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CASE REPORT

Mycosis Fungoides as an Ichthyosiform Eruption

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Ichthyosiform eruption as a specific manifestation of mycosis fungoides is very rare and only a few such cases have currently been reported in the medical literature. A 63-year-old Korean man presented with a 4-year history of a pruritic ichthyotic eruption. There was no personal or family history of ichthyosis or atopy. The ichthyosiform skin changes involved the abdomen, arms, thighs and shins. The face, palms and soles were spared. There was no peripheral lymphadenopathy or organomegaly. The typical lesions of mycosis fungoides were not present. The results of the routine investigations were normal or negative. A skin biopsy specimen revealed the findings of early mycosis fungoides. He was successfully treated with photochemotherapy. **(Ann Dermatol 21(2) 182～184, 2009)**

-Keywords-
Ichthyosis, Mycosis fungoides

INTRODUCTION

Mycosis fungoides (MF) is the most common type of cutaneous T-cell lymphoma (CTCL), and MF is a malignant lymphoma that's characterized by the expansion of a clone of the $CD4^+$ (or helper) memory T cells that frequently lacks other normal T-cell antigens (CD7). The skin rash in MF patients usually consists of patches, plaques or tumors that may have a long natural history; however many atypical variants have also been reported[1]. The MF patients with malignancy sometimes have ichthyosis. Ichthyosiform eruption as a specific manifestation of MF is very rare and this represents 1.8% of all MF cases. Only a few cases of ichthyosiform eruption as a specific manifestation of MF have currently been reported in the English literature[1-8]. We report here on the case of a 63-year-old Korean man who presented with ichthyosiform eruption and a histological pattern of MF.

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CASE REPORT

A 63-year-old Korean man had a 4-year history of a pruritic eruption. There was no personal or family history of ichthyosis or atopy. The ichthyosiform skin changes were widespread, but they were mainly located on the abdomen, arms, thighs and shins (Fig. 1). The face, palms, soles and most flexure areas were spared. Yet it was interesting that we did find a finely scaled eruption on the axilla. There was no peripheral lymphadenopathy or organomegaly. The typical lesions of MF were not present. The results of the routine investigations were normal or

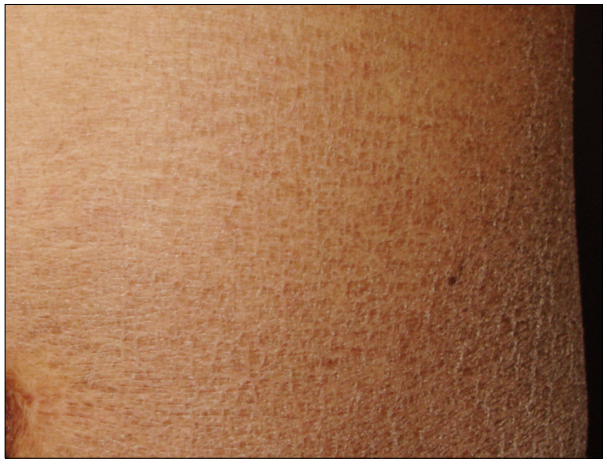

Fig. 1. Poorly defined irregular and polygonal scales on the abdomen.

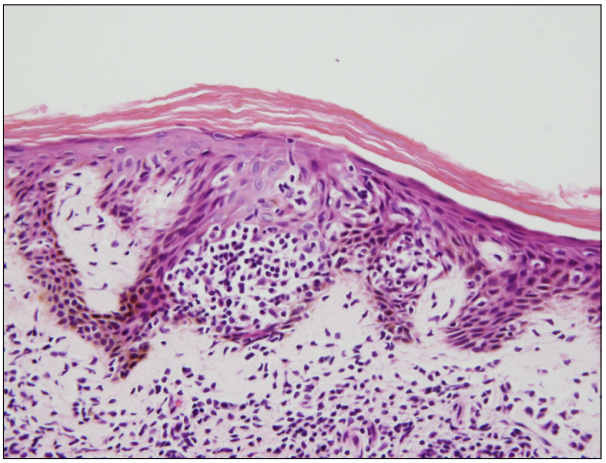

Fig. 2. The biopsy specimen shows a compact, thick orthokeratosis, a thinned epidermis with a slightly decreased granular layer and an upper dermal mononuclear cell infiltrate with Pautrier's microabscess in the epidermis (H&E, ×400).

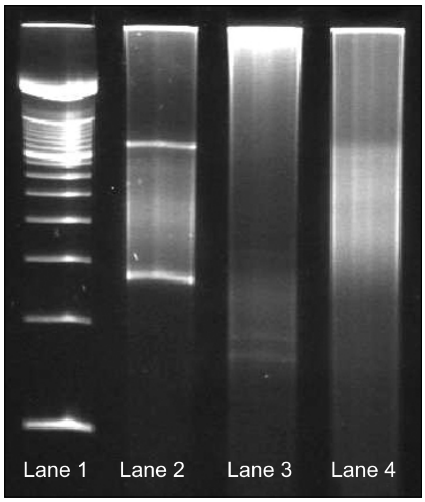

Fig. 3. PCR product run on SSCP gel. Lane 1: molecular weight marker. Lane 2 and Lane 3 show the Vγ1-8 and Vγ11 PCR products from the case, respectively. A well-defined monoclonal pattern was produced with the Vγ1-8 primer. The polyclonal control produced the pattern of smears in lane 4.

negative, including the complete blood count, the differential leukocyte count, the erythrocyte sedimentation rate and the blood chemistry studies. Antinuclear antibodies were not detected. The serologic studies were negative for HIV. No Sézary cells were present in the peripheral blood. Chest x-ray examination, gastroscopy and proctoscopy revealed no abnormalities. A skin biopsy specimen from the ichthyosiform lesion on the abdomen revealed parakeratosis and focally compact orthokeratosis of the epidermis with an underlying thinned granular layer. In the superficial dermis, there was a lichenoid infiltrate that mainly consisted of lymphocytes and histiocytes and it also showed epidermotrophism and microabscess (Fig. 2). The lymphocyte nuclei were hyperchromatic and cerebriform. Immunostaining confirmed the T cell helper pattern of the dermal and epidermal lymphocytes (CD2+, CD3+ and CD4+, but CD20- and CD30-). To assess the clonality in the paraffin-embedded samples, polymerase chain reaction single-strand conformational polymorphism (PCR-SSCP) was performed as previously described[9] to detect T-cell receptor γ (TCR-γ) gene rearrangements. On the results, a monoclonal T-cell clone was detected in the ichthyosiform skin lesion (Fig. 3). Genomic DNA was obtained from the 5-μm sections of the formalin-fixed paraffin-embedded tissues by dewaxing with xylene, proteinase K digestion and phenol/chloroform extraction. The DNA was precipitated in ethanol, dried and dissolved in distilled water. Vγ1-8, Vγ9, Vγ10, Vγ11 and Jγ1/Jγ2 consensus primers were used and the PCR products were analyzed by SSCP combined with 20% polyacrylamide gel electrophoresis. A chronic nonspecific dermatitis sample was used as a polyclonal control.

The diagnosis of Mycosis fungoides stage Ib (T2N0M0) was established. The patient was treated with twice weekly PUVA photochemotherapy for 3 months, and the ichthyosiform eruption regressed.

DISCUSSION

In general, MF is a slowly progressing chronic disorder. It usually begins as flat patches, which may or may not be histologically diagnosed as MF. Many variations of MF have been described, such as the follicular, granulomatous, hypo- or hyperpigmented and unilesional varients[1]. Moreover, ichthyosis-like eruption may very rarely be the first sign of MF. Acquired ichthyosis is recognized as one of the cutaneous manifestations of malignancies[9]. A few cases have recently been reported in which the ichthyosiform eruption proved to be a specific manifestation of MF[2-8].

Acquired ichthyosis may develop in patients of any age and who have several systemic diseases. It has occurred in association with Hodgkin's lymphoma, non-Hodgkin's lymphoma, MF, multiple myeloma and carcinomatosis. Ichthyosiform MF differs from acquired ichthyosis associated with cutaneous lymphoma[10]. In the latter condition, the patient presents with both the cutaneous specific lesions of lymphoma and ichthyosis. However, a skin biopsy of the ichthyotic lesions does not show any pathologic aspect of lymphoma, but rather, it shows only epidermal hyperplasia. In the former condition, on the

Table 1. Summary of the reported cases that present ichthyosiform eruption as the sole manifestation of mycosis fungoides

Authors	Age (years) /Sex	Duration of ichthyosiform eruption (years)	Distribution of ichthyosiform eruption	Suspicious findings of MF	History of icthyosis vulgaris	Other clinical findings
Hodak et al[4]	68/M	1.5	Trunk, arm, leg	None	None	None
	42/M	3	Face, trunk, arm, leg	None	None	None
Badawy et al[6]	67/M	1	Leg	None	NA	None
Marzano et al[7]	44/M	10	Whole body	None	NA	None
Kutting et al[8]	25/M	2	Trunk, arm, shoulder, thigh	None	None	Lymph node enlargement
Present case	63/M	4	Abdomen, arm, thigh, shin	None	None	None

NA: information not available

other hand, ichthyosis can be the only clinical manifestation of MF, and the histologic findings are consistent with both ichthyosis and MF as well. According to the clinical findings, ichthyosiform MF can be divided into 3 types: 1) ichthyosiform eruption as the sole manifestation of the disease, 2) ichthyosiform eruption in conjunction with additional atypical findings of MF and 3) ichthyosiform eruption in combination with the classic types of MF. To date, 5 cases[4,6-8] that showed ichthyosiform eruption as the sole manifestation of MF have been reported (Table 1). Our patient also had the sole manifestation of ichthyosiform MF. Clinical remission in such cases is obtained with nonaggressive therapies such as topical treatments, PUVA therapy and ultraviolet light B (UVB)[6].

Acquired ichthyosis can either indicate the presence of a severe underlying disease or it reveals an atypical form of cutaneous T cell lymphoma. Thus, the patients with acquired ichthyosis should be carefully evaluated by performing a through biopsy.

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