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Dry Scaly Patches and Nodules in a Middle-aged Man

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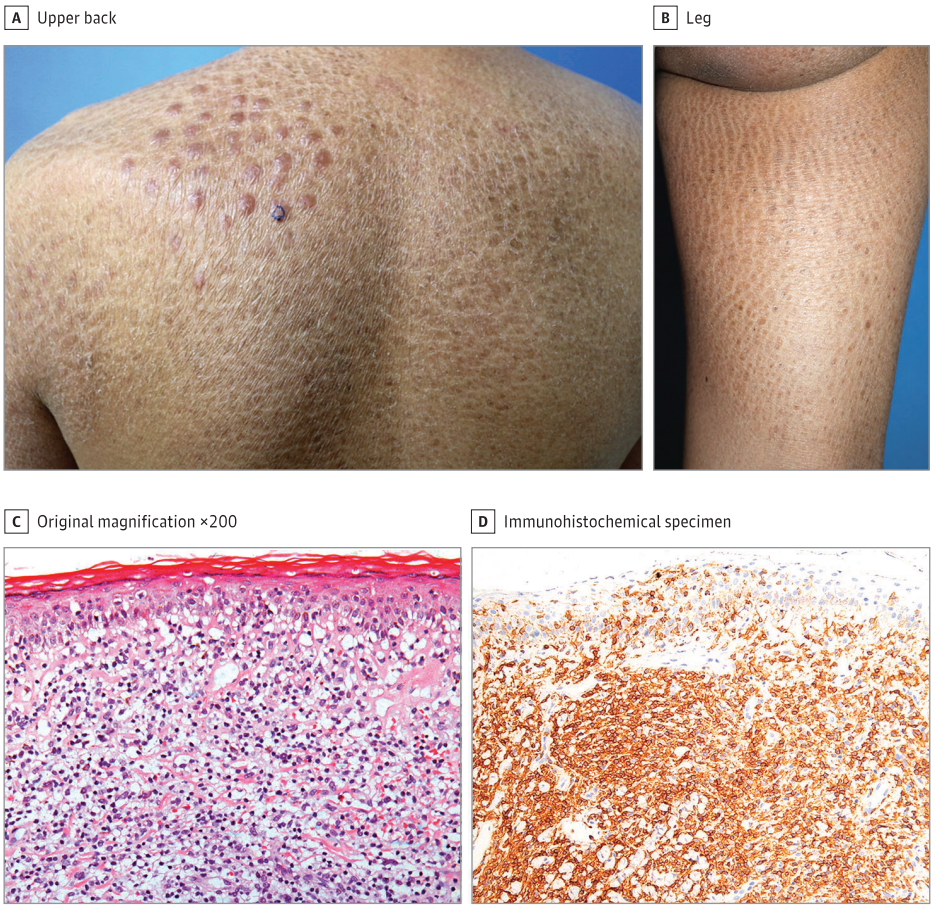

Figure. Clinical images show multiple dome-shaped nodules on the back (A) and extensive, dry scaly patches on the leg (B). Lesional histopathologic images show small to medium-sized atypical cells, and focal areas of epidermotropism (C, hematoxylin-eosin) and CD4 on immunohistochemical analysis (D) (original magnification ×200).

A man in his 50s presented with a 12-year history of itchy eruptions over the whole body. Multiple nodules appeared on the upper back 1 year previously and gradually increased in number and size. The patient was otherwise healthy and denied a family history of inherited ichthyosis or atopy. Physical examination revealed extensive, dry scaly patches on the trunk, arms, and legs. Several 0.5- to 1-cm, dome-shaped, red papules and nodules were found on the upper back (**Figure**, A and B). Mildly enlarged lymph nodes were observed on the cervical, axillary, and inguinal regions. Results of routine blood tests, biochemistry analyses, and peripheral blood flow cytometry tests were within normal limits. No atypical cells were found in the peripheral blood. A biopsy specimen was obtained from the nodules on the patient's back (Figure, C and D).

WHAT IS YOUR DIAGNOSIS?

A. Cutaneous pseudolymphoma

B. Ichthyosiform mycosis fungoides

C. Ichthyosis vulgaris with atopic dermatitis

D. Large plaque parapsoriasis

Diagnosis

B. Ichthyosiform mycosis fungoides

Microscopic Findings and Clinical Course

Histopathologic examination revealed an atrophy of the epidermis with focal orthokeratosis and an extensive, dense infiltrate of lymphocytes in the entire dermis. Small to medium-sized, atypical lymphocytes with focal areas of epidermotropism could be observed (Figure, C). Immunohistochemical staining results were consistent with a T-helper phenotype of the infiltrating lymphocytes, which were positive for CD2, CD3, CD4 (Figure, D), and CD45RO. A few small reactive $CD8^+$ T cells and $CD7^+$ T cells were observed in the dermis. DNA was extracted from paraffin sections, and polymerase chain reaction detected a clonal amplification product for the γ-chain of the T-cell receptor.

The patient was treated with narrowband UV-B 3 times weekly, interferon α-2b, 3 million international units/m^2, 3 times weekly and oral methotrexate, 10 mg once weekly. The patient had complete resolution of the skin lesions and enlarged lymph nodes at 6-month follow-up.

Discussion

Mycosis fungoides (MF) is the most common type of cutaneous T-cell lymphoma, usually with an indolent clinical course.[1,2] Many clinical variants of MF have been reported, such as hypopigmented and hyperpigmented, follicular, and hyperkeratotic-verrucous presentations.[2] Ichthyosiform mycosis fungoides (IMF) was first described by Kütting et al[3] in 1996. It is a rare variant of mycosis fungoides that is present in 1.8% to 3.5% of patients with mycosis fungoides (MF).[4] Since the first report, very few cases have been reported in the literature.[4]

Acquired ichthyosis usually begins in adult life and manifests as dry, scaly patches and plaques or as a generalized eruption resembling ichthyosis vulgaris.[5] This disease can result from a wide range of underlying causes, such as neoplasms, malnutrition, infectious diseases, sarcoidosis, and inflammatory disorders.[6] Acquired ichthyosis is recognized as a cutaneous manifestation associated with malignant diseases, including Hodgkin lymphoma, non-Hodgkin lymphoma, multiple myeloma, and MF.[2]

When acquired ichthyosis is related to MF, it can behave as a paraneoplastic syndrome or as a specific clinical variant of the lymphoma, that is, IMF.[2,6] In the former condition, the ichthyosiform areas are usually completely separate from areas with conventional MF appearance, and a skin biopsy specimen of the ichthyotic lesions only shows epidermal hyperplasia. In IMF, the ichthyosiform eruptions are a specific manifestation of the lymphoma, and its histological presentation includes the typical findings of both MF and ichthyosis vulgaris.[1,5]

Other differential diagnoses for IMF include ichthyosis vulgaris, large plaque parapsoriasis (LPP), and cutaneous pseudolymphoma. Ichthyosis vulgaris is an autosomal dominant disease that usually develops in patients between the ages of 3 months and 5 years with a positive family history.[7] Large plaque parapsoriasis is a clinical condition characterized by erythematous scaly patches or, very rarely, patchy ichthyotic lesions.[8] Approximately 10% to 35% of patients with LPP may progress to definite MF. Lesions of LPP show nonspecific spongiotic dermatitis or interface lymphocytic infiltrate. The typical histopathologic features of MF, such as epidermotropism and atypical lymphocytes, are absent in LPP.[8] Cutaneous pseudolymphomas are benign lymphoproliferative processes that can clinically and histologically resemble MF.[9] However, the absence of a mixed cellular infiltrate of $CD8^+$ T cells, $CD20^+$ B cells, and histiocytes helped us to rule out pseudolymphoma. And the presence of a clonal T-cell receptor γ gene rearrangement supported a diagnosis of MF in this case.

In general, patients with IMF have a good prognosis and respond well to nonaggressive therapies, such as topical treatment, 8-methoxypsoralen plus UV-A or narrowband UV-B therapy, or combined treatment with interferon α and low-dose methotrexate.[6,10]

ARTICLE INFORMATION

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