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

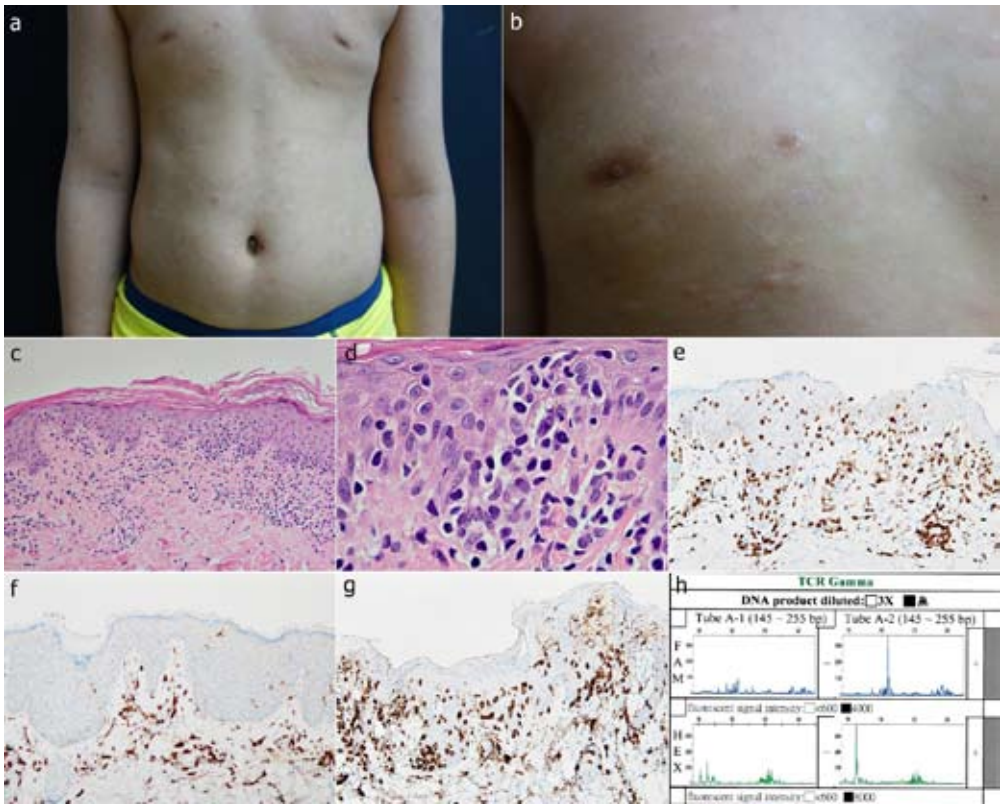

Figure 1. (a) Generalized crusted erythematous papules and white to pink scaly maculopatches on the trunk and upper limbs. (b) Close-up view of the brownish to erythematous crusted scaly papules and hypopigmented maculopatches on the chest. (c) Epidermotropism of atypical lymphocytes with perinuclear 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.

Table 1. Clinical and pathologic features of previously reported 24 pityriasis lichenoid-like mycosis fungoides patients and our patient Patient no./gender/age of diagnosis

	Location	Clinical presentation	Interval[a] (month)	IHC stain	TCR-GRA	Treatment & response
1[1]/M/10	T/E	M/Pa/S	12	CD3 (+), CD20 (-), CD30 (-)	γ chain	PUVA; CR
2[1]/M/5	T/E	M/Pa/PA/S	12	CD3 (+), CD20 (-), CD30 (-)	γ chain	PUVA; Response NA
3[1]/M/4	T/E	Pa/S	24	CD3 (+), CD20 (-), CD30 (-)	γ chain	Loss follow-up
4[2]/F/60	T	Pa/S	6	CD3 (+), CD4 (+), CD8 (-), CD20 (-)	γ chain	Topical steroid; CR
5[3]/F/32	T/E	M/Pa/S	120	CD3 (+), CD4 (-), CD8 (+)	γ chain	NA
6[4]/M/52	T/E	Pa	60	CD3 (+), CD4 (+), CD8 (-), CD20 (-), CD30 (-)	Polyclonal	Multi-therapy[b]; Persistence
7[4]/F/70	T/E	Pa	60	CD3 (+), CD4 (+), CD8 (-), CD20 (-), CD30 (-)	Polyclonal	Multi-therapy[c]; Persistence
8[4]/F/49	T/E	Pa	24	CD3 (+), CD4 (+), CD8 (-), CD20 (-), CD30 (-)	Polyclonal	Multi-therapy[d]; Persistence
9[4]/M/35	T/pubic	Pa	12	CD3 (+), CD4 (+), CD8 (-), CD20 (-), CD30 (-)	Monoclonal	PUVA; CR
10[5]/M/4	T/E	Pa/S	12	CD3 (+), CD4<CD8, CD30 (-)	Monoclonal	PUVA; CR
11[5]/M/7	T/E	M/Pa	6	CD3 (+), CD4<CD8, CD30 (-)	Monoclonal	PUVA; CR
12[5]/M/8	T/E	M/Pa	24	CD3 (+), CD4<CD8, CD30 (-)	Monoclonal	PUVA; CR
13[5]/M/7	T/E	Pa/S	1	CD3 (+), CD4<CD8, CD30 (-)	NA	No treatment
14[5]/M/10	T/E	M/Pa	36	CD3 (+), CD4<CD8, CD30 (-)	Polyclonal	NBUVB; CR
15[5]/M/19	G	Pa/S	14	CD3 (+), CD4<CD8, CD30 (-)	Monoclonal	NBUVB; CR
16[5]/M/44	T/E	Pa/PA	1	CD3 (+), CD4>CD8, CD30 (-)	Monoclonal	NBUVB; CR
17[5]M/10	T/E	M/Pa	5	CD3 (+), CD4>CD8, CD30 (-)	Monoclonal	NBUVB; CR
18[5]/M/59	T/E	M/Pa	60	CD3 (+), CD4<CD8, CD30 (-)	NA	No treatment
19[5]/M/5	G	M/Pa	5	CD3 (+), CD4>CD8, CD30 (-)	Monoclonal	NBUVB; CR; Recurrence
20[5]/F/25	T/E	Pa/PA	2	CD3 (+), CD4<CD8, CD30 (-)	Polyclonal	NBUVB; CR
21[5]/F/7	T/E	M/Pa	1	CD3 (+), CD4<CD8, CD30 (-)	Monoclonal	NBUVB; CR
22[5]/M/11	E	Pa/Pl	20	CD4>CD8, CD30 (-)	NA	NBUVB; CR
23[5]/M/18	T/E	Pa	48	CD4<CD8	NA	NBUVB; CR
24[5]/F/24	T/E	Pa/PA	12	NA	NA	NBUVB; CR; Recurrence
25/M/9 (present case)	G	Pa/S	12	CD3 (+), CD4 (-), CD8 (+), CD20 (-), CD30 (-)	γ chain	NBUVB; CR

Summary

- Total of 25 cases; M/F 18/7; mean age 23.4 years of age (range 4-70 years)
- Location: the whole body (3), trunk & extremities (19), trunk only (2), extremities only (1)
- Symptoms (NA in 5 cases): pruritic/asymptomatic 12/8
- Mean interval between presence of lesions and diagnosis: 23.5 months (range 1-120 months)
- Histological findings: epidermotropism 100% (25/25), haloed lymphocytes 68% (17/25), tagging of lymphocytes along the dermoepidermal junction 64% (16/25), coarse collagen bundles in the dermal papillae 56% (14/25), Pautrier's microabscesses 36% (9/25)
- IHC stain for CD4 & CD8 (NA in 4 case): CD8>CD4 in 57% (12/21), CD4>CD8 in 43% (9/21)
- TCR-GRA (NA in 5 cases): monoclonal 75% (15/20), polyclonal 25% (5/20)
- Treatment: phototherapy in 20 cases, with CR 80% (16/20), poor response 15% (3/20), and response unavailable 5% (1/20)

T: trunk; E: extremities; G: generalized; M: macules; Pa: papules; PA: patch; Pl: plaque; S: scales; IHC stain: immunohistochemical stain; TCR-GRA: T-cell receptor gene rearrangement analysis; NA: not available; CR: complete remission; PUVA: psoralen combined with ultraviolet A; NBUVB: narrow band ultraviolet B

[a]Interval indicates the interval between presence of lesions and being diagnosed with mycosis fungoides

[b]Topical steroids, erythromycin, PUVA

[c]Topical steroids, total skin electron irradiation, PUVA, NBUVB, Bexarotene

[d]Topical steroids, PUVA, interferon

gamma chain (Figure 1, h). PL-like MF was diagnosed. The patient was started on narrowband ultraviolet B (NBUVB) phototherapy. The skin lesions markedly improved after 6 months of treatment. There was no recurrence during the 2 years of follow-up.

There has long been a controversy regarding whether PL is just an inflammatory dermatosis or a genuine 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

(6,7). 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 dermoepidermal 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.

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Teng-Li Lin[1], Yi-Ju Chen[1,2], Yu-Ching Weng[3], Chi-Shun Yang[4], Chao-Kuei Juan[1,2]

[1]Department of Dermatology, Taichung Veterans General Hospital, Taichung, Taiwan

[2]Department of Dermatology, National Yang-Ming University, Taipei, Taiwan

[3]Department of Dermatology, Puli Branch, Taichung Veterans General Hospital, Taichung, Taiwan

[4]Department of Pathology, Taichung Veterans General Hospital, Taichung, Taiwan

Corresponding author:

Chao-Kuei Juan, MD

Department of Dermatology, Taichung Veterans General Hospital

Taichung
Taiwan

No. 1650, Section 4, Taiwan Boulevard

Taichung 40705
Taiwan

zon2zon2zon2zon2@yahoo.com.tw

Received: September 19, 2018

Accepted: February 11, 2019