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A Rare Presentation of Mycosis Fungoides Mimicking Psoriasis Vulgaris

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Key Words
Mycosis fungoides · Cutaneous lymphoma · Psoriasis

Abstract
Mycosis fungoides (MF) is an uncommon primary cutaneous lymphoma with a wide spectrum of clinicopathological manifestations. Diagnosis can be difficult in its early stages given the considerable overlap with more common benign dermatoses. We report an unusual case of MF in a 52-year-old male presenting with psoriasiform plaques on the palms and the soles who rapidly developed additional lesions on the scalp, limps and trunk. Punch biopsy of the face was obtained for routine histology and immunohistochemical stains. Chest X-ray, total body computed tomography scanning and excisional biopsy of the inguinal lymph node were performed. Review of the face biopsy revealed a diffuse dermal infiltrate containing a high number of atypical lymphocytes showing a CD3+, CD4+, CD45RO+, CD8–, CD20– immunophenotype and epidermotropism. Findings were consistent with tumor stage MF (stage IIB, T3 N1 M0). We report a rare presentation of MF mimicking psoriasis vulgaris.

Introduction

Mycosis fungoides (MF) is the commonest variant of primary cutaneous T cell lymphoma, accounting for almost 50% of all primary cutaneous lymphomas [1, 2]. It most commonly affects middle-aged and elderly adults of all races [3]. Typically, neoplastic T cells localize to the skin and produce patches, plaques, tumors or erythroderma [4]. It is characterized by a relatively consistent constellation of clinical, histologic, immunophenotypic and molecular aberrations [5].

MF typically manifests as an indolent cutaneous eruption with erythematous scaly patches or plaques and may progress to generalized erythroderma, cutaneous tumors, or extracutaneous involvement. The initial skin lesions are often confined to sunprotected areas. The rate of progression is unpredictable. Tumors, however, develop only in a minority of patients from patches, plaques, or de novo. They more commonly arise on the

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face and body folds. Leonine facies results from malignant T cell infiltration leading to extensive thickening and skin fold accentuation. The palms and soles can develop hyperkeratosis and skin fissuring. These phases may be distinct or overlapping at diagnosis [6, 7]. Classically, MF shows a CD2+, CD3+, CD4+, CD8–, CD30–, CD45RO+ immunophenotype. Rare cases with a T suppressor CD8+/CD4– phenotype have been reported [8, 9]. This report describes the clinical, histopathologic and immunohistochemical features of a patient with MF mimicking psoriasis vulgaris.

Case Report

A 52-year-old white man was referred to the Department of Dermatology, University of Palermo (Italy) with a six-month history of psoriasiform plaques with superficial erosion and fissuring on the palms (**fig. 1**b) and the soles. Within a few weeks red violet and infiltrated plaques with moderate hyperkeratosis and scaling developed on the scalp and upper limps accompanied by itching. The patient was misdiagnosed as having psoriasis vulgaris. Various mid- and high-potency topical corticosteroids did not offer significant relief. Four months prior to referral he developed red to violaceous nodules which coalesced to form large plaques on the face. Skin examination at the time of hospital admission, approximately six months from the onset of his illness, revealed reddish brown ulcerated tumors and indurated erythematous plaques with superficial erosion on the forehead, glabella, cheeks (fig. 1a), scalp and neck. In addition, numerous plaques, accompanied by itching, were found on trunk and extremities in a generalized distribution. Plaques appeared as sharply delineated, scaly, elevated lesions that were dusky red to violaceous and variably indurated (fig. 1c). His nails were intact without pitting or dystrophy. He had cervical, scalp, neck, axial and inguinal lymphadenopathy. His chest was clear on auscultation. There was no hepatosplenomegaly and his abdomen was soft with normal bowel sounds. His cardiovascular, musculoskeletal and neurological exams were normal. His past medical history was unremarkable. He was not on medications. There was no history of anorexia, weight loss, fever or night sweats. The blood count showed normal red cell count and a slight leukocytosis ($11.42 \times 10^3/\mu l$) comprised of 74.6% neutrophils, 7.2% eosinophils, 0.7% basophils, 11.1% lymphocytes and 6.4% monocytes. Further laboratory investigations of the blood showed alpha 1-globulin 5.25% and an erythrocyte sedimentation rate of 44 mm/h. Chemistry and liver function tests were unremarkable.

A biopsy specimen was obtained from affected areas of the face, fixed in 10% buffered formalin and embedded in paraffin. For routine histology, 5-μm-thick sections were stained with hematoxylin and eosin. Immunohistochemical stains were performed according to the markers presented in **table 1**. Chest X-ray, total body computed tomography scanning and excisional biopsy of the inguinal lymph node were obtained.

Results

Review of the face biopsy on hematoxylin and eosin showed a dense, partly subepidermal and band-like, partly perivascular and periadnexal dermal infiltrate (**fig. 2**a) containing a high number of atypical lymphocytes, eosinophils, plasma cells and macrophages. Many of the lymphocytes were pleomorphic, hyperchromatic, hyperconvoluted and had an increased cytoplasmic to nuclear ratio. Their nuclei were moderately irregular showing several mitoses (fig. 2b). Epidermotropism was prominent with small intraepidermal clusters of lymphocytes forming Pautrier's microabsesses. Mild acanthosis, hyperkeratosis, basal layer damage, edema and fibrosis of the papillary dermis and moderate proliferation of postcapillary venules were also seen. Immunohistologically the malignant cells expressed a mature peripheral T cell phenotype (CD3+, CD4+, CD45RO+, CD8–, CD20–) (fig. 2c–d). Chest X-ray and ultrasonography of the abdomen were without pathological findings. Peripheral blood film for Sézary cells was negative. Total body computed tomography scanning was obtained documenting retroauricular, occipital, deep cervical lymphatic chain, submandibular, submental, axillary, subclavian and inguinal enlarged lymph nodes without visceral involvement. An excisional biopsy of

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the inguinal lymph node showed dermatopathic and reactive lymphadenitis. Findings were consistent with tumor stage MF (stage IIB, T3 N1 M0).

Discussion

MF is generally an indolent malignancy, with slow progression over years or even decades [10]. The onset of the disease is often insidious and initial cutaneous symptoms may be difficult to distinguish from various inflammatory dermatoses such as chronic eczema, psoriasis, atopic dermatitis, etc. [8, 11]. As the disease progresses, patches may evolve over a variable period of time into infiltrated plaques with a more generalized distribution. Plaques can be followed by ulcerated and exophytic tumors. Tumors develop only in a minority of patients, although it is common to have patch, plaque and tumor lesions simultaneously on different parts of the body. Some patient with patch stage MF never progress to other forms of the disease. Tumors are the presenting sign in about 10% of cases. Lymphadenopathy is usually a late occurrence. Visceral dissemination (lungs, spleen, liver, gastrointestinal tract) may develop subsequently. Extracutaneous dissemination is directly correlated to the extent of cutaneous disease [7, 10].

We report a patient who first showed hyperkeratotic psoriasiform plaques on the palms and soles. Involvement of the palms and/or soles occurs at some time during the course of the disease in 11.5% of patients. Mycosis fungoides palmaris et plantaris is a rare variant of MF-type cutaneous T cell lymphoma predominantly affecting or initially presenting on the palms and/or soles. Sometimes the lesions may extend to the fingers, arms and feet. The prevalence of mycosis fungoides palmaris et plantaris among MF patients is 0.6%. In most cases the disease shows a benign course; the lesions remain confined to the initial areas without dissemination and extracutaneous spread [12, 13].

In our patient tumors in the skin and lymphadenopathy developed rapidly over a two-month time span without lengthy preceding erythematous or plaque stages. The initial lesions of MF usually develop in the fifth and sixth decades. The time interval between onset of skin changes and definite pathologic diagnosis of MF is 4–6 years [10, 14]. In the present case the lesions developed into a leonine clinical appearance of the face accompanied by lymphadenopathy, which reminded us the tumor stage MF. Clinical, histopathological and immunohistochemical findings revealed an atypical case of MF. Based on the early involvement of extracutaneous sites, the course of this patient may be more aggressive than typical patch-stage disease, in which mortality is not significantly different from matched controls. The presence of tumor stage disease is associated with a worse prognosis (stage IIB: median survival of 2.9 years) [15].

MF is an uncommon lymphoma with a wide spectrum of clinicopathological manifestations and unpredictable course. Diagnosis can be difficult in its early stages given the considerable overlap with more common benign dermatoses.

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Table 1. Summary of immunohistochemical methods and results

Antibody	Clone	Supplier	Dilution	Incubation time, min	Retrieval	Results
CD3	PC3/188A	Dako	prediluted	40	15 min	positive
CD4	Edu-2	NovoCastra	1:50	60	30 min	positive
CD8	C8/144B	Dako	prediluted	30	60 min	negative
CD20	L26	Dako	prediluted	35	60 min	negative
CD45RO	UCHL-1	Biogenex	1:75	30	50 min	positive

Fig. 1. Erythematous plaques with superficial erosion on the face (**a**), psoriasiform plaques with superficial erosion and fissuring on the palms (**b**) and sharply delineated plaques on trunk (**c**).

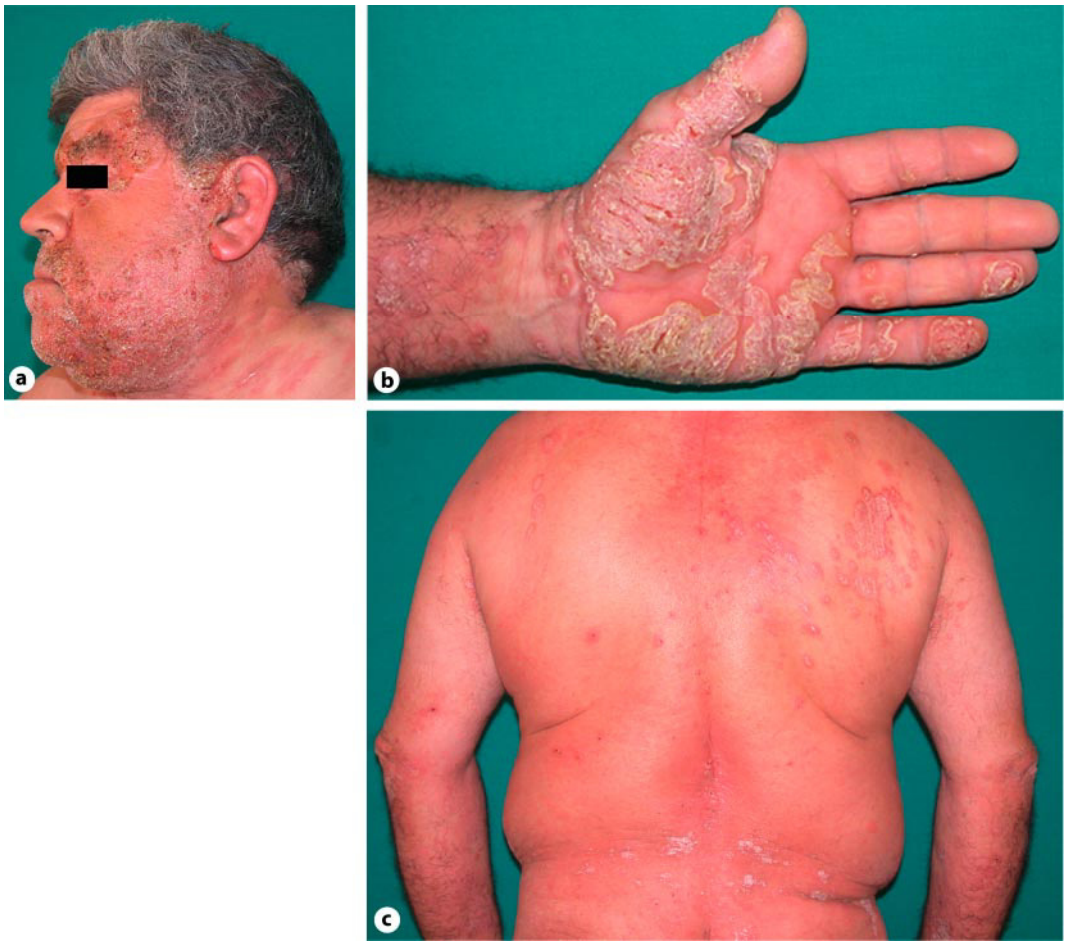

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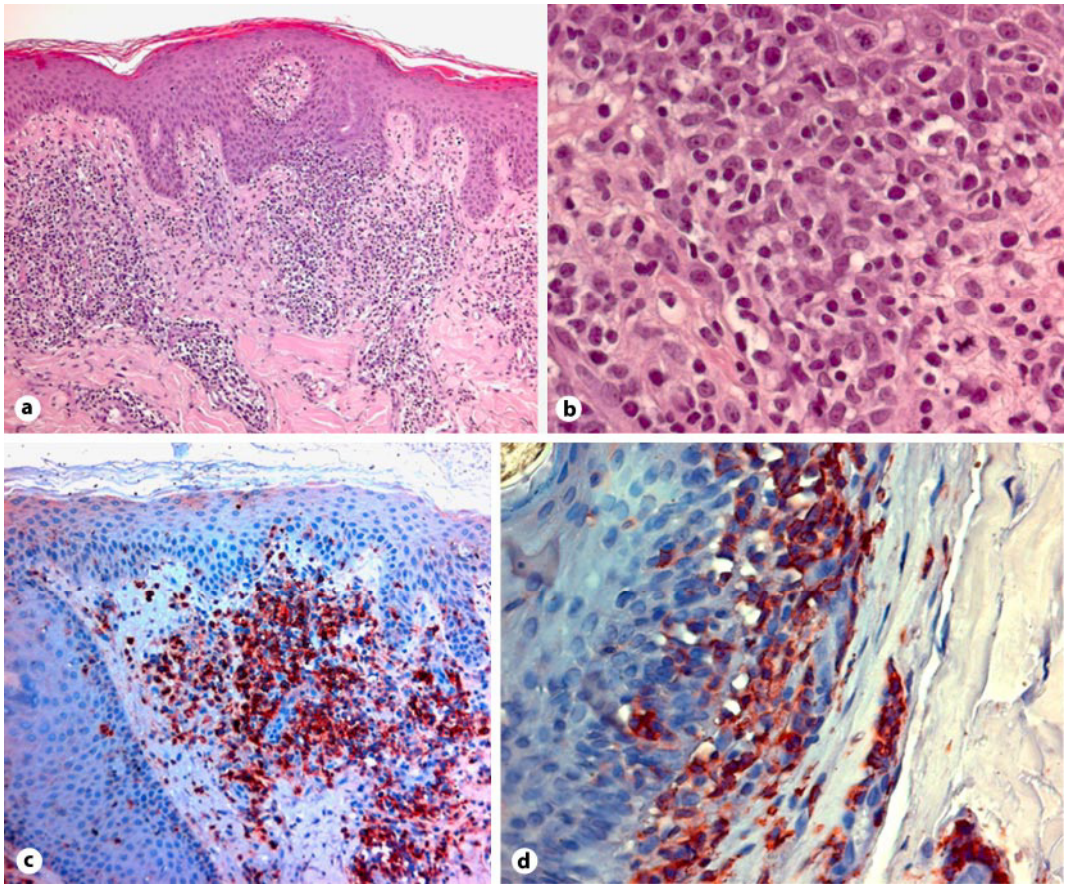
Fig. 2. **a** Histopathology of a skin biopsy showed a dense band-like dermal infiltrate (H&E, original magnification ×125). **b** Abnormal lymphocytes with moderately irregular nuclei showing several mitoses (H&E, original magnification ×640). **c** Immunoreactivity for CD45RO (original magnification ×160). **d** Immunoreactivity for CD45RO (original magnification ×500).



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