Atypical Presentation of Mycosis Fungoides- A Rare Case Report

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Abstract

Commonest cutaneous T cell lymphoma is mycosis fungoides. Clinical stages are patch, plaque, tumor ;erythroderma and poikoiloderma . Extracutaneous spread takes place in late stages where any organ may be involved. The majority of reported cases are with typical presentation. A rare case of mycosis fungoides in a 45-year-old male with papular eruption is described.

Key words: *Mycosis Fungoides Atypical Presentation*

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Introduction

It's a cutaneous T cell lymphoma which typically begins as slowly progressive dermatitis like patches and plaques, when untreated evolves into nodules and leads to eventual systemic dissemination[2]. The patch/plaque stage of the disease is the result of medium sized malignant T cells while in the more advanced stage develops as a consequence of exclusively dermal involvement of non-epidermotropic malignant T cells[2, 3]. The characteristic cell of mycosis fungoides is a small or medium sized lymphocyte with a cerebroid nucleus. Mycosis fungoides is a disease predominantely of adult males, although children are occasionally affected[2, 4]. It is an indolent type of cutaneous T cell lymphoma that evolves slowly through skin limited disease to systemic involvement [4]. The disease can become biologically high grade in its later stages.

Case report

A 45 year old male was brought to skin department of a tertiary care hospital with history of papular rashes all over the body, fever and pruritus for the past 8 months. Bilateral symmetrical erythematous grouped fine, dry, non adherent scaly papules distributed on nose, face, back of neck, behind the ears, extensor surface of arms and forearms of both upper extremities, dorsal aspect of both hands, extensor surface of both knee joints. Papules are erythematous, round shaped, 3-6 mm in diameter, discrete and firm.

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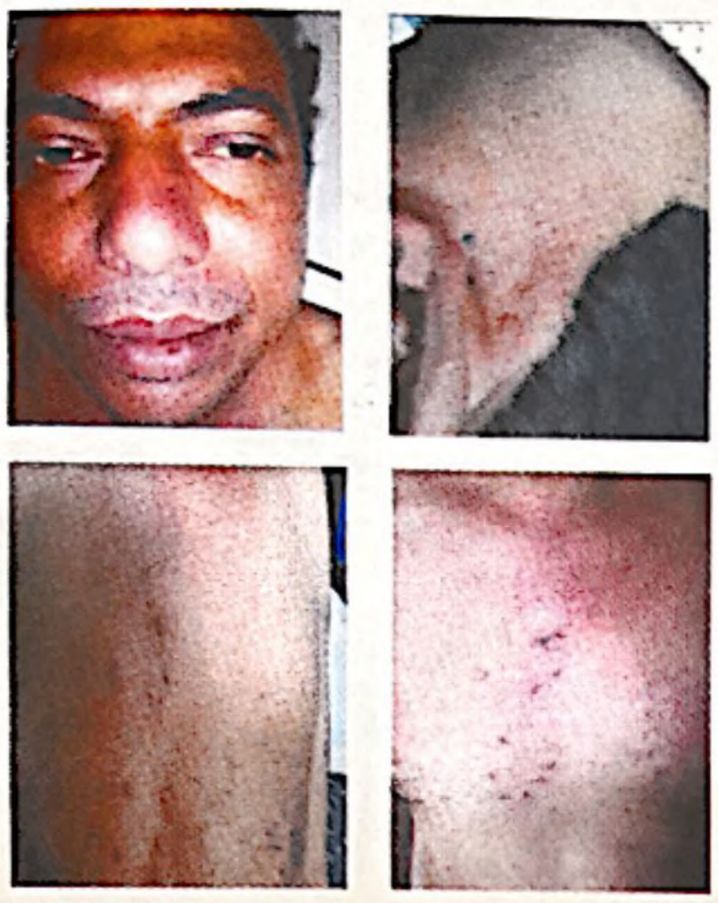

Fig: Multiple erythematous papules on face, postauricular region, chest

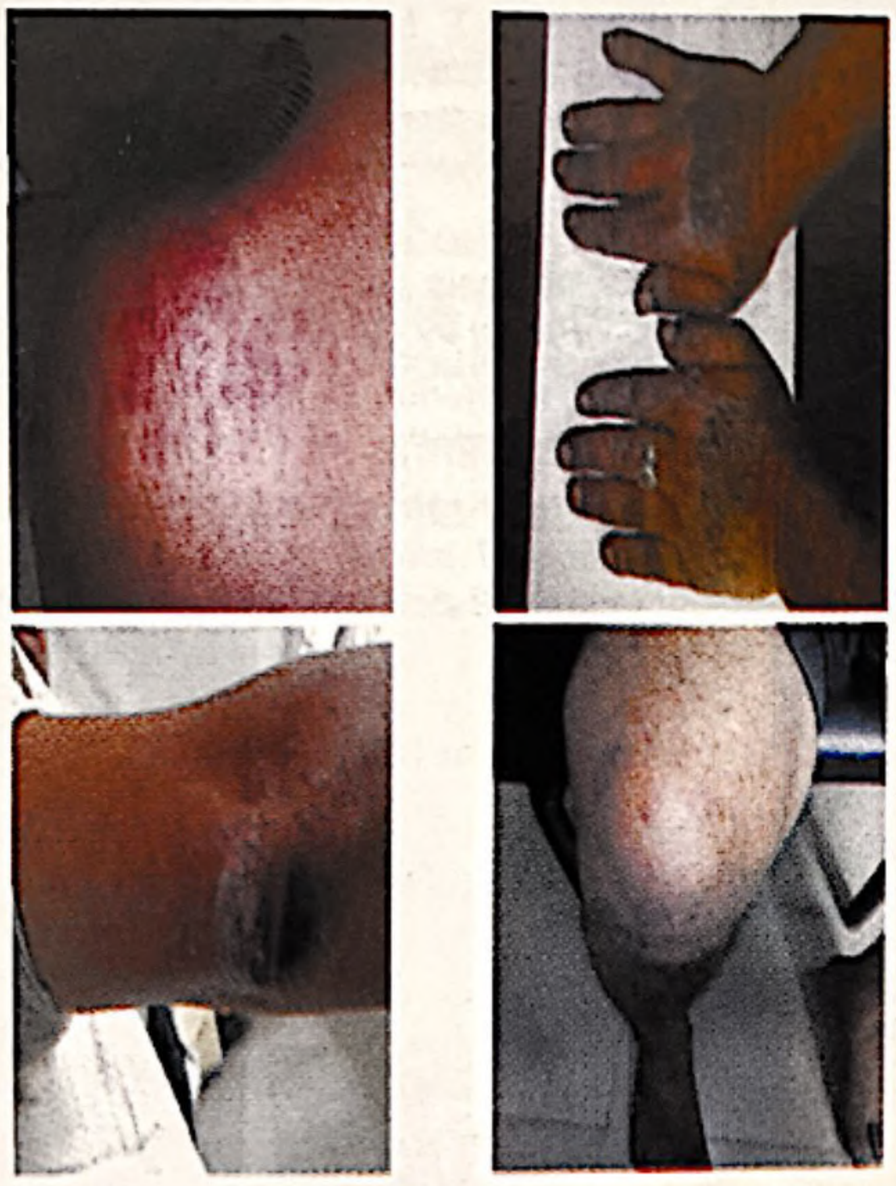

Fig: Multiple erythematous papules on trunk, dorsal hand, knees

On laboratory investigations, hemoglobin was 8.0 g%, total leukocyte count was 8500/cumm and platelet count was 240,000/cumm. Peripheral smear showed no atypical cells. There was no involvement of the bone marrow. Specimens were from right shoulder and dorsal hand.

Histopathological findings:
Sections of skin reveal mild hyperketatosis with acanthosis and exocytosis by lymphocytes.

The superficial dermis reveals moderate infiltrate of lymphocytes mixed with histiocytes
Comments: Suggestive of mycosis fungoides

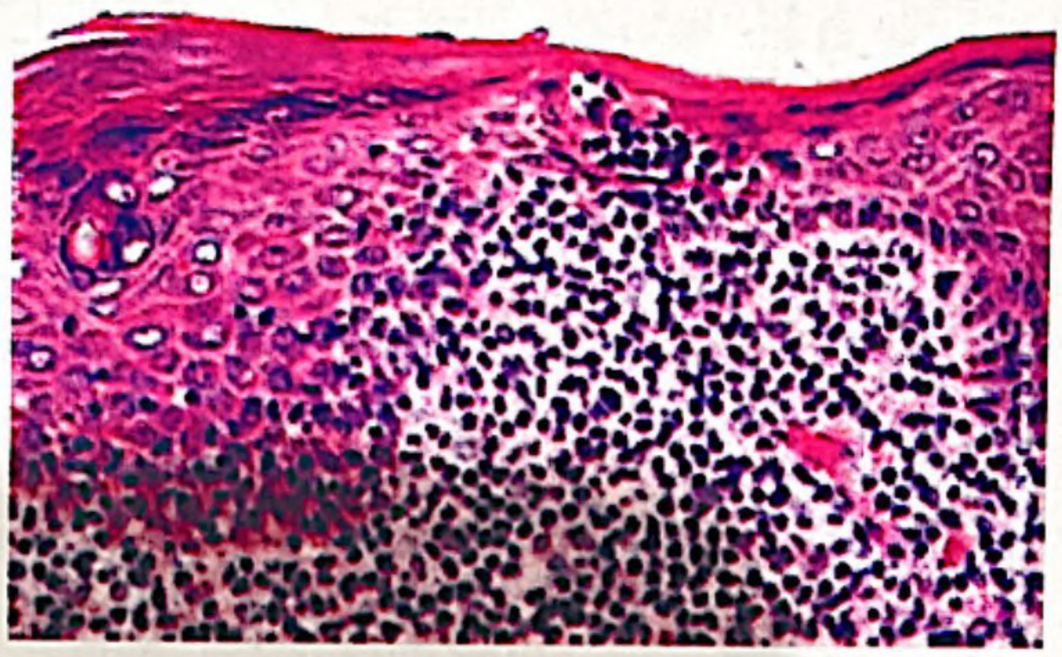

Fig: Histopathological findings of the patient

Immunohistochemistry showed...

- LCA (CD45): Positive
- CD_3 : Positive, CD_3 positive T Lyphocytes are also present in the epidermis.
- CD_5 : Positive, CD5positive T Lyphocytes are also present in the epidermis.
- CD_{20} : Most of the Lymphocytes in the dermis are negative.
- Comment: Mycosis fungoides

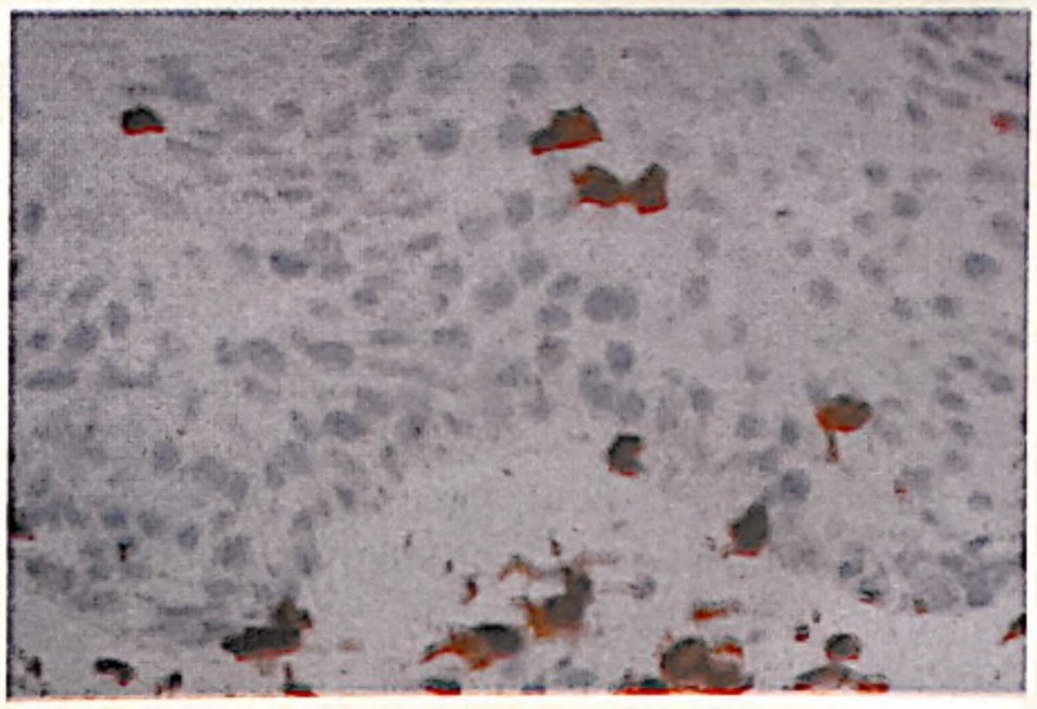

Fig: Immunohistochemistry of the patient

These lymphoid cells were atypical and large with convoluted nuclei. Histopathological diagnosis of mycosis fungoides-tumor stage was made. CD3, CD4 and LCA positivity (Fig. 3).

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Discussion

Mycosis fungoides is the T cell lymphoproliferative disorder that arises primarily in the skin and that may evolve into generalized lymphoma[3, 5]. A viral etiology has been suspected because of certain similarities to HTLV-1 associated adult T cell leukemia. Lymphomabut has not been proved as yet[6]. The majority of the cases occur in adult males but adolescents can also be affected[7]. It has various manners of presentation and progression.Traditionally, mycosis fungoides is divided into three stages-premycotic, mycotic and tumor stage[2, 7]. In the premycotic stage, the skin is erythematous, scaly and pruritic. The microscopic appearance may be non-diagnostic small number of frankly atypical lymphoid cells. These cells may evade the dermis to form Pautrier's microabscess. The characteristic cell of mycosis fungoides is a small or medium sized lymphocyte with a cerebroid nucleus. This term cerebroid nucleus refers to the highly irregular contour of the thick nuclear membrane, which results in an appearance somewhat reminiscent of brain convolutions[2]. In our case, the patient presented with tumor stage of the disease. He had lymphadenopathy and hepato-splenomegaly indicating an extracutaneous spread. However there were no atypical cells in the peripheral blood, ruling out the possibility of Sezary Syndrome. In mycosis fungoides in cases of extracutaneous spread, lymph node, liver, spleen and lungs are often involved, in addition to the peripheral blood[2, 8]. Transformation to large highly atypical lymphocyte with development of an aggressive biological course [4]. Although mycosis fungoides is a malignant lymphoma of low grade malignancy with prolonged survival, important prognostic parameters are at diagnosis, needed absence cutaneous spread and there should be as prognostic parameters have a frequent terminal complication[2,9].

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