Pityriasis Lichenoid-like Mycosis Fungoides in a 9-year-old Boy: A Case Report

Dear Editors,

Pityriasis lichenoides (PL)-like mycosis fungoides (MF) is a rare variant of MF, presenting clinical findings of PL but histological features of MF. It was first reported by Ko et al. (1) and only a few cases have been reported since (2-5). Herein we report the case of a boy with PL-like MF and review the related literature.

A 9-year-old boy presented with a 1-year history of multiple pruritic crusted erythematous papules and scaly pink maculopatches on the face, trunk, and extremities (Figure 1, a and b). Histologic examination of a papule revealed lymphocytic epidermotropism and lymphocytes tagging the dermoepidermal junction. The nuclei of the lymphocytes were hyperchromatic and irregular (Figure 1, c and d). Immunohistochemically, the infiltrating lymphocytes revealed positivity for CD2, CD3, CD5, CD7, and CD8, but were negative for CD4, CD20, CD30, CD68, and CD163 (Figure 1, e-g). T-cell receptor gene rearrangement analysis (TCR-GRA) demonstrated the rearrangement of the

Figure 1. (a) Generalized crusted erythematous papules and white to pink scaly maculopatches on the trunk and upper limbs. (b) Close-up view of the brownish to erythematous crusted scaly papules and hypopigmented maculopatches on the chest. (c) Epidermotropism of atypical lymphocytes with perinuclear halo and lymphocytes tagging the dermoepidermal junction (hematoxylin and eosin (HE), ×200). (d) Atypical lymphocytes with hyperchromatic irregular, convoluted, or cerebriform nuclei (HE, × 400). Immunohistochemical stain (original magnification ×200): (e) positive for CD3, (f) negative for CD4, and (g) positive for CD8. (h) T-cell receptor gene rearrangement analysis demonstrating rearrangement of the gamma chain.
There has long been a controversy regarding the relationship between PL and MF: (A) PL with a dominant T-cell clone, (B) PL subsequently progressing into MF, and (C) PL-like MF. In the first category, PL is a monoclonal T-cell lymphoproliferative disease. Wang et al. (2) proposed three categories for the relationship between PL and MF: (A) PL with a dominant T-cell clone, (B) PL subsequently progressing into MF, and (C) PL-like MF. In the first category, PL is a monoclonal T-cell-mediated inflammatory disorder, in which T-cell clones were found in about 50% of patients.
The second category involves progression from long-term PL to MF (8,9). The average time-to-progression is about 8 years. It has been speculated that the PL-related immunologic microenvironment is favorable for developing a tumoral clone. Our patient presented with PL-like lesions clinically, while biopsy findings, results of immunohistochemistry, and TCR-GRA all suggested that this case was MF. Due to the short duration (only one year) of his lesions, we established the diagnosis of PL-like MF de novo, rather than evolution from PL to MF.

The features of previously reported cases of PL-like MF and those of our patient are summarized in Table 1 (1-5). Men were predominant (18:7) among the total of 25 patients. Most patients were children or young adults (mean age of 23.4 years). The interval between presence of lesions and diagnosis varied from 1 month to 10 years. The cutaneous eruptions were all PL in appearance and almost all involved both the trunk and extremities. Pruritus was reported by approximately half of the patients. Histologically, the scaly papules were usually indistinguishable from classical MF, showing epidermotropism, haloed lymphocytes, lymphocytes aligning along the dermoeidermal junction, and Pautrier's microabscesses. Immunohistochemically, all tested cases demonstrated positivity for CD3 but were negative for CD20 and CD30. Cases with predominantly CD8-positive cells were twice as prevalent as cases with predominantly CD4-positive cells. TCR-GRA was performed in 20 cases, 15 of which revealed monoclonality. Most patients received psoralen combined with ultraviolet A or NBUVB phototherapy, and demonstrated either a complete or partial response. Recurrence was reported in only 2 cases (5).

In summary, PL-like MF is a rare variant of MF. It has some features distinct from classic MF, such as a higher incidence in young men and predominantly CD8-positive T-cells infiltration. Phototherapy can be used as the first line of treatment. A good response and a favorable prognosis can be expected.

References:

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