HYPOPIGMENTED MYCOSIS FUNGOIDES TREATED SUCCESSFULLY WITH PUVA

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Hypopigmented lesions are rarely encountered in mycosis fungoides. We here report a 22-year-old female patient who presented with a 5-year history of asymptomatic progressively increasing discrete and confluent hypopigmented macules and a 1-year history of a few itchy erythematous, scaly, indurated plaques. The histological features were consistent with a clinical diagnosis of mycosis fungoides. She was successfully treated with PUVA therapy.

Key Words : Mycosis fungoides, PUVA

Introduction

Mycosis fungoides (MF) is a form of cutaneous T-cell lymphoma which usually presents as infiltrated scaly plaques, nodules and tumors.[1] Atypical manifestations of MF include follicular mucinosis, lymphomatoid papulosis, granulomatous slack skin, pustular, bullous, hyperkeratotic and verrucous forms.[2-4] Cutaneous pigmentary changes are well known in MF. Hypopigmentation and hyperpigmentation are regular features of poikiloderma atrophicans vasculare as well as healed lesions of MF.[5] However, pure macular hypopigmentation is an extremely uncommon presentation of MF.[6,7] We here report a case of MF in which vitiligo-like hypopigmentation was the only initial manifestation of MF.

Case Report

A 22-year old young woman presented with complaints of asymptomatic hypopigmented macules on the extremities, face and the trunk. She first noticed these lesions on the extensor surface of the forearms 5 years back and since then the lesions gradually progressed to involve the other parts of the body within three years. Individual macules gradually coalesced to form large areas of hypopigmentation interspersed with islands of normal skin. For the last one year, the patient had developed itchy, erythematous, scaly plaques on the left side of the neck, right shoulder, left axilla and both thighs. Plaques slowly progressed in size with no remissions or seasonal variations. There was no history of significant weight loss, fever or anorexia. Family and personal histories were non-contributory.

General physical examination of the patient revealed bilateral, discrete, non-tender, firm and mobile axillary lymph nodes of 2-3 cms size. There was no hepatosplenomegaly. Cutaneous examination showed hypopigmented, non-scaly macules on the face, trunk and extremities. Some of the lesions on the face and the trunk had coalesced to form large area of hypopigmentation with small discrete islands of normal skin. The lesions showed no atrophy or telangiectasia. In addition, the patient had

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erythematous, scaly, indurated plaques varying in size from 5x5 cms to 5x10 cms involving the left side of the neck, left axilla, both thighs and the right shoulder. The plaque on the right shoulder showed mild atrophy but no telangiectasia.

Skin biopsy from both the hypopigmented as well as the indurated plaque showed essentially similar histopathological features. The dermis was infiltrated with atypical appearing lymphoid cells. At places the cells were seen to invade into the epidermis and form intra-epidermal collections suggestive of Pautrier's micro-abscesses. These cells were positive for leucocyte common antigen and negative for B-cell markers. Routine haemogram, liver and renal function tests and skiagram of the chest were within normal limits. No atypical cells were seen in peripheral smear. Fine needle aspiration cytology of axillary lymph node showed features of dermatopathic lymphadenopathy. Utrasonogram of abdomen was normal. Bone marrow aspiration cytology revealed no involvement with tumor cells.

The patient was treated with 8-methoxypsoralen plus UVA light (PUVA) for stage IIA MF. Treatment was given thrice a week. After 4 months of PUVA therapy (33 treatment sittings) the patient showed almost complete clearance of the indurated plaques. The hypopigmented lesions which started showing peri-follicular repigmentation after 12 treatments had significantly repigmented after 4 months of therapy. A post-treatment histopathological examination revealed no evidence of tumor cells.

Discussion

Hypopigmented macules without associated poikiloderma is a rare presentation of mycosis fungoides. So rare is the presentation that it has been reported in less than 10 patients in the literature. Interestingly, all the earlier patients have been coloured and with the exception of one patient have all been males. It is also interesting that almost all the patients, including our patient were either adolescents or young adults[7] - this is in contrast to MF in general in which 81% of patients are above 45 years of age.[9]

The hypopigmented lesions of MF occasionally progress to plaque lesions.[6] Sometimes, however lesions retain their macular character for years and are variously misdiagnosed as pityriasis alba, pityriasis versicolor, leprosy and early vitiligo. All these conditions are common in the coloured races and in the absence of any clinical leads, skin biopsy is recommended in patients when the hypopigmented lesions are persistent or recur despite appropriate therapy.

The cell marker studies, from the hypopigmented lesions have shown predominantly CD4 antigen-positive cells with complete loss of CD7 antigen.[7] CD7 antigen is otherwise normally expressed by 100% of CD8 positive cells (T-suppressor / cytotoxic cells) and 90% of the CD4 positive cells (T-helper / inducer cells). However, the value of cell marker studies in the diagnosis of mycosis fungoides in still controversial.[7]

Ultrastructural studies from five patients with hypopigmented mycosis fungoides have shown focal invasion of the epidermis by mycosis cells. The adjacent melanocytes and keratinocytes showed degenerative changes. The majority of melanocytes exhibited swelling of cytoplasmic organelles and defective melanogenesis with production of spherical incompletely melanized melanosomes. These changes would appear to be a non-specific response to cell injury associated with inflammation and may be related to ischaemia consequent upon disruption of the normal epidermal architecture.[5]

In some patients the hypopigmented lesions progress to plaques sometimes after as long as 20 years.[3,6] It is not known whether biopsy specimens taken from the hypopigmented macules in these, prior to the development of plaques, showed any features suspicious of my-

cosis fungoides. Our patient had the hypopigmented lesions for almost 5 years before she developed the plaque lesions. She had been variously treated as pityriasis alba, pityriasis versicolor and vitiligo. An earlier biopsy taken during the macular stage of the disease was non specific and the patient had then been treated with dapsone probably with a diagnosis of leprosy.

Most of the patients of MF with hypopigmented lesions have been treated with PUVA therapy with repigrnentation and decrease or disappearance of the infiltrate.[5] Our patient also responded to PUVA therapy both clinically and histologically.

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