Mycosis fungoides and alopecia areata: A case report

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Mycosis fungoides (MF), the most common type of cutaneous T-cell lymphoma, is histologically characterized by atypical T lymphocytes with cerebriform nuclei that infiltrate the epidermis. Alopecia can be a manifestation of mycosis fungoides (MF) but alopecia areata (AA) is not usually associated with MF. We present a case of hypopigmented erythematous MF and AA with a childhood onset.

Keywords: mycosis fungoides, alopecia areata, childhood, hypopigmentation

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INTRODUCTION

Mycosis fungoides (MF) is the most common type of cutaneous T-cell lymphomas.

It is characterized by pleomorphic lesions including patches, plaques, cutaneous tumors, dyspigmentation, or erythroderma and alopecia is not common [1]. Alopecia areata (AA) is also a T-cell mediated disorder directed at anagen hair follicles presenting with non-scarring patchy hair loss that can progress to total alopecia. AA is associated with other autoimmune diseases such as vitiligo, thyroid diseases, pernicious anemia, and atopic dermatitis but is not usually associated with MF [2]. We present a case who developed an erythematous plaque, a few hypopigmented patches, and multiple alopecic regions on his legs simultaneously in childhood.

CASE REPORT

A 22-year-old male with skin phenotype IV was visited in the dermatology outpatient clinic of Razi Hospital, Rasht, Iran. He had an erythematous plaque without any symptoms on the anteromedial aspect of his right thigh with a childhood onset. In addition, there were multiple patches of non-scarring hair loss on his both legs without any symptoms that were noticed simultaneously when he was almost six years old for the first time (Figure 1.a-c).

On clinical examination, there was a 10cm × 8 cm pink scaly hairless plaque on the anterior aspect of the right thigh. Multiple regions of patchy alopecia were also seen on both lower limbs and buttocks without exclamation mark hair, skin atrophy, and erythema. The pull test was negative. Alopecic patches covered about 15 to 20% of the body surface area.

Two weeks after the initial examination, two hypopigmented patches were noticed in the alopecic area beside erythematous plaques on his right thigh and back of the right shin but we were not sure whether these lesions were new or old.

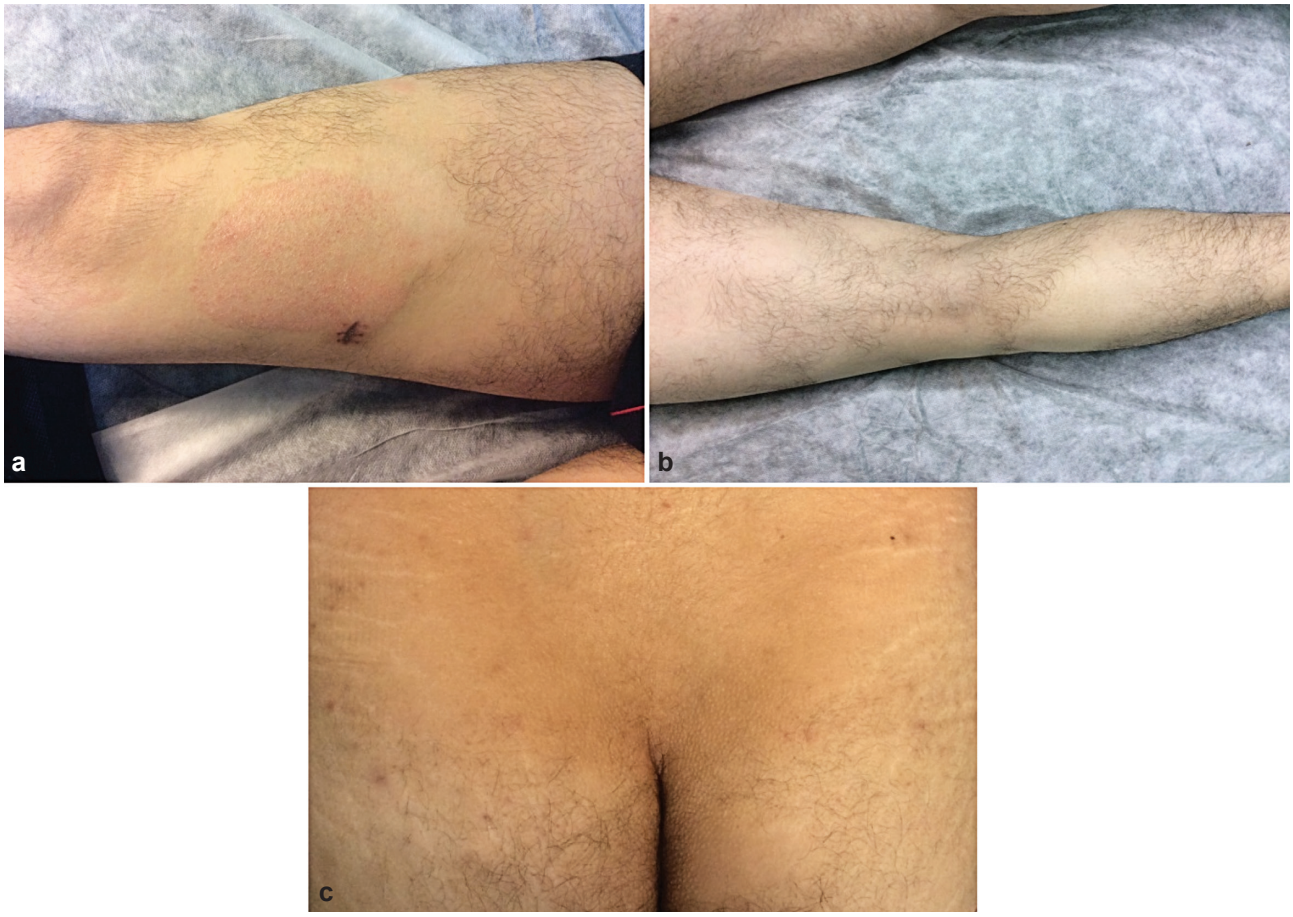

Figure 1.1.a. An erythematous plaque and alopecic patches on the right thigh.**1.b.** Multiple alopecic patches on the left thigh. **1.c.** Alopecic patches and folliculitis on the buttocks.

Our patient was atopic and his family history was negative for alopecia areata, vitiligo, and leprosy. He was a farmer and had contact with pesticides.

On physical examination, there was no organomegaly or lymphadenopathy. There was no alopecia on the scalp and face. His nails were normal and rest of the skin physical examination revealed folliculitis in the lumbosacral area.

The patient was treated with topical steroids (triamcinolone acetonide ointment) for the erythematous plaque with no significant improvement but he refused to receive additional treatment regarding alopecic patches on the lower limbs.

Skin biopsy was made from three sites: the erythematous plaque, hypopigmented patches near the erythematous plaque, and the alopecic area on his right thigh.

Pathological study of the erythematous lesion revealed lymphocyte alignment along the dermoepidermal junction with focal migration of atypical lymphocytes into the epidermis and formation of Pautrier microabscesses. There were also superficial perivascular lymphocytic infiltration, pigmentary incontinence, and cytoid bodies in the upper dermis (Figure 2.a).

Immunohistochemistry revealed epidermal infiltration of atypical T lymphocytes with a predominance of CD4 over CD8 (Figure 2.b) and negative CD30.

Neither follicular involvement by atypical cells nor histology of follicular mucinosis was present. Skin biopsy of the hypopigmented area revealed reduced melanocytes in the basal layer. T-cell clonal study was not performed.

Vertical sections of the skin of the alopecic patch showed a normal epidermis, mild congestion of superficial dermal vessels, decreased follicular density, perifollicular fibrosis, degenerative changes of the follicular epithelium, and sparse infiltration of perifollicular inflammatory cells. No epidermotropism and dermal mucin deposition

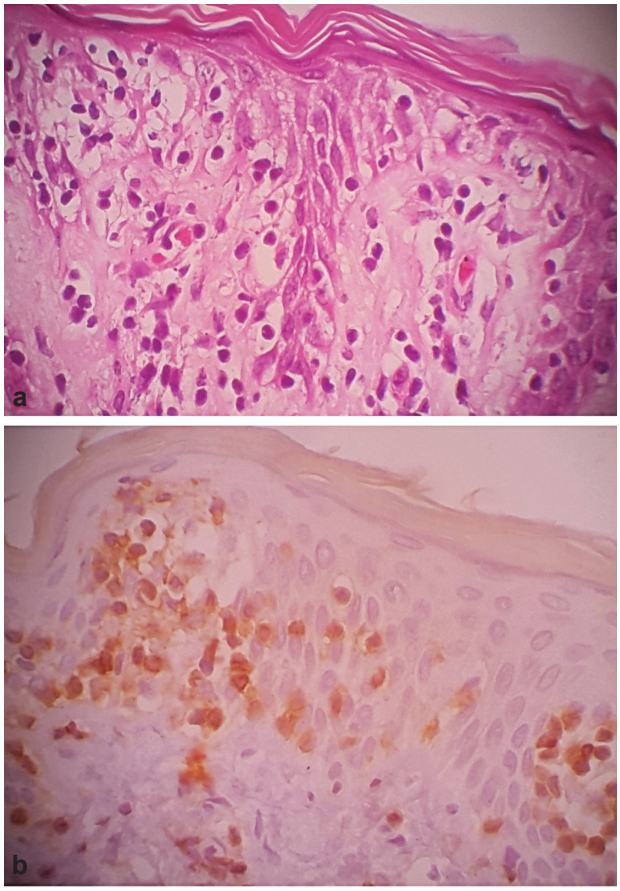

Figure 2. 2.a. Focal migration of atypical lymphocytes in to the epidermis and formation of Pautrier microabscess and cystoid body in upper dermis (H&E, 40×). **2.b.** Immunohistochemistry showed epidermal infiltration of atypical T lymphocytes with a predominance of CD4 (CD4 receptor staining, 40×).

was detected. All these findings are seen in late stages of alopecia areata, so folliculotropic MF was ruled out.

Other laboratory tests and abdominopelvic ultrasound were normal and Sezary cells were absent on the peripheral blood smear.

DISCUSSION

Our patient was interesting because he had alopecia areata and plaque type and hypopigmented MF that started simultaneously when he was six years old.

In 2011, Yang Bi *et al*. reported three clinical patterns of hair loss in MF as follows: patchy hair loss similar to alopecia areata, localized hair loss within overt MF lesions, and total-body hair loss which was seen only in patients with generalized erythroderma and SS. In this study, hair loss mimicking clinically alopecia areata was found in 34% of the patients with MF/SS and was more common in the scalp without pathologic criteria of AA. However, there was no self- report of coexisting AA and MF in the national registration system for AA patients [3].

Follicular involvement with either benign or malignant T cell infiltration could result in alopecia. Hence, clinical suspicion is very important in evaluation of patients with AA like hair loss.

Folliculotropic MF may also be an imitator of AA [4], but our patient did not have folliculotropic MF on pathologic study.

In general, atypical T lymphocyte infiltration within alopecic areas suggests that MF contributes to hair loss but benign peribulbar or intrabulbar T-cell infiltration indicates AA.

Interestingly similar HLA-DR and DQB alleles have been found in ME and AA, which are responsible for antigen presentation restriction. So, common peptide antigens could trigger T cells in both AA and MF. These peptide antigens may be derived from either self or other agents. On the other hand, a common pathogenesis has been proposed for benign AA and malignant F-MF (Folliculotropic MF) [3]. Therefore, AA may change to folliculocentric or patch stage MF as a disease spectrum [5].

Our patient could also be considered as a case of juvenile and hypopigmented MF. The incidence of juvenile MF is low, but hypopigmented MF has been frequently reported in children and dark skin patients like our patient [6,7].

Our patient also had folliculitis, especially on the buttocks. Interestingly, Creed *et al.* reported two patients with clinical presentation of AA preceding MF who both had coexisting folliculitis with gram positive cocci [5]. *Staphylococcus* might play a role via its putative antigen in MF and persistent antigen stimulation may be a triggering factor [8].

In such cases, AA could be a preceding feature for MF or a misdiagnosed MF. There is probably a different unknown spectrum of immune dysregulation that begins from benign T-cell proliferation affecting hair follicles in the form of AA to malignant T-cell proliferation in the form of folliculotropic MF [5].

In conclusion, this case was interesting because he had simultaneous presentations of benign and malignant T-cell dependent immune dysregulations with a childhood onset.

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