

SHORT COMMUNICATION

Recurrent Course and CD30 Expression of Atypical T Lymphocytes Distinguish Lymphomatoid Papulosis From Primary Cutaneous Aggressive Epidermotropic CD8⁺ Cytotoxic T-cell Lymphoma

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Accepted Jan 8, 2014; Epub ahead of print Feb 18, 2014

Lymphomatoid papulosis (LyP) is characterised by a chronic course of years to decades of recurrent papulonodular lesions, each of which undergoes spontaneous regression after weeks or months (1).

Recently LyP type D was identified as a new histopathological variant simulating a primary cutaneous aggressive epidermotropic CD8⁺ cytotoxic T-cell lymphoma (pCAE-CD8⁺ CTCL) (2). Despite histologically alarming features, the patients have a clinical presentation and indolent course that are similar to those with typical cases of LyP.

CASE REPORT

A 24-year-old Japanese woman with a 10-year history of slightly itchy erythematous papules and plaques on the whole body was referred to our hospital. Individual lesions were self-healing within a few months, even though her papules and plaques never disappeared completely. Although she was initially diagnosed

with atopic dermatitis, topical corticosteroids were ineffective.

Physical examination revealed reddish papules and plaques with a crusted surface, and brownish macules, scattered on her trunk, thighs and arms (Fig. 1, upper). There was neither palpable lymphadenopathy nor hepatosplenomegaly. She reported neither fever nor weight loss. Laboratory data showed no elevated levels of sIL-2R or LDH. PET-CT scanning showed no remarkable FDG uptake, including the lymph nodes and skin lesions. Histopathologically, a biopsy from a plaque on her waist showed prominent epidermotropism. The atypical lymphocytes were characterised by large pleomorphic cells. On immunohistochemical staining, the infiltrating lymphocytes expressed CD3, CD8, CD30, granzyme B and TIA-1 (Fig. 2). They were negative for ALK, CD56 and EBER-1. Southern blot analysis showed TCR V β /J β 1 rearrangement. From the combination of clinical, histological, and immunohistochemical features, we diagnosed her as having LyP type D.

The eruptions improved with no active treatment after 7 months, without progression or signs of extracutaneous involvement (Fig. 1, lower).

DISCUSSION

LyP type D is difficult to differentiate from pCAE-CD8⁺ CTCL (2). pCAE-CD8⁺ CTCL is characterised by the rapid onset of plaques and tumours, frequently exhibiting necrosis and ulceration (3). The clinical course is aggressive, with a median survival of 32 months. Histologically, pCAE-CD8⁺ CTCL show striking epidermotropism of atypical lymphocytes with a CD8⁺ cytotoxic phenotype.

Although we initially suspected our patient of having pCAE-CD8⁺ CTCL, clinical presentation showed waxing and waning papules that resolved spontaneously. Furthermore, we found that the atypical lymphocytes expressed abundant CD30. Therefore, we finally diagnosed our patient as having LyP type D.

As shown in Table I, LyP is divided into 4 subtypes by a diagnostic criteria for CD30⁺ LyP (1). Epidermotropic infiltrate of atypical CD30⁺ or CD30⁻ lymphoid cells is also found in type B. In our case, most CD30 cells in epidermis expressed CD8 that histologically

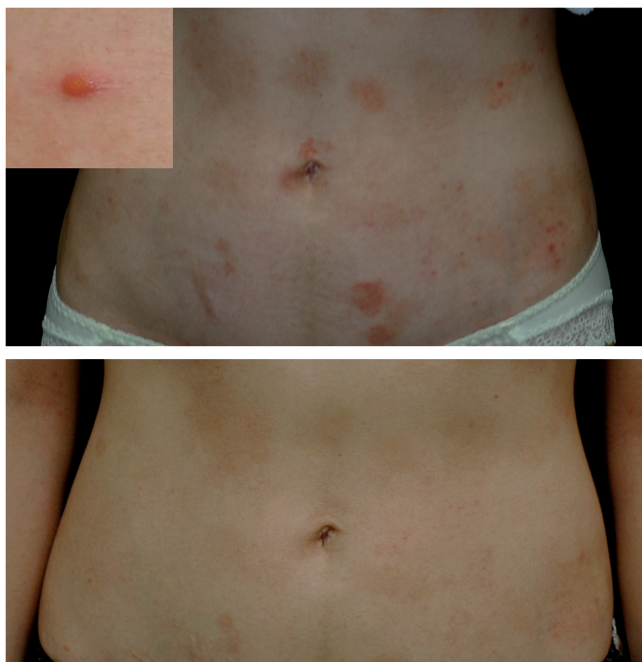


Fig. 1. Erythematous papules and plaques on the abdomen (upper). The insert is the close-up of the papule on the waist. 7 months later, most of the eruptions have disappeared without any active treatment (lower).

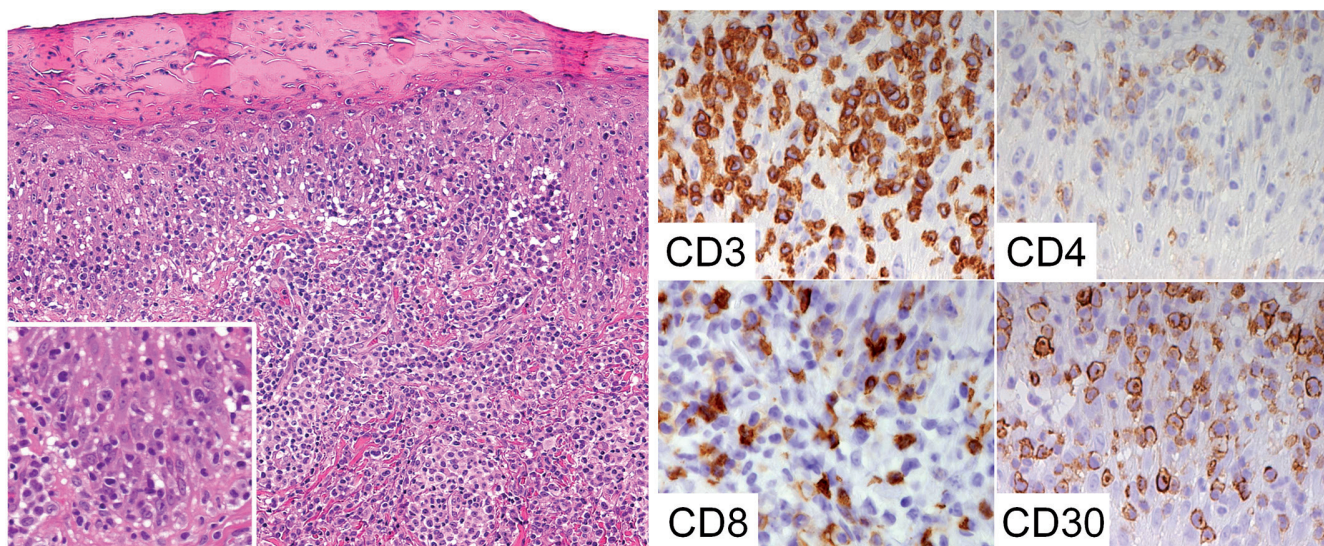


Fig. 2. Haematoxylin and eosin staining of the biopsy specimen sampled from a plaque on the waist. Massive proliferation of large pleomorphic lymphoid cells with remarkable epidermotropism. The inset is the close up of infiltrating lymphocytes in the epidermis. Immunohistochemical staining showed CD3, CD8, and CD30 expression by the infiltrating lymphocytes.

resembles pCAE-CD8⁺ CTCL, leading to the diagnosis of LyP type D.

Gormley et al. (4) suggest that low-grade CD8⁺ cases of CTCL fulfill immunophenotypic and often histologic criteria for pCAE-CD8⁺ CTCL, but follow a benign chronic course without progression or signs of extracutaneous involvement, and have prognosis similar to patch-stage mycosis fungoides (MF). The considerable histologic and immunophenotypic overlap between the more indolent and aggressive cases means that distinction must be made based on clinical features, history of disease course, and results of patient physical examination.

Another possible differential diagnosis of this case is a CD8⁺ MF or CD30⁺ MF variant. Prince et al. (5) suggested that the spontaneous remission occurs in MF and Sezary syndrome. Although MF was involved in the differential diagnosis from the clinical features this possibility would be less likely because the degree of epidermotropism in MF is not so marked (6).

Table I. Histologic criteria of lymphomatoid papulosis (1)

Type A	Wedge-shaped infiltrate with scattered or clustered CD30 ⁺ tumor cells, intermingled with numerous inflammatory cells, such as small lymphocytes, neutrophils, eosinophils, and histiocytes. Type A is the most common histologic presentation.
Type B	Epidermotropic infiltrate of small atypical CD30 ⁺ or CD30 ⁻ lymphoid cells with cerebriform nuclei that histologically resembles MF.
Type C	Cohesive sheets of CD30 ⁺ large atypical lymphoid cells with only a few admixed reactive inflammatory cells.
Type D	Epidermotropic infiltrate of small-to medium-sized atypical CD8 ⁺ and CD30 ⁺ lymphoid cells that histologically resembles primary cutaneous aggressive epidermotropic CD8 ⁺ cytotoxic T-cell lymphoma.

In conclusion, we described LyP type D characterised by striking similarities to pCAE-CD8⁺ CTCL. Despite histologically alarming features, the patients have an indolent course, as do other variants of typical cases of LyP. The recognition of features of LyP type D is important for the appropriate management of the patients. This variant must be distinguished from the pCAE-CD8⁺ CTCL to avoid overly aggressive treatment.

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