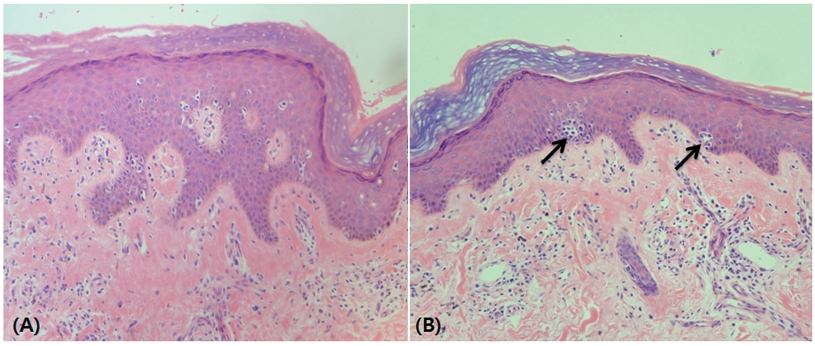

Fig. S1. (A) Biopsy specimens showing scattering of atypical lymphocytes, some forming small collections in hyperplastic epidermis. (B) Histopathological findings show compact orthokeratosis with underlying thinned granular layer and Pautrier's microabscesses (arrow) (haematoxylin and eosin (H&E) ×100) (patient 6).

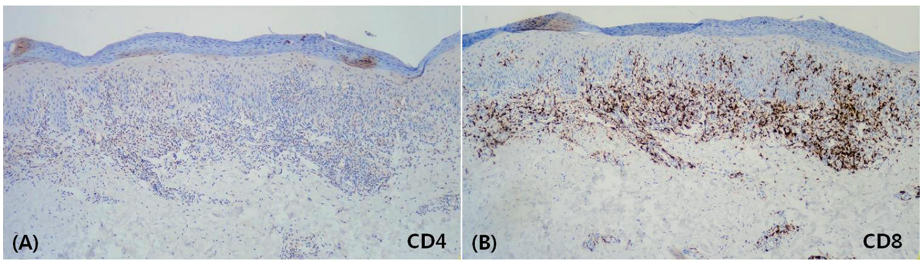

Fig. S2. Predominance of a $CD8^{+}CD4^{-}$ phenotype detected on immunohistochemistry (A, B) (patient 9).

Appendix S1

MATERIALS AND METHODS

Patients

Medical records of patients with MF seen in the dermatology department of Kosin University Gospel Hospital, South Korea, over the last 22 years were searched for patients with ichthyosis-like eruptions. Diagnosis of MF was based on a combination of clinical, histopathological/immunohistochemical and T-cell receptor (TCR)-γ gene analysis data. Data on patient's age, sex, duration of disease, location and form of skin lesion, symptoms, clinical diagnosis, medical history, therapy, response to treatment, outcomes and course of the disease were collected. Stage was determined by tumor–node–metastasis (TNM) classification, revised in 2011 by a joint working group of the International Society for Cutaneous Lymphomas (ISCL), the US Cutaneous Lymphoma Consortium and the Cutaneous Lymphoma Task Force of the European Organisation for Research and Treatment of Cancer (EORTC) (2).

Histological, immunohistochemical and molecular evaluation

Histopathological findings were assessed based on the findings of early MF suggested by Pimpinelli et al. (3).

An avidin–biotin complex immunoperoxidase technique (ABC Vectastain Kit; Vector Laboratories, Burlingame, CA, USA) was applied to paraffin-embedded tissue using monoclonal antibodies to CD3 and CD7. To further distinguish T-helper cells from T-suppressor cells, antibodies to CD4 and CD8 were applied and the CD4/CD8 ratio was calculated. Polymerase chain reaction was used to determine the TCR gene rearrangement.

Table SIII. *Histopathologic, immunophenotypic, and genotypic findings of 10 patients with ichthyosiform mycosis fungoides (IMF)*

Patient	1	2	3	4	5	6	7	8	9	10	Patients (n = 10) *n* (%)
Histological features of conventional mycosis fungoides											
Parakeratosis	–	+	+	+	–	+	–	–	+	–	5 (50)
Pautrier's microabscess	–	–	+	+	–	+	–	+	+	–	5 (50)
Lymphocytes aligned along basal layer	+	+	–	–	–	+	+	+	+	–	6 (60)
Epidermotropism	+	+	+	+	+	+	+	+	+	+	10 (100)
Intraepidermal lymphocytes larger than lymphocytes in dermis	+	–	+	–	–	+	+	–	–	–	4 (40)
"Haloed" lymphocytes in the epidermis	+	+	+	+	+	+	+	+	+	+	10 (100)
Atypical cells with hyperchromatic nuclei	+	–	+	+	–	+	+	+	+	+	8 (80)
Psoriasiform lichenoid pattern	+	+	+	+	–	+	+	+	+	–	8 (80)
Dermal papilla was stuffed with lymphocytes	–	–	–	+	–	+	–	+	+	–	4 (40)
Coarse collagen bundles in the papillary dermis	+	+	+	+	+	+	+	+	+	+	10 (100)
Folliculotropism	–	–	–	–	–	–	–	+	–	–	1 (10)
Histological features of acquired ichthyosis											
Compact orthokeratosis	+	+	+	+	+	+	+	+	+	+	10 (100)
Thinning or absent granular layer	+	+	+	+	+	+	+	+	+	+	10 (100)
Histological features of vasculitis (fibrinoid necrosis)	–	–	–	–	+	–	–	–	–	–	1 (10)
Immunophenotype[a]											
CD3	+	+/–	+	+	+	+/–	+/–	+	++	++	
CD4/CD8 ratio	=1	>1	=1	>1	<1	=1	=1	>1	1<	1>	
CD7	+/–	–	–	+/–	+	–	–	–	–	+/–	
T-cell receptor-γ gene rearrangement	+	–	+	+	+	–	–	+	–	+	6 (60)

[a]–: Less than 10% of cells; +/−: 10–30% of cells; +: 30–50% of cells; ++: 50–70%; +++: ≥ 80% of cells.

CD4/CD8 ratio < 1, CD8 expression is more predominant than CD4; =1, expressions of CD4 and CD8 are similar; >1, CD4 expression is more predominant than CD8.

Table SII. *Clinical outcomes, treatment, follow-up and course of patients with ichthyosiform mycosis fungoides*

Patient	Treatment and response	Follow-up duration after diagnosis, years	Course
1	PUVA → PR, 6 years later relapse, NBUVB → CR	18	No new lesions appeared
2	PUVA → PR	8.2	Responded to treatment but the lesions remained
3	PUVA, Aci → PR	10.8	Responded well after therapy, but lesions remained
4	PUVA, Aci → PR, MTX (for 31 months), Ste → PR	3	Responded to treatment but the lesions remained
5	PUVA, Aci → PR, MTX→ PR, UVA1, Ste → PR	8	Responded to treatment but the lesions remained
6	PUVA, Aci→ PR, Aci → PR	5	Responded to treatment but the lesions remained
7	PUVA → PR	1	Repigmentation occurred after PUVA, but the lesions remained
8	NBUVB → no response, PUVA, Aci → PR, PCALCL → Excision, IFN-α → PR	5	Not responded well after NBUVB, but combined treatment with PUVA and Aci showed partial remission Follicular lesions and newly emerging nodules also appeared
9	PUVA → PR	1	Responded well after therapy, but lesions remained
10	NBUVB → PR	0.5	Responded well after therapy, but lesions remained

PUVA: psoralen plus ultraviolet A; NBUVB: narrowband ultraviolet B; UVA1: ultraviolet A1; MTX: methotrexate; Aci: oral acitretin; ste: topical steroid; CR: complete remission; PCALCL: primary cutaneous anaplastic large cell lymphoma; PR: partial remission; IFN-α: interferon α.

Table SI. *Clinical data for patients with ichthyosiform mycosis fungoides (IMF)*

Patient	Age, years/ Sex	Medical history	Duration, years, before MF	Symptom	Clinical diagnosis	Age at onset of MF, years	Age at onset of ichthyosiform eruptions, years	Location of ichthyosiform lesions	Additional MF lesions	Associated lymphoma	TNM/stage
1	15/M		2	Pruritic (VAS=3)	Psoriasis, Parapsoriasis, MF	13	13	Trunk, buttocks, upper and lower limbs	Patch	–	$T_2N_0M_0$/IB
2	8/F		5	Pruritic (VAS=4)	Ichthyosis vulgaris	3	3	Scalp, face, trunk, buttocks, upper and lower limbs	None	–	$T_2N_0M_0$/IB
3	17/F		10	Asymptomatic	Ichthyosis vulgaris	7	7	Trunk, buttocks, upper and lower limbs	None	–	$T_2N_0M_0$/IB
4	63/M	DM, Fatty liver	2	Pruritic (VAS=8)	Ichthyosis vulgaris, exfoliative dermatitis	61	61	Trunk, buttocks, upper and lower limbs	Plaques	–	$T_2N_0M_0$/IB
5	30/M	BPH	5	Asymptomatic	MF	25	25	Trunk, buttocks, upper and lower limbs	Multiple ulcerated lesions	–	$T_2N_0M_0$/IB
6	47/M	Tenosynovitis	3	Pruritic (VAS=6)	MF	44	49	Trunk, upper and lower limbs	Patches and plaques	–	$T_2N_1M_0$/ IIA
7	28/M	Pulmonary Tuberculosis	10	Asymptomatic	Ichthyosiform MF, Hypopigmented MF	18	18	Trunk, lower limbs	Hypopigmented lesions	–	$T_2N_0M_0$/IB
8	29/M		3	Pruritic (VAS=7)	MF	26	30	Trunk, buttocks, upper and lower limbs	Patch and follicular lesions	PCALCL	$T_2N_0M_0$/IB
9	26/F		6	Asymptomatic	Ichthyosiform MF, Ichthyosis vulgaris	20	20	Trunk, lower limbs	None	–	$T_2N_0M_0$/IB
10	57/M		20	Asymptomatic	Ichthyosis vulgaris, Ichthyosiform MF	37	37	Face, trunk, buttocks, upper and lower limbs	None	–	$T_2N_0M_0$/IB

DM: diabetes mellitus; BPH: benign prostatic hyperplasia; MF: mycosis fungoides; PCALCL: primary cutaneous anaplastic large cell lymphoma; M: male; F: female; VAS: visual analogue scale; TNM: tumor–node–metastasis.