POIKILODERMATOUS MYCOSIS FUNGOIDES: CLINICAL CASE

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Poikilodermatous mycosis fungoides (MF) is a rare distinct clinical variant of cutaneous T-cell lymphoma, formerly referred to as poikiloderma vasculare atrophicans or parapsoriasis variegata. Mycosis fungoides (MF) is a malignant neoplasm of T-lymphocyte origin, most commonly memory CD4+ T-cells [1–3].

Apart from classic form, there are poikilodermatous and erythrodermic variants (the latter should not be confused with Sézary's syndrome). There are wide range of rare atypical presentations including hypo- and hyperpigmented, verrucous, hyperkeratotic, follicular, lichenoid papular, palmoplantar psoriasiform, granulomatous, vesicular, bullous, and pustular variants, which have been described in the literature [1]. These are clinically unusual cases that run a similar course to that of classic MF. These are clinically unusual cases that run a similar course to that of classic MF were described as a complex dermatologic disease characterized by telangiectasia, pigmentation, and atrophy, were termed poikiloderma vasculare atrophicans (PVA). Later, it was believed that PVA represented a stage or an outcome of various dermatoses, such as mycosis fungoides, parapsoriasis, dermatomyositis, scleroderma, lupus erythematosus, lichen ruber planus, genodermatoses, and so on. Nowadays poikiloderma vasculare atrophicans is recognized as a clinical variant of patch stage MF [4–6]; and poikilodermatous findings on non-sun-exposed areas should be considered MF until proven otherwise.

The first manifestation of poikilodermatous MF usually occurs at an earlier age than that of classic MF, and a male predominance was reported for both forms.

Histopathology of poikilodermatous lesions discloses an atypical T-cell infiltrate in the papillary dermis, often with evident epidermotropism [1]. However, Pautrier microabscesses are not as common in comparison to classic MF. Melanophages and melanin incontinence are also observed, along with ectasia of the superficial dermal vessels and epidermal atrophy.

Immunohistological staining commonly shows either a prevalence of the CD4+, CD8− pattern or CD8+, CD4− immunophenotype, which is more often seen in hypopigmented variants of MF [7].

We report a 29-year-old patient with generalized poikilodermatous skin lesions, whose diagnosis of mycosis fungoides was made only a few years after the onset of his disease due to its bizarre clinical behavior and a natural reluctance to diagnose this disease in children and adolescents.

The first eruption appeared on his skin at the age of eleven. At that time there were some few separate well-defined asymptomatic hypopigmented patches on his chest and left shoulder, which resolved spontaneously without treatment. Several different diagnoses have been declared for this patient: vitiligo, morphea (after transformation into hyperpigmented plaques with slight atrophy), lichen ruber planus (two years earlier according to the histopathological findings, although immunohistological studies were not performed at that time). Lately the whole surface of the skin was very thin, crinkled and scaly, and had characteristic wrinkled, "cigarette-paper" appearance. The skin affections had diffuse distribution with only small several islands of uninvolved skin on the trunk and low extremities. Skin lesions presented a confluent poikilodermatous patches and plaques with mottled hyper- and hypopigmentation, atrophy and teleangiectases. Almost all of these patches, especially those located on the thighs and the lateral aspects of the trunk, were also remarkable for the net-like distributed plane lichenoid papules. On the anterior chest and in the paraumbilical area a few ill-defined erythematous patches could be observed. They were both visibly and palpably slightly infiltrated. The anterior aspects of the shins showed several confluent plaques with grayish-brown tint and evident infiltration.

Taking into consideration the aforementioned features and the past medical history data we were inclined to regard the condition as a rare poikilodermatous form of MF. It was decided to obtain four punch biopsy specimens from representative areas: erythematous patch on the anterior aspect of the chest, typical poikilodermatous patch on the right flank, lichenoid papule on the right thigh and the plaque on the anterior aspect of the left shin respectively. The diagnosis «Poikilodermatous MF» was confirmed by histopathological examination. The immunohistologic studies of all the specimens also revealed the unusual pattern with simultaneous presence of both CD4+ and CD8+. Suspecting misdiagnosing it was decided to reassess the biopsy findings which were received two years earlier with additional sectioning of preserved paraffin blocks. An appraisal of both slides was made. While the first one made three years ago showed the histopathological features of

lichen planus, the second one received by additional sectioning revealed the signs of MF, though they were not apparent.

Conclusions: The variability of atypical clinical presentations of MF and its similarity to the benign inflammatory and noninflammatory skin disorders may become a source of considerable confusion and controversy, challenging a dermatologist to make a nonprecise diagnosis. Multiple biopsies with additional block sectioning and immunohistochemistry may appear essential to reach the genuine diagnosis. Therefore, scrupulous clinicopathological correlation is the absolute necessity.

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