A 33-year-old woman was visited at the dermatological clinic of Loghman-e-Hakim Hospital with multiple hypopigmented patches on her right arm, buttocks and flanks. Her lesions developed over the past year with no regression. Lesions were resistant to multiple antifungal therapies. Clinical examination revealed multiple circular or oval patches on the right arm, flank and sacral area. There was no erythema but fine scales were observed on the lesions (Figure 1). No cutaneous sensory deficit was detected and no abnormality was found on physical examination. Also, there was no family history of similar problems. Abdominal ultrasonography and chest x-ray were normal. We performed biopsy from one of the lesions.

What is your diagnosis?

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Conflict of interest: none to declare

Received: January 15, 2011  
Accepted: August 1, 2011
Histopathology finding

Histopathological examination showed hyperkeratosis and irregular acanthosis accompanied by exocytosis of atypical lymphocytes in lower part of the epidermis, some of them with large and irregular hyperchromatic nuclei. There was no spongiosis. Pautrier’s microabscesses were observed within epidermis. Infiltration of lymphocytes and shedding of pigment with mild RBC extravasation was detected within papillary dermis (Figure 2).

Hypopigmented Mycosis Fungoides

Hypopigmented Mycosis Fungoides (MF) is a rare variant of cutaneous T-cell lymphoma, first described in 1978. Since then, only a hundred cases have been reported. Hypopigmented MF is typically observed in dark-skinned individuals of Asian or African descent, especially children 

The pathogenesis of hypopigmented MF remains to be clarified. It is suggested that the hypopigmentation is due to the cytotoxic effect of T-suppressor lymphocytes on the adjacent melanocytes. Ultrastructural studies show focal invasion of the epidermis by mycosis cells admixed with degenerative changes in neighboring melanocytes and keratinocytes. The majority of melanocytes exhibit disorganized melanogenesis with production of spherical partly melanized melanosomes. The precise incidence of hypopigmented MF is yet unknown, probably because of under or misdiagnosis

Mycosis fungoides can manifest with distinct various clinical patterns. In addition to the well-known plaque and poikiloderma forms, atypical features include pustular, bullous and verrucous forms. Hypopigmented patches are an uncommon manifestation of MF and belong to the group of CD8+ cutaneous T-cell lymphomas in the majority of cases.

In hypopigmented MF, the predominant location of the hypopigmented lesions is on non-photodistributed areas of the body and their persistent nature can be a clue to diagnosis. The differential diagnosis of hypopigmented macules includes leprosy, pityriasis lichenoides chronica and most commonly, post-inflammatory hypopigmentation, vitiligo, atopic dermatitis, tinea versicolor, pityriasis alba, parapsoriasis and sarcoidosis. Histopathologic examination is still the best method for the diagnosis of hypopigmented MF. The gold standard for treatment appears to be PUVA, but repigmentation has been reported following treatment with Carmustine (BCNU) and Mechlorethamine. Hypopigmented MF usually responds well to therapy and has a biologically benign course, but recurrences after therapy are common.

In conclusion, persistent or odd hypopigmented lesions should be evaluated through biopsy to avoid delay in the diagnosis of MF, especially in young people.

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