AN INTERESTING CASE OF MYCOSIS FUNGOIDES

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Abstract: Mycosis fungoides is the most common of the primary cutaneous T cell lymphomas. We describe a 58 year old male who presented with features of plaque stage of Mycosis fungoides. The lesions initially started around the hip as hyperpigmented plaques which was followed by similar lesions over legs, trunk, arms and other regions of the body gradually in 10 yrs. History of loss of weight and appetite was present. Dermatological examination revealed multiple well-defined hyperpigmented thickened scaly plaques of varying sizes present over trunk, face and extremities. Right axillary lymphnodes were enlarged, firm, discrete and nontender. Immunohistochemistry showed CD 3 and CD 4 positivity and histopathological examination was consistent with Mycosis fungoides. The patient was treated with 6 cycles of Total Skin Electron Beam Radiotherapy, followed which he showed dramatic improvement in skin lesions. We report this case for its rarity and interesting presentation.

Keyword: Immunohistochemistry, Epidermotropism, Mycosis fungoides, Total Skin Electron Beam Radiotherapy.

INTRODUCTION:
We report an interesting case of Mycosis fungoides (MF) in a male, who was successfully managed. MF is the most frequent variant of Cutaneous T Cell Lymphomas (CTCL), generally associated with an indolent clinical course and characterized by well-defined clinicopathological features. It usually begins in mid to late adulthood (median age at diagnosis is 55 to 60 years) with a male to female ratio of 2:1.

CASE REPORT:
A 58 year old male, presented with history of itchy skin lesions over trunk and extremities since 10 years and non-healing ulcer over the right arm of 1 year duration. The lesions initially started around the hip as hyperpigmented plaques which was followed by similar lesions over legs, trunk, arms and other regions of the body gradually in 10 yrs. History of loss of appetite and weight was present. Dermatological examination revealed multiple well-defined hyperpigmented thickened scaly plaques of varying sizes present over trunk, face and extremities. Hyperpigmented patches were seen over the face and neck. A single nodular ulcerative plaque of 5 × 4 cm size was present over the middle one-third of lateral aspect of right arm. Right axillary lymphnodes were enlarged, firm, discrete and nontender. Scaling was seen over the palms and soles. Oral mucosa showed no lesions.

Figure 1: Hyperpigmented scaly plaques over front of trunk and arms.
Investigations revealed no abnormality in his complete blood count. Blood urea, sugar, creatinine and liver function tests were normal. Peripheral smear showed no atypical lymphocytes. X-ray chest, ultrasonogram abdomen and pelvis revealed normal study and there was no evidence of internal organ involvement. With these findings, a provisional diagnosis of cutaneous lymphoma was made and the patient was subjected to further investigations. Bone marrow biopsy showed cellular marrow with mild lymphocytosis. Histopathological examination revealed epidermis with hyperkeratosis, confluent parakeratosis with a small focus of atypical lymphocytes, irregular acanthosis with broad elongated rete pegs. Dermis showed band-like infiltrate of atypical lymphoid cells [Figure 5, 6], moderate nuclear pleomorphism and few mitotic figures. [Figure 7]. Immunohistochemistry revealed CD3, CD4 positivity and CD 30, CD 20 negativity, consistent with Mycosis fungoides. Thus, a final diagnosis of Mycosis fungoides, stage 2 A, T2/ N1/ M0 / B0 was made.

The patient was treated with 6 cycles of Total Skin Electron Beam Radiotherapy (TSEB) for 6 weeks, after which he showed dramatic improvement in skin lesions. [Figure 8, 9, 10]
DISCUSSION:
Mycosis Fungoides is the most common variant of Primary CTCL, with an indolent clinical course and well-defined clinopathological features. The etiology is unknown, but one of the suggested theories is that it is a disease of antigen (unknown) persistence associated with chronic lymphocyte stimulation and eventual transformation of benign lymphocytes to a low grade malignant T cell lymphoma[1].

Classical MF is characterized by typical cutaneous stages of disease consisting of patches and plaques involving less than 10% of the body surface area (stage T1/IA), more than 10% of the body surface area (stage T2/IB), tumours (stage T3/IB) and erythrodermic stage (stage T3/III). The various stages in MF are patch stage, plaque stage, tumour stage and erythrodermic stage. Patch stage is characterized by subtle fine scaly and often atrophic erythematous patches on the trunk, usually involving the limb girdle areas, breast and buttocks [2]. Plaques are more obvious persistent polymorphic erythematous lesions with a similar distribution, but with the development of stage T2/IB, usually involving head, neck, limbs and trunk. There may be associated scaling and pruritus. Rarely individual plaques may become eroded, ulcerated and painful. Tumours show considerable variations in size and the patients with erythrodermic stage usually have severe pruritus. Histological involvement of lymph nodes and other organs is a poor prognostic sign. Any organ such as the liver, spleen, bone marrow, lungs, bone and central nervous system can get affected.

The rare variants of MF are verrucous/hyperkeratotic MF, bullous type, pustular, ichthyosiform, panniculitislike, hyperpigmentary MF, folliculotrophic hypopigmented variant, pagetoidreticulosis, poikilodermatous type and granulomatous slack skin. Histopathological changes include epidermis with presence of mononuclear cells without spongiosis (epidermotropism). Clusters of atypical lymphocytes (pautrier microabscess) and selective single colonization of a typical T cells along the basal layer. Dermis reveals predominant lymphocytic infiltrate in the papillary dermis. As disease progresses, dermal infiltrate becomes more diffuse with more mitotic figures. Immunohistochemistry will show tumour cells with CD 2, 3, 4, 5 positivity and CD 7, 8 negativity. The differential diagnosis includes chronic dermatitis, psoriasis, contact dermatitis, atopic dermatitis, eczema and tinea corporis for the patch / plaque stage, and B cell lymphoma, sarcoidosis, deep fungal infections a typical mycobacterial infections, leprosy and leishmaniasis for the tumour stage. Treatment options [6] include PUVA or UVB with/without INF - alpha, topical corticosteroids (for limited patches, thin plaques), local radiation therapy (single lesion) or TSEB (for generalised thick plaques), topical or systemic cytotoxic drugs for stage IA to IA disease; PUVA + INF - alpha, oral retinoids + INF - alpha / PUVA, TSEB and local radiation therapy for stage IB disease; PUVA A + INF - alpha / oral retinoids, topical or systemic cytotoxic drugs, TSEB or radiation therapy to all skin in the body, extracorporeal photopheresis for stage III disease and systemic cytotoxic drugs, systemic retinoids, biologic response modifiers – denileukin diftitox or IL-12, TSEB and/or radiation therapy to all skin in the body for stage IV disease.

CONCLUSION:
This case is being reported to keep a high index of suspicion of cutaneous T cell lymphoma in any patient presenting with psoriasiform dermatitis, as early diagnosis and early intervention gives cure or near cure to the patient and improves the quality of life, unlike other cases of lymphoma

REFERENCES:
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