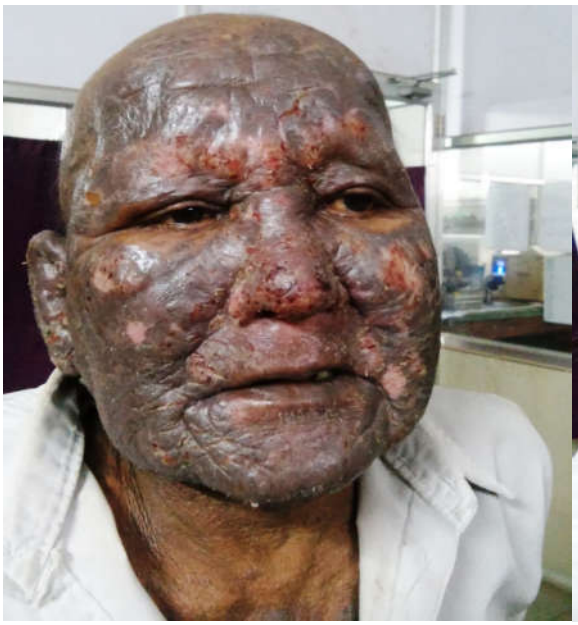
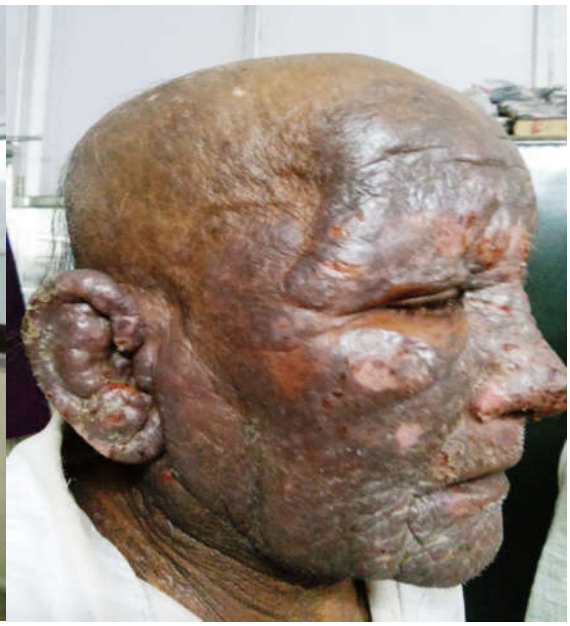


Fig 1(a)



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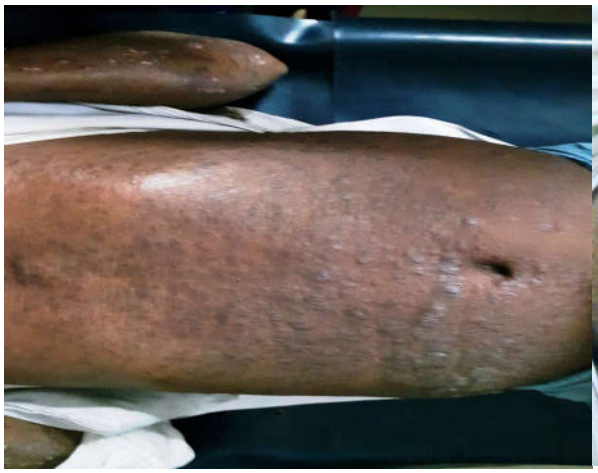

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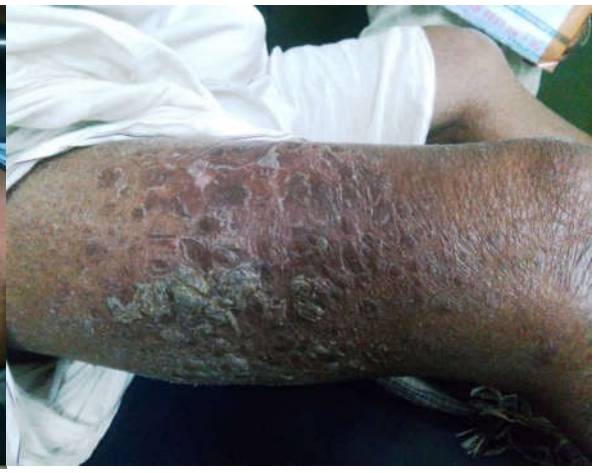


Fig 1(d)

Fig 1 a,b Thickening and lichenification of skin all over the face, ear with few excoriations.Fig 1(c,d)- Thickened plaque like growths over thigh and trunk.

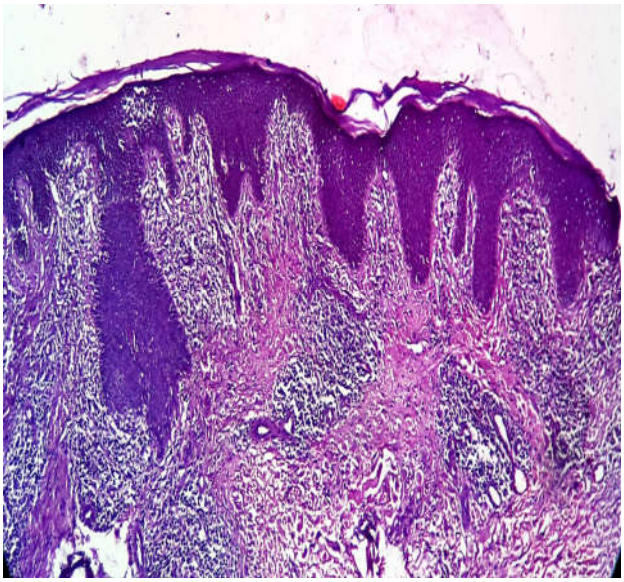


Fig 2a Histopathology (4x,scanner)- skin showed irregular acanthosis, elongation of rete pegs

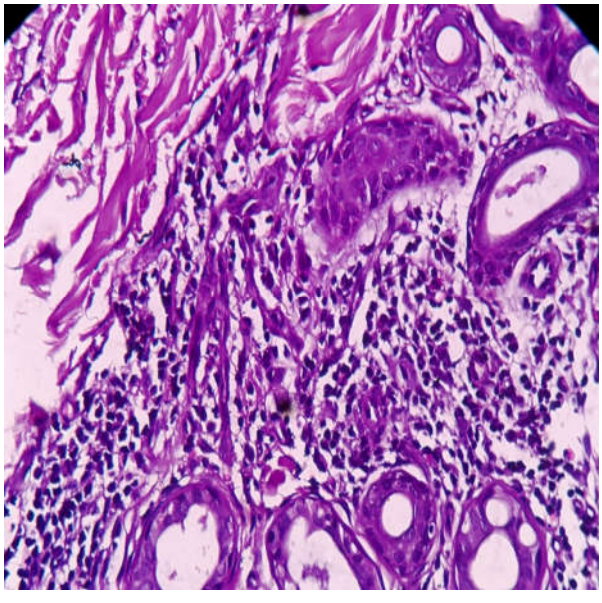


Fig 2 b

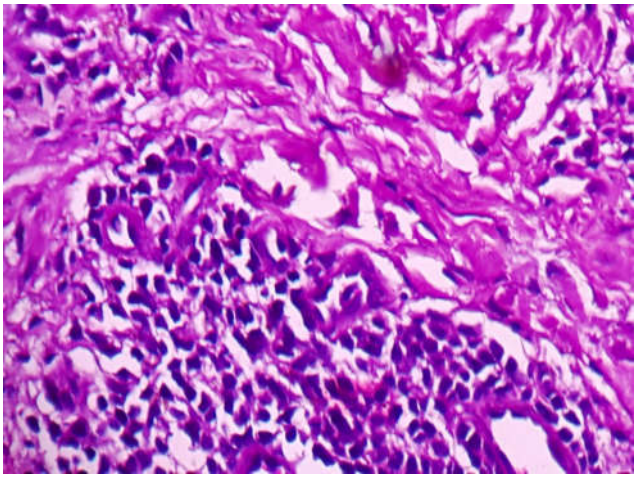
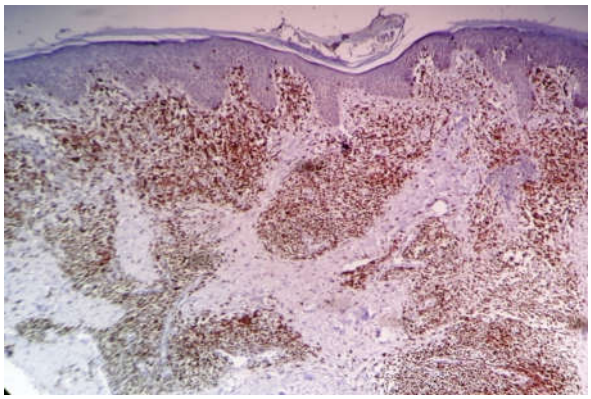
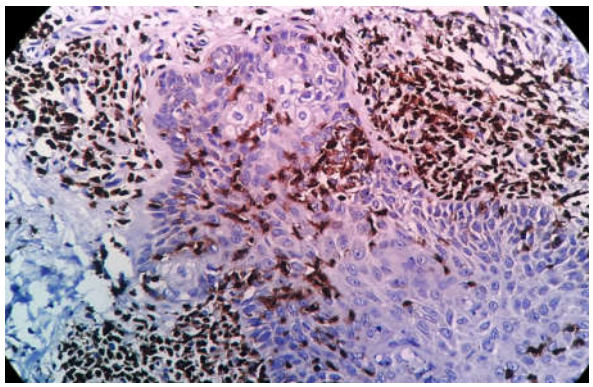


Fig 2 b,2c Histopathology (40x)HP View -Showing atypical lymphocytes in mid dermis around dermal appendages.





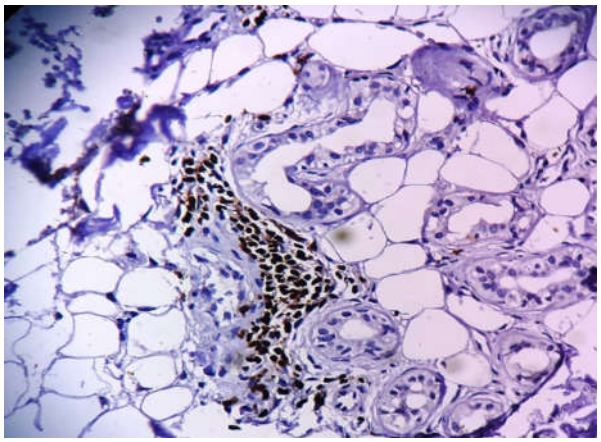


Fig 3 a b c IHC study showing atypical lymphocytes positive for CD 3 positive.

A recent series–[4] noted that between 4 and 5 percent of patients with CTCL had the disease by 20 years of age. Male: female ratio is 2:1.[1]

Patients may give a history of long standing pruritic, inflammatory patches or plaques present for more than a decade. To further complicate matters, the use of topical steroids in the early stages, may diminish the lesion in intensity supporting an incorrect diagnosis of a benign process. Although the patient and physician may be reassured by this seemingly well-controlled dermatitis, the correct diagnosis is delayed by topical corticosteroid treatment.[2]

Beginning with a premycotic erythematous or eczematous stage, cases of cutaneous lymphoma slowly progress to an infiltrative 'plaque' stage and eventuate in the 'tumour' state.[5] Variants like verrucous, bullous, pustular, purpuric, hyperkeratotic, hypopigmented, erythrodermic and poikilodermic lesions have also been described.[6]

Diagnosis of Mycosis fungoides histopathologically is based on the finding of dense diffuse infiltration of small to medium size atypical lymphocytes with cerebriform nuclei in upper dermis often as haloed cells either singly or in a linear distribution and around dermal appendages with features of epidermotropism .[7]. IHC of CD3 was positive for tumor cells with CD 2O being negative which clinched the diagnosis of “cutaneous T cell lymphoma-Mycosis fungoides”. Other panel of IHC markers which are usually positive are CD2,CD5,CD4.[8].

The clinical presentation of MF brings chronic actinic dermatitis and sezary syndrome as differential diagnosis which can be ruled out by distinctive histomorphology .Chronic actinic dermatitis presents with polymorphous perivascular infiltrate and absence of clonal lymphocytes with few to no mitosis. Sezary syndrome on other hand is the systemic manifestation of mycosis fungoides with presence of sezary cells in peripheral blood and bone marrow which was were absent in this case.

Single most prognostic factor in mycosis fungoides is the extent of cutaneous and extracutaneous disease as reflected in clinical stage, once the disease becomes systemic, the prognosis is significantly worse[9]. Age over 60 years and elevated LDH are other adverse prognostic parameters [10]. The treatment of CTCL is difficult as the disease shows a slow evolution with wide progression in many patients. Patients with limited skin involvement topical therapies are desirable. Other chemotherapy agents that are effective in treating CTCL are group of alkyiating agents mechlorethamine hydrochloride or nitrogen mustard. Treatment with psoralen plus ultraviolet A(PUVA) appears to be promising in early MF with skin lesions only.[11] Patients with more extensive disease require systemic therapy to prolong survival. Oppurtunistic infection is the usual cause of mortality in patients who die of this disease.

CONCLUSION

The diagnosis of MF remains challenging and a definitive diagnosis of this uncommon tumor depends on distinctive histomorphology and IHC. A diagnosis of primary cutaneous lymphoma should be considered if a chronic psoriasiform or eczematous dermatitis has not responded appropriately to treatment. Differentiation of true MF from dermatologic conditions mimicking MF is important to ensure proper

management of the patient's symptoms and treatment, and to provide accurate prognostic information. The diagnosis may be missed and the patient left untreated for years because of the benign appearance of this disorder. Maintaining an index of suspicion for the disease, with multiple skin biopsies over time and vigilant patient follow-up are essential to effectively treat this disorder in its early stages and prevent progression to a life-threatening malignancy.

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