

## Mycosis fungoides- adnexotropic variant -a rare case report

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### Abstract

Mycosis fungoides accounts for <1% of non hodgkins lymphomas. It is the commonest cutaneous T cell lymphoma comprising 44% of cutaneous lymphomas. It is clinically characterised by patch, plaque and tumor nodules. Herein we present a case of a female patient of 56 years who presented with multiple papular and nodular lesions of extremities, chest wall and ulcerated nodular lesion of scalp since 6 months.

**Key words:** Adnexotropic, Cutaneous, Lymphoma, Mycosis Fungoides

### Introduction

Mycosis fungoides was first described by Alibert in 1806 as a mushroom like skin neoplastic condition [1]. There is predominance in patients who are between 55-60 years of age being more common in male patients [2]. Initial lesions are similar to inflammatory dermatoses, with the development of disease, lesions become infiltrated as erythematous plaques [3]. Morphologically, the neoplastic lymphoid infiltrate is epidermotropic and composed predominantly of small-to intermediate-sized atypical lymphocytes with enlarged hyperchromatic, cerebriform nuclei and clear cytoplasm (haloed cells). These atypical lymphocytes often colonize the basal layer of epidermis singly or in a linear fashion, forming a "string of pearls." Pautrier microabscesses, which consist of small aggregates of atypical lymphocytes often in association with Langerhans cells, can be helpful in the diagnosis [4, 5]. Traditionally, mycosis fungoides is divided into three stages-premycotic, mycotic and tumor stage [6, 7]. In the premycotic stage, the skin is erythematous, scaly and pruritic. The microscopic appearance may be similar to non specific dermatitis. In mycotic stage infiltrative plaques appear and biopsy shows polymorphous inflammatory infiltrate in dermis that contains some atypical lymphoid cells. In tumor stage dense infiltrates of atypical lymphoid cells with characteristic cerebriform nucleus expand the dermis [6].

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### Case Details

This case of ours is a 56 year old female patient who presented with multiple ulcerative nodules in size ranging from 1-3cm over the scalp. Multiple scaly itchy patches seen all over the body since 8 months.(figure.1). General examination revealed bilateral cervical and epitrochlear lymphadenopathy. Her general condition was fair. Her respiratory and cardiovascular systems were within normal limits.

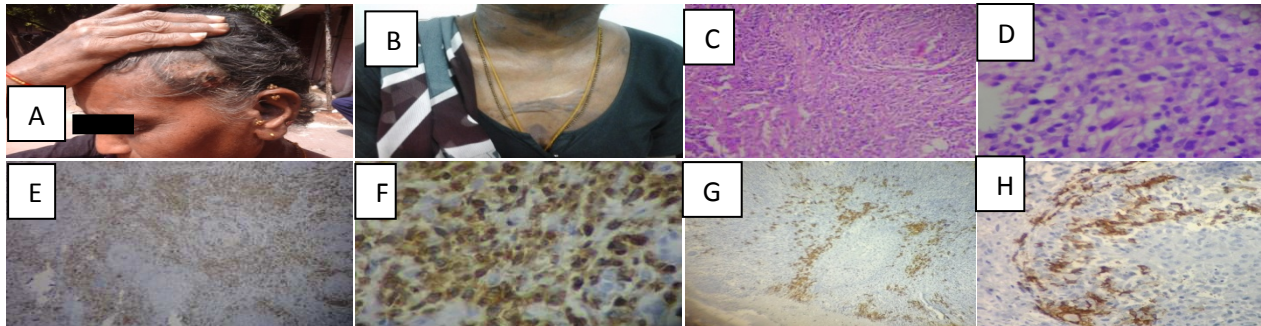
**Laboratory investigations** showed peripheral blood smear showed normocytic normochromic blood picture with adequate number of platelets and WBC count within normal limits. Mantoux and VDRL test were negative. Liver function tests and renal function tests were within normal limits. Ultrasonography of abdomen and Computerised tomography scan showed no organomegaly.

**Skin biopsy findings:** (figure 1) grossly received elliptical skin bits of 0.2cm. Routine paraffin embedding and processing was done. Section studied showed epidermis and dermis. Epidermis showed mild hyperkeratosis, thinning and ulceration focally. Dermis is infiltrated by atypical lymphoid cells, plasma cells and monocytoid cells. Epidermotropism and pautrier microabscess are also seen in the sections. Provisional diagnosis was given as atypical lymphoid hyperplasia /Mycosis fungoides and the tissue was subjected to immunohistochemical workup.

**Immunohistochemistry:** CD3 (PS1) diffuse cytoplasmic and membrane positivity in T cells and CD20 (L-26) positive in B CELLS.

Definitive diagnosis of mycosis fungoides was considered.

**Figure-1**



**A:** ulcerative nodule on the scalp. **B:** plaque over skin on the chest. **C, D:** skin biopsy, **E,F:** CD3 positivity in T cells. **G, H :** CD20 positivity in the B cells.

**Cervical Lymph node- Fine Needle Aspiration Cytology:** (figure 2) Cytosmears were cellular with few mature lymphocytes, atypical lymphoid cells that are two times larger than mature lymphocyte with nuclear lobation, plasma cells, clusters of epithelioid histiocytes, binucleate and multinucleated forms. Mitosis also noted. Provisional diagnosis as lymphoproliferative disorder was considered and excision was advised.

**Lymph node biopsy:** Grossly received lymph node specimen measuring 3\*2 \*1cm size. Cut section showed grey white areas. Tissue was subjected to routine paraffin embedding and processing.

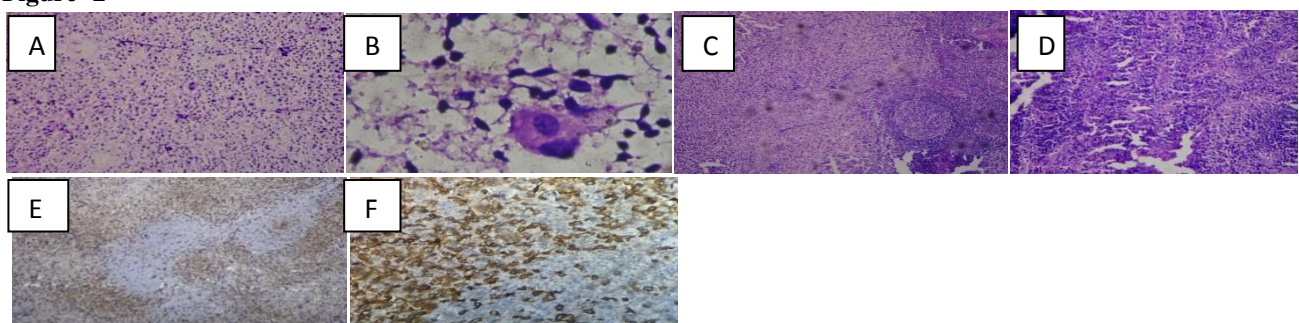
Sections showed complete effacement of lymph node with few reserved follicles. Parafollicular zones are expanded with diffuse sheets of atypical lymphoid cells with moderate amount of eosinophilic cytoplasm, cerebriform nuclei in some. Mitotic activity, Pigment laden histiocytes, sinusoidal dilatation with high endothelial proliferation was seen.

Provisional diagnosis of dermatopathic lymphadenitis/Nonhodgkins lymphoma-Tcell type was considered. Sections were submitted to immunohistochemical studies.

**Immunohistochemistry:** CD3 (PS1) diffuse positivity in the malignant cells, CD 68(KP1) negative, ALK-1(SP8) focal positivity.

Definitive diagnosis of Cutaneous T cell lymphoma infiltration into lymph node was considered.

**Figure- 2**



**A&B:** FNAC of lymph node. **C&D:** H & E stained section of lymph node. **E&F:** CD3 positivity of lymphnode

## Discussion

Mycosis fungoides is the T cell lymphoproliferative disorder that arises primarily in the skin and that may evolve into generalized lymphoma [8,9]. A viral etiology has been suspected because of certain similarities to HTLV-1 associated adult T cell leukemia- lymphoma [10].

The majority of the cases occur in adult males but adolescents can also be affected [7]. It has various manners of presentation and progression. Important prognostic parameters are stage at diagnosis, absence of complete remission after first treatment [11]. Once cutaneous spread takes place, prognostic parameters have no influence on survival and prognosis is bad [9].

Adnexotropic mycosis fungoides [12]. Skin of the face and scalp are most commonly involved. Hair follicles are infiltrated by characteristically atypical T cells resulting in the formation of Pautrier microabscesses and be associated follicular mucinosis where keratinocytes forming affected follicles produce intracellular mucin.

Folliculotropic MF is a variant of MF, which is characterized by the presence of follicular infiltrates of atypical cerebriform CD4+ T lymphocytes often sparing the epidermis. Most cases show numerous degenerations of the hair follicles (follicular mucinosis) but cases without follicular mucinosis have been reported.

Pagetoid reticulosis is a variant of MF is characterized by the presence of patches and plaques with an intraepidermal proliferation of neoplastic T-cells. The term should only be used for localized type. Granulation slack skin is also a variant of MF [13,14].

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