

Telangiectatic Mycosis Fungoides: A New Clinicopathological Presentation Mimicking Acquired Naevoid Telangiectasia

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Mycosis fungoides (MF) is the most common subtype of cutaneous T-cell lymphoma. The classical clinical picture usually evolves through 3 stages: patch, plaque and tumour stage. However, multiple clinicopathological variants of MF have been reported and a considerable number of reports have documented MF lesions mimicking different inflammatory and non-inflammatory cutaneous disorders. In these situations the diagnosis can be challenging and additional studies (histopathological, immunohistochemical and molecular analysis) may be necessary in order to establish a definite diagnosis.

We report here an uncommon clinical presentation of MF manifested by multiple isolated non-atrophic patches with prominent telangiectasias and, histopathologically, by an epidermotropic lymphocytic infiltrate accompanied by dilated capillaries in the reticular dermis. This “telangiectatic” variant of patch-stage MF seems to expand the clinicopathological spectrum of early MF lesions.

CASE REPORT

A 52-year-old man with personal history of arterial hypertension treated with perindopril was referred to our department for evaluation of a 6-month history of asymptomatic erythematous patches on the face, trunk and upper extremities. Physical examination revealed an apparently healthy man with several round-to-oval-shaped patches ranging from 2 to 15 cm in diameter on the abdomen (Fig. 1a), left arm (Fig. 1b) and left temple. The lesions were discrete erythematous patches with multiple, blanching, scattered, 1–3 mm telangiectasias diffusely distributed over the surface. Neither skin atrophy, nor hypo- or hyper-pigmented areas were present. The rest of the physical examination was unremarkable. The patient denied any alcohol consumption or previous treatments with oral calcium blockers or topical corticosteroids. Dermoscopy revealed abundant telangiectasias and

tortuous vessels associated with red-brownish patchy areas on the entire lesion (Fig. 1c).

Skin biopsies from 2 different patches were performed, which revealed an atypical band-like epidermotropic lymphocytic infiltration along the basal layer and upper dermis, surrounding prominent dilated vessels (Fig. 2a). Scattered intraepidermal atypical lymphocytes were observed without clear-cut Pautrier’s microabscess formation (Fig. 2b). No epidermal atrophy was present. Immunohistochemically, atypical lymphocytes were positive for CD3 and CD4 (Fig. 2c) and a decreased expression of CD7 antigen was noted. B-cell markers were all negative (CD20, CD10, CXCL13), as well as CD30 and c-kit (CD117). Ki67 labelling index was approximately 10%. A monoclonal lymphoid T-cell population was detected by PCR for both T-cell receptor (TCR) gamma and beta chains. Results of blood examination including blood cell count, liver function tests, lactate dehydrogenase and B2-microglobulin blood levels, as well as results of chest radiography and abdominal ultrasonography were unremarkable.

Treatment with potent topical corticosteroids was prescribed and a clinical improvement in the lesions was observed. During the following months 2 new telangiectatic patches developed on the abdomen and right armpit, of smaller size than the previous ones.

DISCUSSION

MF has a wide range of clinicopathological manifestations, some of which differ substantially from the classical presentation. Many different clinical variants of MF have been described including poikilodermic, follicular, bullous, granulomatous, hypopigmented, hyperpigmented, hyperkeratotic, pustular, ichthyosiform or erythrodermic forms (1–4). In addition, MF and, specifically, in early stages may adopt atypical clinical presentations and mimic a wide range of cutaneous disorders (5).

Our patient presented several non-atrophic patches with prominent linearly distributed telangiectasias

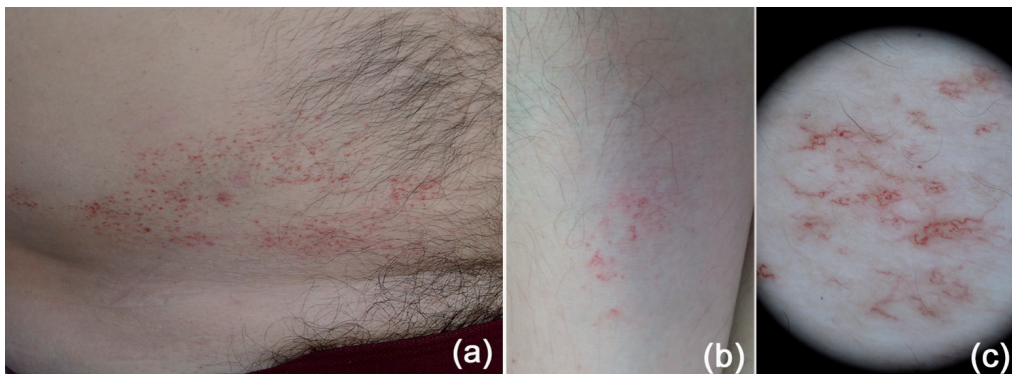


Fig. 1. Clinical presentation and dermoscopic features: erythematous asymptomatic patches with prominent telangiectatic vessels on (a) the abdomen and (b) the left arm. (c) Abundant telangiectasia and smaller tortuous vessels associated with red-brownish patchy areas at dermoscopy.

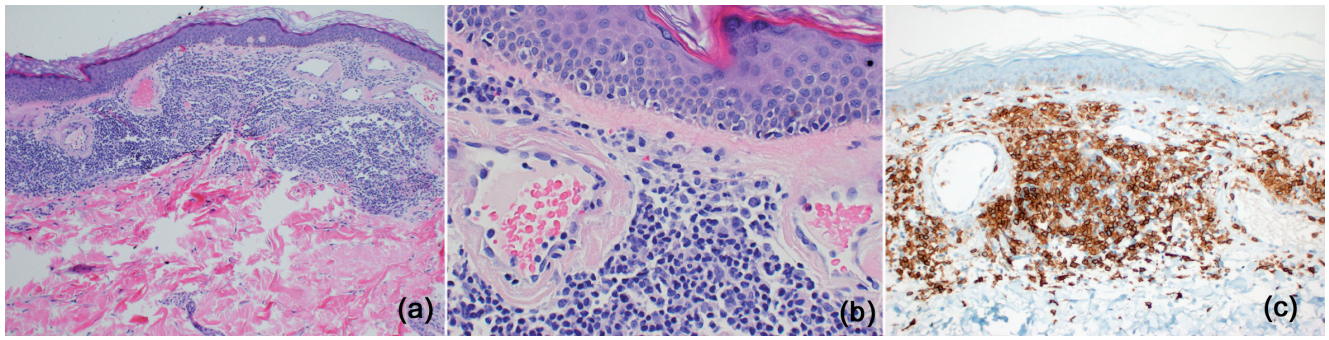


Fig. 2. Histopathological/Immunohistochemical features. (a) Band-like lymphocytic infiltrate along the basal layer and upper dermis surrounding ectatic blood vessels (hematoxylin and eosin, original magnification x100). (b) Epidermotropism of lymphoid cells (hematoxylin and eosin, original magnification x200) that (c) corresponded to mature T-helper cells expressing CD4 antigen (x100).

clinically mimicking acquired naevoid telangiectasia. Only a discrete underlying erythema was present and characteristic erythematous MF patches were not observed. Clinically, the differential diagnosis should be established with several disorders manifested by localized multiple clustered telangiectasias, including acquired (unilateral or bilateral) naevoid telangiectasia (6), localized telangiectasia macularis eruptiva perstans (7), reticular telangiectatic erythema secondary to subcutaneous implantation of medical devices (8) and collagenous vasculopathy. The association of photo-distributed telangiectasia with the use of calcium-channel blockers has also been reported (9). However, the presence of a band-like epidermotropic atypical dermal lymphoid infiltrate could easily rule out these diagnoses. The definite diagnosis of MF was established on the basis of the observed histopathological immunophenotypic and molecular genetic features.

Clinically non-apparent dilated superficial blood vessels seem to be a frequent feature of early MF lesions (10). In fact, fine short linear vessels in combination with orange-yellowish patchy areas seem to represent the most frequent dermoscopic pattern in early MF. Moreover, spermatozoa-like vessels might represent a rather specific finding of the disease. These features can help to distinguish early MF from other chronic dermatitis that frequently show dotted vessels, white and yellow scales, but hardly ever linear vessels (11). The possibility that an exaggerated neoangiogenic stimulus could explain the development of “telangiectatic MF lesions” can be hypothesized.

Prominent dilated vessels is a characteristic feature of poikilodermatous MF (poikiloderma vasculare atrophicans), a clinicopathological variant characterized by large plaques with areas of hypopigmentation and hyperpigmentation, atrophy, and telangiectasia typically involving the major flexural areas and trunk. Histologically, poikilodermatous MF shows an atrophic epidermis with dilated dermal blood vessels, fibrosis and melanophages in the upper dermis, along with scattered epidermotropic atypical lymphocytes around dermal blood vessels. The observed clinicopathological

features in our patient do not correspond to those of poikilodermatous MF; however, the possibility that may correspond to an early stage of this uncommon variant of MF cannot be ruled out.

In conclusion, we describe here a novel variant of MF clinically manifested by patches of grouped telangiectasias simulating acquired naevoid telangiectasia, which showed a good response to topical corticosteroids. This “telangiectatic variant” seems to both expand the clinical spectrum of MF and the list of dermatoses that could be mimicked by MF.

The authors declare no conflicts of interest.

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