Mycosis Fungoides: Tumour d'emblee

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ABSTRACT

Mycosis Fungoides is a Cutaneous T-cell lymphoma characterized by infiltration of skin with patches, plaques, and nodules composed of T-lymphocytes. It is the most common type of Cutaneous T-Cell Lymphoma and accounts for almost 50% of all primary cutaneous lymphoma. Tumour d' emblee is the term used for the patient presenting with skin tumors not preceded by patches or plaques. We report a rare case of mycosis fungoides d' emblee variant with tumors of only 3 months duration without any preceding skin lesions.

Key words: Cutaneous T-cell lymphoma, Mycosis fungoides, nodules, tumour d'emblee

INTRODUCTION

Mycosis Fungoides (MF) is a Cutaneous T cell lymphoma (CTCL) characterized by infiltration of skin with patches, plaques and nodules composed of T-lymphocytes. The disease is extremely variable in its clinical course and presentation. Intense pruritus is a common symptom. Trunk and body folds are commonest site of involvement. It has various stages, premycotic, patch, plaques, nodules, tumours and erythroderma. Tumour d'emblee is a variant of tumour stage, which develops from normal skin without prior patch or plaque stage.[1,2]

CASE REPORT

A 55-year-old Hindu male, tobacco farmer by occupation, presented with multiple infiltrated plaques and nodules of 3 months duration over the face and scalp. The nodules started from ears and spread all over the face and scalp, increasing in size and number. Patient had a history of severe itching episodes for past three years which were not relieved by antihistaminics. There was no history of any preceding skin lesions. There was history of episodes of fever, weight loss, anorexia, and nausea for the past 2 months. He was a chronic smoker for the past 35 years. On examination, multiple infiltrated plaques and nodules with few erosions and foul smelling superficial ulcers were present over face and scalp. The infiltrated skin over forehead, nose and ear lobules and loss of eyebrows gave a leonine face appearance [Figures 1 and 2]. Bilateral cervical, post auricular, axillary and inguinal lymphadenopathy was noted. The nodes were discrete, non-tender, mobile, and firm in consistency. Loss of hair was seen on scalp, eyebrow, and axillary region. Systemic examination was normal and there was no hepatosplenomegaly.

Investigations revealed a hemoglobin level of 8.6% gm and the Erythrocyte sedimentation rate (ESR) of 110 mm/hour. Other routine blood and urine investigations were within normal limits. Mantoux test and slit skin smear for acid fast bacilli were negative. Chest X-ray, lymph node aspiration cytology and bone marrow examination did not reveal anything abnormal. Ultrasonography of abdomen and pelvis and computed tomography (CT) scan of head and neck, chest, abdomen, and pelvis were normal.

Histopathological examination of the excision biopsy from a nodule over face showed a lymphocytic infiltrate in the papillary dermis and around the hair follicle and pilosebaceous unit. [Figure 3] Many cells had cerebriform nuclei and clear cytoplasm. Epidermotropism was noted with formation of well defined pautrier's microabscess at places [Figure 4]. Follicular mucinosis was noted.

Immunohistochemistry showed positive CD3 and LCA markers as and CD 30 and CD 20 negativity. The investigatory evidence hence was consistent with the clinical diagnosis of Mycosis fungoides. A final diagnosis was Mycosis fungoides stage T3N1M0B0 (II-B).

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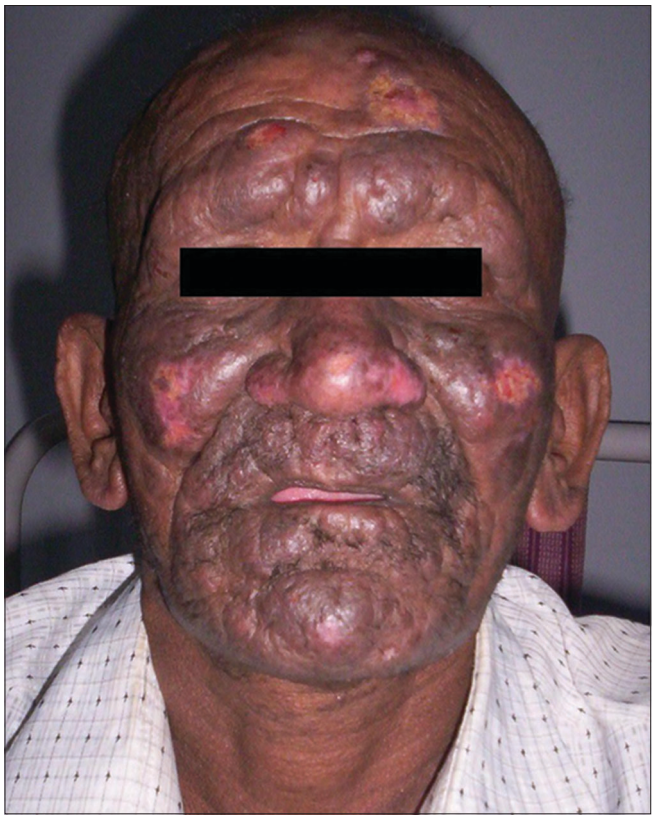

Figure 1: Nodules plaques and erosions over face

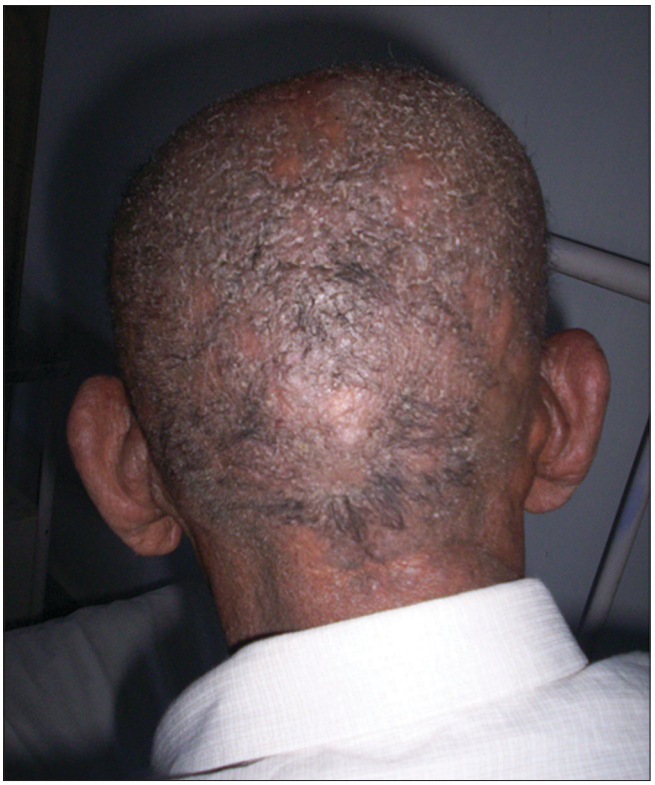

Figure 2: Lesions over scalp

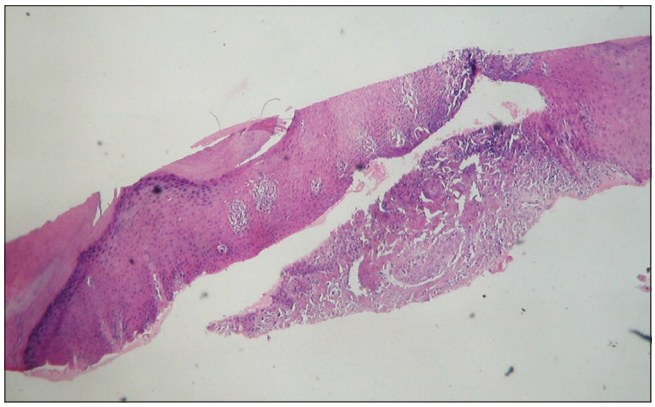

Figure 3: Biopsy showing epidermotropism (H and E, ×40)

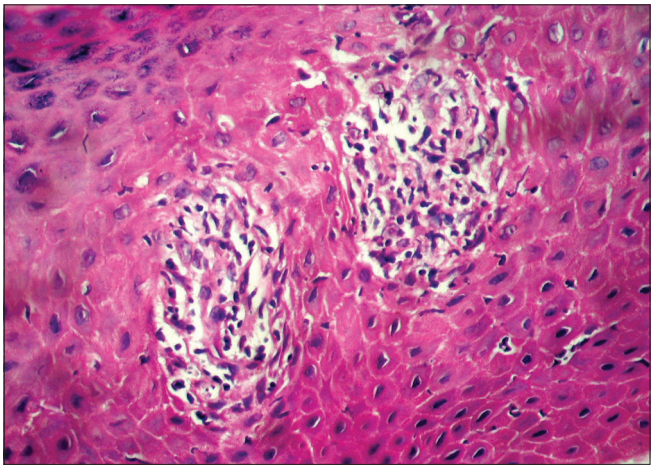

Figure 4: Biopsy showing clusters of atypical lymphocytes within the epidermis (pautrier microabscesses) (H and E, ×400)

DISCUSSION

Alibert first described the classic plaque form of Mycosis fungoides in 1806.[2] He termed it mycosis fungoides because of the resemblance of the lesions to "mushrooms." In 1885, Vidal and Brocq described mycosis fungoides d' emblee for a patient presenting with skin tumours not preceeded by patch or plaques.[3] In this type of MF, the tumors develop suddenly without the usual progression from eczematous or plaque stage. Tumours are the initial presentation in approximately 10 % of the patients.[4]

MF is the most common type of CTCL and accounts for almost 50% of all primary cutaneous lymphomas.[2,5] However other lymphoproliferative disease also involve the skin including Ki-1 +anaplastic large cell lymphoma, peripheral T-cell lymphoma, cutaneous B-cell lymphoma, adult T-cell leukaemia/ lymphoma, T-cell lymphoid leukaemia and cutaneous Hodgkin's disease.[4,6]

Incidence of MF has been estimated to range from 0.06 to 0.1 per 10,000 cancer cases per year in the USA. MF is approximately twice as common in men as in women. Blacks have twice the incidence of whites as suggested in some studies. Most cases are diagnosed in 5th and 6th decades (55–60 yrs).[2,7,8]

The term tumour d' emblee is now falling into disrepute and these tumors may, in fact, be pleomorphic CD 30 negative cutaneous T-cell lymphoma (peripheral T-cell lymphoma), which have undergone large cell transformation.[9,10] Many of these cases are likely to be classified by immunophenotyping as various types of non-MF T-cell lymphoma or even B-cell lymphoma of the skin.[11]

Such type of MF d' emblee has been reported rarely in past.[12-14] Many cases described as the d'emblee variant in the past may have represented other types of lymphomas.[15]

The CD 30 negative large CTCL and small/ medium sized pleomorphic CTCL have been described in literature to be presenting with tumors without prior or concurrent patches or plaques along with histological presentation sometimes similar to that of MF. The CD 30 negative large CTCL (5 year survival of 15%) has a poor prognosis compared with small/ medium sized pleomorphic CTCL (5 year survival of 60%).[16]

Usually the mean interval between appearance of skin lesions and definite diagnosis by histopathology is approximately 6 years,[2] However, in our case it was only 3-4 months. The patient was treated with CHOP regimen [cyclophosphamide, hydroxydaunorubicin (doxorubicin), oncoverin (vincristine), prednisone] plus methotrexate. Our patient died within 10 months of diagnosis despite initial improvement with chemotherapy.

Taking into account that this case of tumour d' emblee also showed typical histopathological changes along with CD30 negativity and the eventual death of the patient within short span of time after the diagnosis, it is possible that the patient had CD 30 negative large CTCL, which could not be confirmed owing to limited resources in our hospital set up.

This case is reported because of an acute and masquerading presentation of Mycosis fungoides reiterating the fact that CTCL can pose an enormous diagnostic challenge.

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