

## SHORT COMMUNICATION

### Lymphomatoid Papulosis Type D or an Aggressive Epidermotropic CD8<sup>+</sup> Cytotoxic T-cell Lymphoma?

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Lymphomatoid papulosis (LyP) is defined as a chronic, recurrent, self-healing eruption of papules and small nodules, characterised by a waxing and waning course, and by histological features of a CD30<sup>+</sup> cutaneous T-cell lymphoma CTCL. Classically, 3 histologic subtypes of LyP are recognised: Type A (histiocytic), type B (MF-like) and type C (anaplastic like CTCL) (1).

Together with primary cutaneous anaplastic large T-cell lymphoma, LyP is classified as a part of the primary cutaneous CD30<sup>+</sup> lymphoproliferative disorders in the WHO-EORTC classification of cutaneous lymphomas (2).

Recently, a new histologic LyP variant termed type D, simulating an aggressive epidermotropic CD8<sup>+</sup> cytotoxic T-cell lymphoma, but with the typical clinical presentation and indolent course of LyP, has been described (3–5). We describe a new case of this variant, of which diagnosis is particularly challenging and emphasises the importance of cross-disciplinary collaboration.

#### CASE REPORT

A 48-year-old woman referred from a private dermatologist, presented with a 3-year history of asymptomatic small inflammatory red papules distributed over her trunk and limbs. The papules erupted and healed within weeks or months leaving the area hyperpigmented (Fig. 1).

On admission the patient did not have any other systematic symptoms as fever, weight loss or night sweating. Apart from the skin rash, the physical examination, including examination for lymphadenopathy or organomegaly, did not reveal any abnormalities. Except for a diagnosis of chronic obstructive lung disease the patient was otherwise healthy.

The first biopsy taken by the private dermatologist revealed a markedly epidermotropic infiltrate, and superficial dermal band-like and deep perivascular infiltrates of small slightly atypical lymphocytes (Fig. 2). The lymphocytes showed a cytotoxic CD3<sup>+</sup>, CD8<sup>+</sup>, Tia-1<sup>+</sup>, Perforin<sup>+</sup>, CD56<sup>+</sup> immunophenotype and also expressed the pan-T-cell markers CD2 and CD5, but partly lacked CD4 and CD7 (Fig. 3). Molecular genetic analysis showed clonal rearrangements of T-cell receptor genes. The histopathologic conclusion was a cutaneous CD8<sup>+</sup> epidermotropic cytotoxic clonal T-cell infiltrate suspicious of an aggressive epidermotropic CD8<sup>+</sup> cytotoxic T-cell lymphoma.

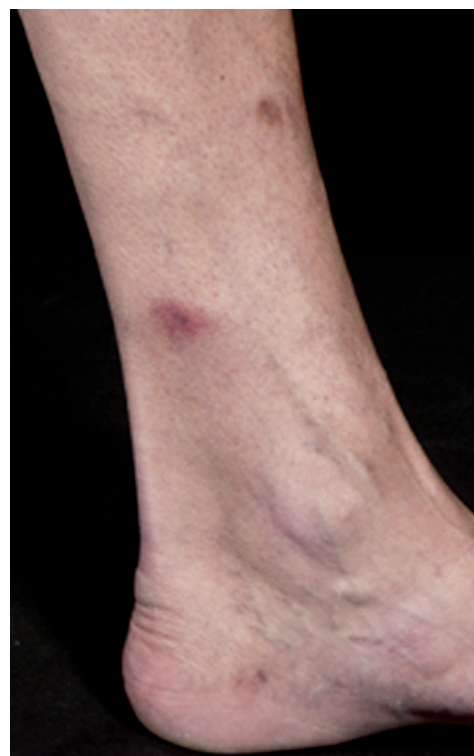


Fig. 1. The patient's ankle showing a new lesion and an old lesion with hyperpigmentation.

The second biopsy taken at the department of Dermatology from a new lesion showed on comparison identical histological, immunophenotypic and genetic

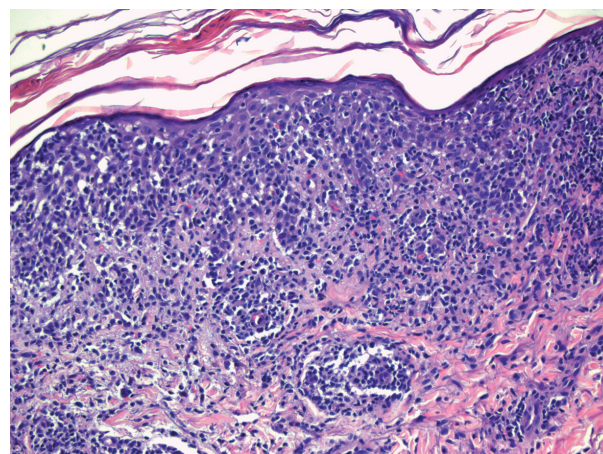


Fig. 2. Histological evaluation of the first biopsy taken. Epidermotropic infiltration by small atypical lymphocytes (H&E, original magnification  $\times 20$ ).

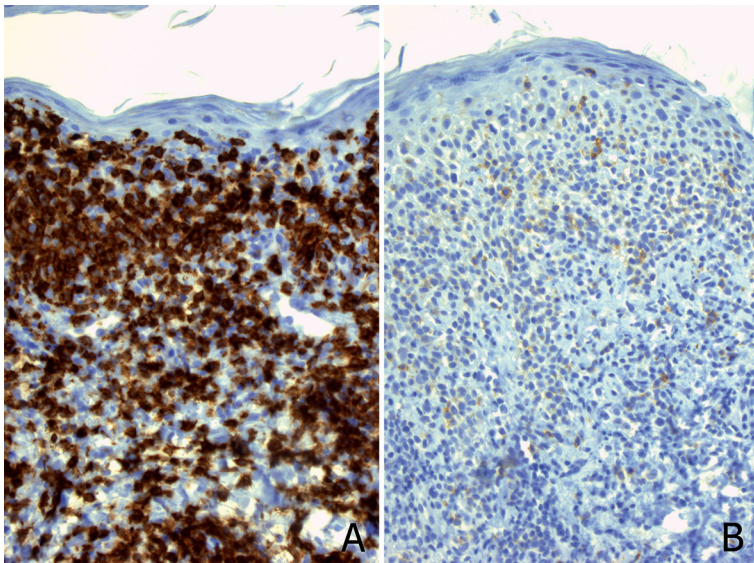


Fig. 3. Strong immunohistochemical reaction of infiltrating lymphocytes for CD8 (A) and occasional CD30<sup>+</sup> lymphocytes (B) (original magnification  $\times 20$ ).

features. An additional staining for CD30 of both the primary and secondary biopsy showed weak expression of occasional intraepidermal and dermal lymphocytes.

The patient underwent further diagnostic investigations including laboratory blood tests, a combined whole body and positron emission tomographies and a bone marrow biopsy. No systemic disease was revealed by the investigations. The combination of clinical and immunohistopathological features suggested the diagnosis of LyP, type D.

The patient was initially treated with topical steroids with no effect. Therefore the treatment was changed to oral methotrexate (Paranova, Ballerup, Denmark) 15 mg a week with excellent effect without emergence of new papules.

## DISCUSSION

This case stresses the importance of collaboration between the clinician and pathologist for correct diagnosis of cutaneous lymphomas. The first diagnosis which was proposed by the pathologist, aggressive epidermotropic CD8<sup>+</sup> cytotoxic T-cell lymphoma, is a condition with rapid onset of patches, plaques, nodules and tumours and often complicated with ulcerations and necrosis (6). The disease may disseminate to unusual sites, such as the lung, testis, central nervous system, and oral cavity, but not to the lymph nodes. It is known to have an aggressive course with a median survival of 32 months. The positive immunohistochemical staining for CD30, albeit weak, and the clinical presentation were inconsistent with these diagnoses, and a rare case of LyP, type B was suggested. However, LyP, type B is characterised by marked epidermotropism but this case differed immunophenotypically by strong reaction for CD8 instead of the usual pure CD4 reaction, and the newly suggested LyP, type D included all of these features. LyP is characterised as an indolent, sometimes self-limited condition

with a disease-specific 5-year survival rate close to 100% (2, 3). The final diagnosis was therefore LyP, type D, which is in agreement with the fact that our patient had no systemically involvement and no progression of the disease despite an anamnesis of 3 years.

This case is another example of the newly described LyP, type D with an excellent prognosis despite the aggressive pathological appearance. It is important to recognise this subtype of LyP in order to avoid unnecessary worrying and aggressive therapeutic measures.

*The authors declare no conflicts of interest.*

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