SHORT COMMUNICATION

Dorsal Hand and Foot Mycosis Fungoides: Looking Beyond Mycosis Fungoides Palmaris et Plantaris

Kee Suck SUH, Jang Hwan JUNG, Sun Mun JEONG, Min Soo JANG and Jong Bin PARK*
*Department of Dermatology, Kosin University College of Medicine, 262 Gamcheon-ro, Seo-gu, Busan, 49267, South Korea. *E-mail: dermashining@gmail.com*
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Mycosis fungoides palmaris et plantaris (MFPP) refers to a variant of mycosis fungoides (MF) that is localized on the palms and soles (1). In 2013, Nakai et al. (2) proposed a new concept, hand and foot MF (HFMF), to encompass all cases involving the hand and foot, including not only the palmar, but also the dorsal sides. However, there has been no study focusing on HFMF showing apparent predominance of the lesions on the dorsal hands over the palmar side, which we have termed dorsal HFMF (DHFMF).

MATERIALS AND METHODS

A retrospective study was conducted on 6 patients with DHFMF to investigate clinicopathological findings. Inclusion criteria were: (*i*) diagnosis of MF, based on the diagnostic algorithm for early-stage MF (3) via a combination of clinical, histopathological, and T-cell receptor (TCR) γ gene rearrangement analyses, (*ii*) MF lesions confined to the hands and feet without involving any other body parts, and (*iii*) the dorsal side of the hands being the most prominent site of involvement. Patients who did not show monoclonality in TCR γ gene rearrangement analysis were excluded. Clinical data were collected through review of medical charts and clinical images, and a detailed history was taken from 5 available patients to identify possible causative factors. Histopathological findings were evaluated by reviewing the slides for each patient.

RESULTS AND DISCUSSION

Histopathological findings are summarized in **Table I**. Typical features of MF were seen, along with frequent orthohyperkeratosis, parakeratosis, and acanthosis (**Fig. 1**).

Table I. Histopathological findings in 6 cases of dorsal hand and foot mycosis fungoides

Criteria	Patient number 1	2	3	4	5	6	Cases *n* (%)
Epidermal features							
Orthohyperkeratosis	+	+	–	+	+	+	5 (83.3)
Parakeratosis	+	–	+	+	+	+	5 (83.3)
Acanthosis	+	+	+	+	+	+	6 (100.0)
Epidermotropism	+	+	+	+	+	+	6 (100.0)
Pautrier's microabscess	+	–	–	+	+	+	4 (66.7)
Atypical lymphocytes with hyperchromatic nuclei	+	+	+	+	+	+	6 (100.0)
Haloed lymphocytes	+	+	+	+	+	+	6 (100.0)
Lymphocytes in the epidermis larger than those in the dermis	–	–	+	+	+	–	3 (50.0)
Lymphocytes aligned along the basal layer	–	+	–	+	+	+	4 (66.7)
Spongiosis	–	+	+	–	–	–	2 (33.3)
Dermal features							
Lichenoid infiltration	+	+	–	+	–	+	4 (66.7)
Dermal papillae stuffed with lymphocytes	+	+	–	+	+	+	5 (83.3)
Coarse collagen bundles in the papillary dermis	+	+	–	+	+	+	5 (83.3)

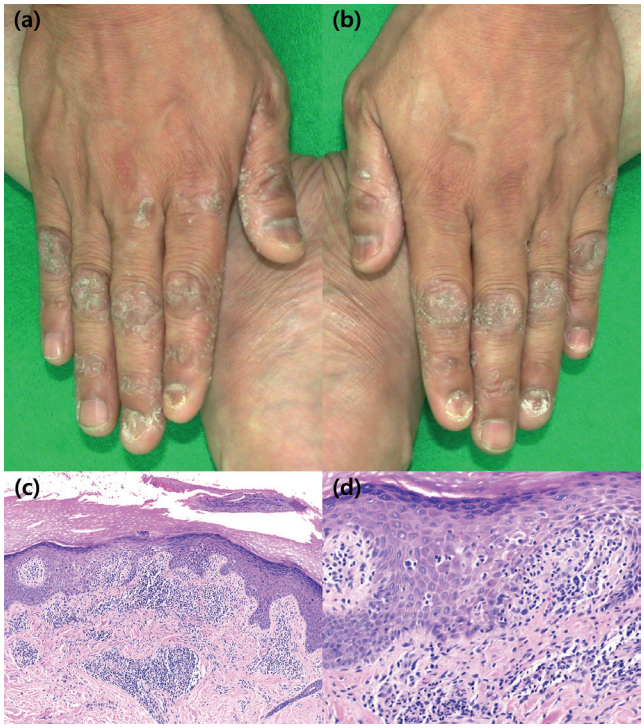

Fig. 1. Clinical and histopathological findings in dorsal hand and foot mycosis fungoides. (a, b) Hyperkeratotic plaques are predominantly distributed on the knuckles (interphalangeal and metacarpophalangeal joints) in patient 1. (c, d) Histopathological findings in patient 4. (c) Patchy lichenoid infiltration, marked orthohyperkeratosis, parakeratosis, and epidermotropism (haematoxylin and eosin stain (H&E) ×100). (d) Large atypical haloed lymphocytes in the spinous layer along with basal alignment of lymphocytes (H&E ×400).

Occasional spongiosis were noted, consistent with the known findings in MFPP (4). Because these features may confound the diagnosis of DHFMF, immunohistochemical and molecular biological studies should be performed in combination. All 6 patients showed a predominance of $CD4^+$ over $CD8^+$ T cells.

Clinical data are shown in **Table II**. The mean duration from onset to diagnosis was 1.8 years. All lesions presented as localized pruritic patches or plaques that were hyperkeratotic or scaly, corresponding to the typical morphology of MFPP (Fig. 1) (4, 5). The lesions were predominantly located on the metacarpophalangeal or interphalangeal joints of the dorsal hands (**Fig. 2**). These findings show that DHFMF cannot be clinically differentiated from benign dermatoses, such as psoriasis, hand and foot eczema, and dermatomyositis, without histopathological confirmation.

Table II. Clinical data in 6 cases of dorsal hand and foot mycosis fungoides

Pat No.	Age, years/ sex	Duration (years)	Distribution	Symptom	Stage[a]	Occupation	Exposure duration (years)	Treatment and response	Outcome	Follow-up duration (months)
1	51/M	1	D+P+S	Pruritus	IA	Not known	–	MTX (4 mo) → PR	Responded well but lesions remained	4
2	53/M	2	D+P	Pruritus	IA	Fisherman	20	MTX (3 mo) → CR	Cleared up	3
3	39/M	2	D+P+S	Pruritus	IA	Store worker (transporting goods)	10	UVA-1 (6 mo) → PR	Responded well but lesions remained	6
4	52/F	3	D+P	Pruritus	IA	Cook (trimming and grilling)	18	UVA-1 (8 mo) → CR → Recur	Cleared up but recurred 2 years later	58
5	55/M	1	D+P+S	Pruritus	IA	Steel industry worker	16	UVA-1 (1 mo) → CR	Cleared up	1
6	63/M	2	D	Pruritus	IA	Farmer	21	Ali (1 mo) → CR	Cleared up	6

[a]Clinical staging was performed according to the International Society for Cutaneous Lymphoma and the European Organisation for Research and Treatment of Cancer classification for cutaneous lymphomas (2011).
Ali: oral alitretinoin; M: male; F: female; CR: complete remission (> 95% clinical improvement); mo: months; D: dorsal side of the hands; MTX: oral methotrexate; P: palmar side of the hands; PR: partial remission (50–95% clinical improvement); S: soles; UVA-1: ultraviolet A1.

All 5 patients for whom detailed history taking was available had occupations involving a lot of manual work for quite a long time (mean 14.2 years) until first onset of the disease. The lesions were located mainly on the protruding parts of the hands (knuckles), which are susceptible to external impacts. Therefore, a possible relationship between trauma and DHFMF could be inferred, as Paul et al. (6) suggested that chronic trauma may be a factor in antigenic stimulation responsible for causing MF, and Lebas et al. (7) claimed that MF might exhibit the Koebner phenomenon. Meanwhile, 4 patients had been repeatedly exposed to chemicals, such as motor fuels (patient 2), detergents and byproducts of grilling (patient 4), metals, cutting oils, and solvents (patient 5), and pesticides (patient 6). These might also have served as causative factors, since previous studies have suggested relationships with various chemicals (8–10). These studies have shown that at least several years or sometimes decades of exposure are required to develop malignant transformation following chronic antigenic stimulations, differing from inflammatory dermatoses (e.g., contact dermatitis) that develop via various immunologic mechanisms (e.g., hypersensitivity reaction) within a relatively short time after exposure to causative agents. Further studies are required to clarify the exact nature of the antigens involved and to what extent they play a role in the development of MF.

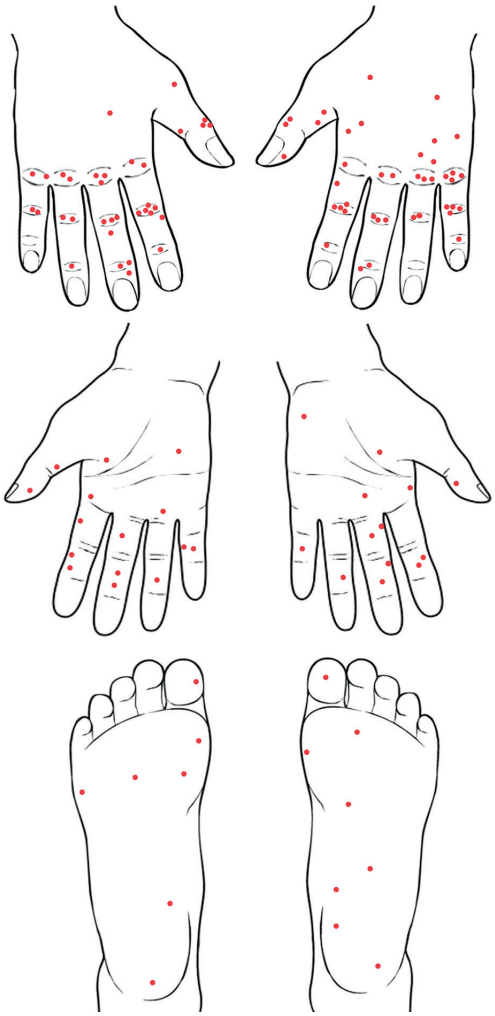

Fig. 2. Distribution of lesions in dorsal hand and foot mycosis fungoides. The distribution of the lesions in 6 patients is shown simultaneously. Each *red dot* represents 1 lesion. Note the knuckle-tropic distribution with the interphalangeal and metacarpophalangeal joints of the dorsal hand are the most prominent site of involvement.

Similar to MFPP having a relatively indolent course (2), all patients with DHFMF were well-controlled. Four patients achieved complete remission, and 2 showed partial remission. Local recurrence occurred in 1 patient 2 years after achieving complete remission. Patients responded well to ultraviolet A1 irradiation (65 J/cm^2, 3 times per week), oral methotrexate (15 mg/week), and oral alitretinoin (30 mg/day), previously known to be effective in the treatment of MFPP (4, 11).

The hyperkeratotic morphology, occasional spongiosis on histopathology, and favourable response to treatments indicate overall clinicopathological similarities between DHFMF and MFPP (2, 4, 5). Therefore, it is plausible that they should be encompassed under a single comprehensive concept; HFMF. Since DHFMF may be overlooked due to its overlapping features with benign dermatoses, combined analysis of histopathological, immunohistochemical, and molecular biological studies should be performed, based on a high index of suspicion when faced with long-standing localized hyperkeratotic knuckle-tropic lesions on the hand and foot.

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The authors have no conflicts of interest to declare.

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