Hyperpigmented Mycosis Fungoides – A Rare Entity

Dear Editor,

A 67-year-old man of Kosovar-Albanian ethnic origin (skin phenotype IV) presented to our dermatology clinic with generalized hyperpigmented patches and plaques all over the body, so-called melanoerythroderma (Figure 1). The lesions, which first appeared nearly six years ago, developed gradually; they were diagnosed as mycosis fungoides (MF), and were subsequently treated only with topical corticosteroids. We performed further examinations upon admission to our department. Relevant laboratory parameters – blood cell count, LDH, urinalysis, and serum chemistry – were within normal limits. Endocrinological examination excluded Addison disease, and the patient was not receiving any drugs that could cause skin hyperpigmentation. Chest-abdomen-pelvis computed tomography (CT) scan and sternal puncture were normal. Flow cytometric immunophenotyping revealed less than 5% aberrant T-cells. Histopathology and immunohistochemistry of skin specimens revealed lichenoid infiltration of small- to medium-sized atypical T-lymphocytes within the upper dermis, epidermotropic lymphocytes with several Pautrier's microabscesses (Darier's nests), pigment incontinence, abundant melanophages in the papillary dermis (Figure 2, a, b), and the T-cell $CD4^{+}CD7^{-}CD8^{+}$ phenotype (Figure 2, c, d). Based on the clinical picture, histopathology, and immunohistochemistry the diagnosis of hyperpigmented mycosis fungoides (MF) stage IIIA (T4N0M0B0) was established. Skin-oriented therapy (retinoids-PUVA) resulted in slight improvement.

Hyperpigmented MF is a rare, uncommon, clinical variant of MF, with a predilection for dark-skinned people (1). Only a few cases of hyperpigmented MF have been reported so far, and our case being one of them (2-5). Hyperpigmented patches or/and plaques

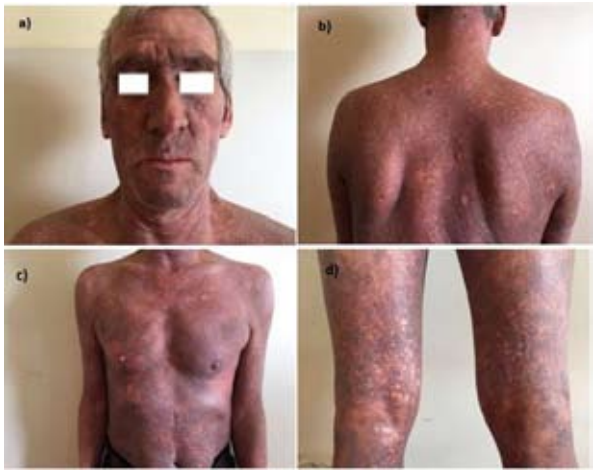

Figure 1. Clinical presentation of hyperpigmented mycosis fungoides – generalized hyperpigmented patches and plaques (melanoerythroderma).

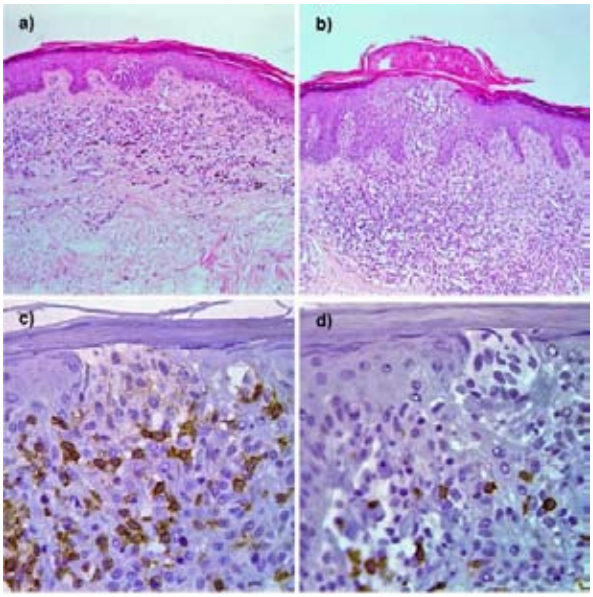

Figure 2. (a, b) Lichenoid infiltration of small- to medium-sized atypical T-lymphocytes within the upper dermis, epidermotropic lymphocytes with several Pautrier's microabscesses (Darier's nests), pigment incontinence, abundant melanophages in the papillary dermis (hematoxylin and eosin ×4). (c) Immunohistochemistry; staining for CD4 highlights the positive T-lymphocyte cells. (d) CD8+ (hematoxylin and eosin ×40).

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dominate the clinical picture. Hyperpigmented MF is characterized by a predominantly $CD8^+$ epidermotropic T-cell phenotype, although different phenotypes have been reported ($CD4^+$ or $CD4^-CD8^-$) (2). Histopathologically, interface changes, pigment incontinence and melanophages are usually found in addition to the classical findings of early MF (1). The exact mechanism of hyperpigmentation is not well understood. Hyperpigmented MF had an indolent course in most reported cases, and skin-directed therapy is therefore the treatment of choice. Although MF and its hyperpigmented variant is a lymphoma of low-grade malignancy, large-cell transformation ($CD30^+$) of hyperpigmented MF can occur (1).

These rare cases of special clinical MF variants are extremely valuable and can help us investigate and understand the pathophysiology of the disease. Treatment and close follow-up is mandatory in the hyperpigmented variant of MF.

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