

LETTERS TO THE EDITOR

Lymphomatoid Papulosis Showing $\gamma\delta$ T-cell Phenotype

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Lymphomatoid papulosis (LP) is a chronic papulo-necrotic or papulonodular skin disease with histological features suggestive of malignant lymphoma (1, 2). The disease is characterized by recurrent pruritic papules at different stages of development, which arise on the trunk and limbs. LP is a part of primary cutaneous CD30⁺ lymphoproliferative disorders.

$\gamma\delta$ T cells represent a small subset of T cells that possess a distinct T-cell receptor (TCR) on their surface. The exact function of $\gamma\delta$ T cells remains unknown. Primary cutaneous T-cell lymphoma, in which tumour cells show $\gamma\delta$ T-cell phenotype, is classified into primary cutaneous $\gamma\delta$ T-cell lymphoma in the current World Health Organization (WHO) classification regardless of the pathology (1). Some cases show subcutaneous panniculitis-like infiltrates, while others show predominantly dermal infiltrates with or without epidermotropism. Most cases clinically show panniculitis-like ulcerated plaques or patches resembling mycosis fungoides. To our knowledge, there has been no reported case whose clinical behaviour is typical of LP.

CASE REPORT

In January 2009, a 37-year-old Japanese man was referred to our department with a 10-year history of multiple red papules on his limbs (Fig. 1). Each papule healed in a few weeks, but new lesions continued to appear. Clinical history and general physical examination were unremarkable. Histopathological examination of the biopsy specimen from the left thigh revealed a prominent epidermal infiltrate of large atypical lymphocytes

and dermal infiltrate of mixture of small and large lymphocytes (Fig. 2a). Dermal small lymphocytes were CD3⁺, CD4⁺, CD8⁻, CD20⁻, CD45RO⁺, and CD30⁻ (some populations were CD4⁻, CD8⁺), while large atypical lymphocytes were CD3⁺, CD4⁻, CD8⁺, CD45RA⁺, and CD30⁺ (Fig. 2b, and data not shown). Those large cells also expressed Granzyme B (Fig. 2c), perforin, and T-cell intracellular antigen-1 (data not shown), which were all granule-associated proteins of cytotoxic lymphocytes. Interestingly, large atypical cells mainly in the epidermis were TCR β F1⁻ and TCR- δ 1⁺ (Fig. 2d and data not shown). Systemic examination with gastrointestinal endoscope, computed tomography, and positron emission tomography showed no evidence of disease. The patient was diagnosed with LP. There has been no sign of development into lymphoma, but waxing and waning of the skin lesions has been ongoing in the 20 months after the initial diagnosis.

DISCUSSION

LP is considered as a benign disease that has an excellent prognosis. A recent study showed that only 4% of patients developed to systemic lymphoma and only 2% died of systemic disease over a median follow-up period of 77 months (2). LP histologically shows dermal infiltrate of large atypical lymphocytes positive for CD30. Those atypical lymphoid cells have variable expression of the pan-T-cell antigens, CD2, CD3, CD5, CD7, and CD45 (1). While most cases of LP show CD4⁺ phenotype, cases with CD8⁺ LP have also been noted in the literature (3–5). There has been no paper

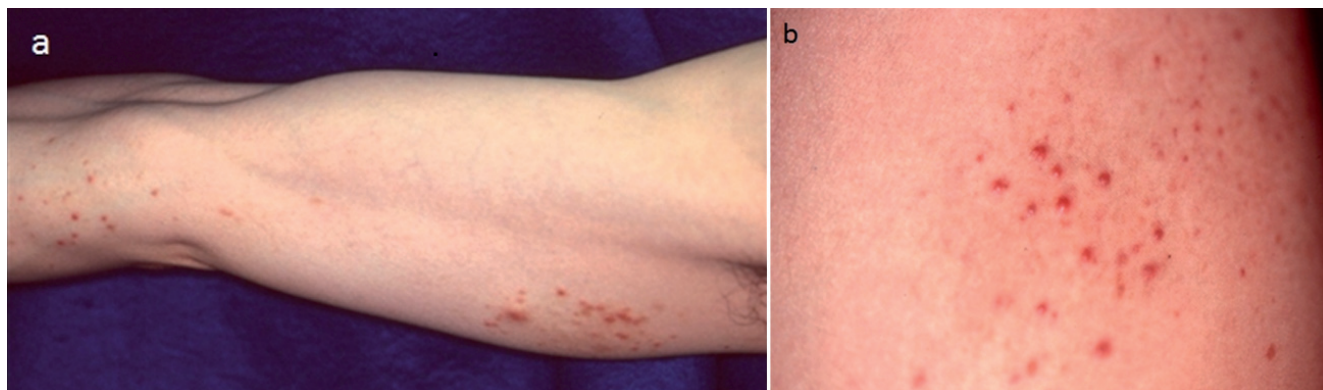


Fig. 1. (a) Multiple papules on the upper extremities. (b) Close-up of non-necrotic red-to-purple papules.

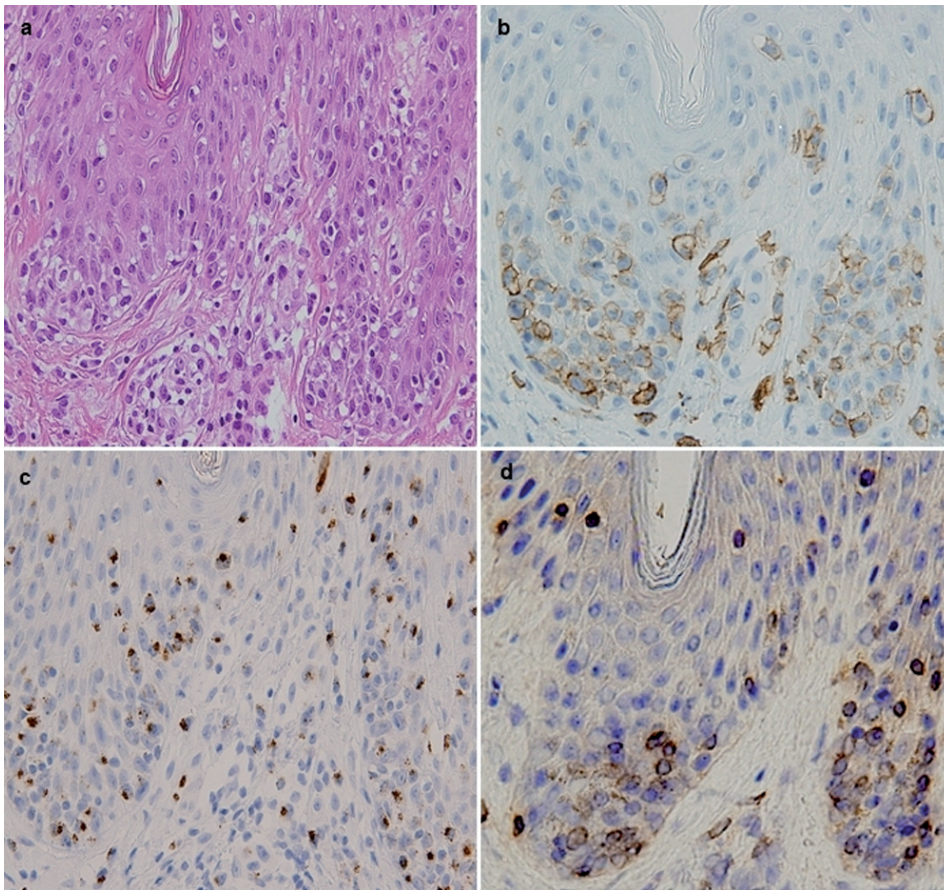


Fig. 2. (a) Infiltration of small-to-large lymphocytes in the epidermis and superficial dermis. Some large cells show nuclear atypia (haematoxylin-eosin stain, original magnification $\times 400$). (b) CD30 staining. (c) Granzyme B staining. (d) T-cell receptor- $\delta 1$ staining.

studying whether large atypical lymphocytes express TCR $\alpha\beta$ or $\gamma\delta$. To our knowledge, this is the first case of LP showing $\gamma\delta$ phenotype.

Primary cutaneous $\gamma\delta$ T-cell lymphoma, which is a clonal proliferation of mature, activated $\gamma\delta$ T cells with a cytotoxic phenotype, is a new entity in the current WHO classification (1). The prognosis of $\gamma\delta$ T-cell lymphoma is usually very poor. However, cases with more indolent clinical courses have been reported (6, 7). So far, not all cases with cutaneous lymphoma have been examined for TCR $\alpha\beta$ or $\gamma\delta$ expression because immunohistochemistry using frozen samples or flow cytometric analysis has been necessary. As a new method for detecting TCR $\gamma\delta$ using paraffin-embedded sections is now available (8), more cases of TCR $\gamma\delta$ phenotype will be reported, and our concept of primary cutaneous $\gamma\delta$ T-cell lymphoma may change.

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