Case Report

Mycosis Fungoides: Uncommon Presentation

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Abstract

Mycoses fungoides is a common cutaneous T cell lymphoma. Tumor and ulcerative stages are advanced lymphoma. We report a case of mycosis fungoides that presented with ulcerated plaques and nodules over the body and infraorbital region, he was being treated as leprosy without improvement of the lesions. Diagnosis was established with clinical presentation and histopathology of the lesion.

Keywords: *Dermatitis, lymphoma, mycosis fungicides, ulcerated plaques*

Introduction

Mycosis fungoides (MF) is the most common presentation of primary cutaneous T cell lymphoma. (CTCL) MF has slow progression with well-defined clinicopathological features.[1] CTCL accounts for 4% of non-Hodgkin lymphoma. MF and sezary cell lymphoma are the two main subtypes of lymphoma. MF involves skin but in advanced stage it spreads to blood, lymph, and visceral organs. MF simulates various skin disorders (chronic dermatitis, leprosy, and psoriasis) leading to delayed diagnosis and treatment. We report a case that presented with ulceration on plaques and nodules with infraorbital ulcer being treated as a case of leprosy in elsewhere. This is a rare case as patient reported with bilateral ulceration in infraorbital area which was the main presenting complaint to the author.

Case Report

A 46-year-old male farmer presented to Doon Hospital with widespread patches, plaques, nodules ranging from 5–6 mm to 5–10 cm over trunk, limbs, and face for 3 years, few ulcerated plaques and nodules were present over chest, abdomen, back and left infraorbital area for 6 months. [Figure 1 and 2] The patient started developing asymptomatic red patches over thigh and back for 3 years which gradually involved limbs; trunk, neck, and face within a year. They were slowly progressive and associated with slight itching. The development of multiple red raised scaly lesions brought the patient to local doctor where he was started with antileprotic treatment, patient took treatment for 1 year but condition deteriorated and he developed nodules and ulcers over some lesions with loss of appetite and weight which prompted his referral to hospital. Physical examination revealed multiple erythematous slightly scaly patches, plaques, and nodules ranging from 5–6 mm to 5–10 cm present over both upper and lower limbs, with extensor predominance. [Figure 2]

Ulcerated plaques and nodules were present over chest, abdomen, back, and face. These ulcers were having necrotic center. Largest ulcer was present in the left infraorbital area, this was 9–10 cm in size, nontender, involving lower eyelid. Local examination revealed cervical lymphadenopathy. Systemic examination was unremarkable except mild anemia (Hb 9 g%). Chest X-ray, ultrasonography of whole abdomen showed no organomegaly. Lesional biopsy was done that showed band-like papillary dermal lymphoid infiltrate and intraepidermal lymphocytes out of proportion with spongiosis (epidermotropism) with pautrier microabscesses, lymphocytes tagging the junction and within the epidermis showing haloes and variable nuclear pleomorphism, dermal reticular fibroplasia was also appreciable. Description based on clinical presentation and histopathology a final diagnosis of MF was entertained. The patient was started with oral steroids 40 mg/day and referred for further management. This patient was subsequently improved with chemotherapy.

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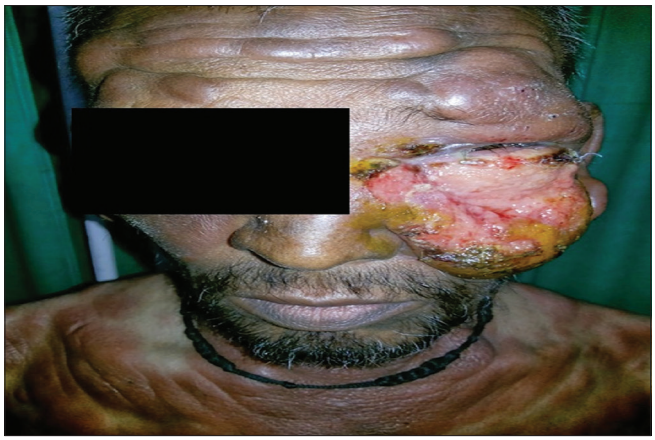

Figure 1: Case of mycosis fungoides: Closer facial view

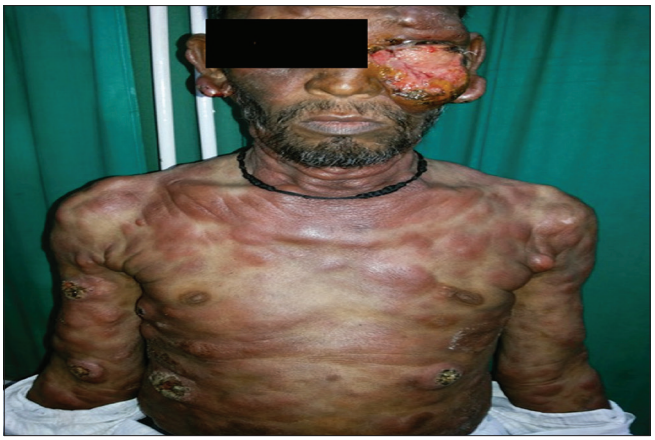

Figure 2: Case of mycosis fungoides: Far view

Discussion

MF is a T cell lymphoproliferative disorder that arises primarily in the skin and may evolve into generalized lymphoma.[2,3] It has protean manifestations premycotic, mycotic and tumor are the three stages of MF. Premycotic stage is characterized by erythematous, scaly, and mildly pruritic lesions. In mycotic stage, infiltrative plaques enlarge slowly and cover approximately 10% of body resembling exfoliative dermatitis. Tumor stage lesions resembles mushrooms, tumor appear rounded and lobulated ranging 1–15 cm in diameter. In tumor d'emblee form of the disease, affected individuals may develop large nodules without previous formation of plaque.

The incidence of CTCL increases with age, with an average onset between 50 and 60 years. Although this disease is less prevalent in children, people of all ages can be affected. MF with *de novo* ulceration is a rare presentation. Ulcer usually develops in tumors and this occurs because of fast tumor growth and this is associated with poor prognosis and high morbidity.[1,4] In this index case, ulcers were present over plaques. Few such cases have been reported in this (plaque) stage of MF who are being treated with methotrexate and in coexistence with CD30+ cutaneous T-cell lymphoma mimicking pyoderma gangrenosum in a patient with ulcerative colitis.[5,6] The index case presented *de novo* with ulceration without any history of ulcerative colitis or taking methotrexate. Estimated MF prevalence in the eyes and surrounding structures is low (2%) and among those having visual symptoms it is 26.7–40.4%.[7-9] Incidence is possibly underestimated given the difficulty in diagnosing it and late diagnosis, in addition to clinical variants. Primary eye conditions, prior to skin repercussions, are extremely rare. The eye lids and eye surface are usually the most affected areas, particularly in more advanced stages. Ectropion is the most common manifestation on the eyelids, caused by tumors or cutaneous infiltration. Case management is complicated regardless of the mechanism and may compromise eye surface integrity.[7-9] Ophthalmic abnormalities in patients with cutaneous T-cell lymphoma are relatively uncommon, the symptoms of the disease are wide and different leading to difficulty in treatment.[10] This patient presented with erythematous scaly ulcerated plaques which were earlier being treated as leprosy with multi drug therapy but were not responding to treatment. In cases of nonresponding ulcerative plaques, careful correlation of clinical and pathological findings, repeated biopsy may be required to avoid missed and delayed diagnosis of MF.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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