

## An Unusual Case of Peripheral T-cell Lymphoma

V. Broshtilova<sup>1</sup>, M. Gantcheva<sup>2</sup>

<sup>1</sup>*Department of Dermatology and Venereology, Faculty of Medicine, Medical University, Sofia, Bulgaria*

<sup>2</sup>*Institute of Experimental Morphology, Pathology and Anthropology with Museum,  
Bulgarian Academy of Sciences, Sofia, Bulgaria*

A 24-year-old woman presented with 2-year history of erythematous patches, papules and plaques on her trunk and extremities. Skin lesions were unpainful, slightly itchy, and showed tendency to resolve spontaneously. Medium to large atypical lymphoid cells with focal epidermotropism were seen on routine histology. Immunophenotyping revealed CD2+, CD3+, CD5+ and partly CD4+, PD-1+, Bcl-6+, CXCL-13+,  $\beta$ F1+. TCR-gamma, TIA-1, Granzyme B, CD8 were negative. Only few cells stained positive for CD56 and CD30. This constellation was more suggestive of peripheral T-cell lymphoma, not otherwise specified (PTCL/NOS). The lack of negative prognostic factors such as multifocal skin involvement with ulceration, concurrent extracutaneous disease, age older than 70 years and predominantly large cell morphology favor the diagnosis of atypical immunohistochemical variant of mycosis fungoides (MF). PTCL/NOS are extremely rare cases with bad prognosis and rapid fatal outcome, which must always be ruled out from all cases of MF by complete work-up and accurate clinical history.

*Key words:* mycosis fungoides, peripheral T-cell lymphoma, not otherwise specified.

### Introduction

Peripheral T-cell lymphomas, not otherwise specified (PTCL, NOS), represent a heterogeneous group of T-cell neoplasms that do not fit to any defined T-cell entities [2, 3]. Most cases represent late stage of nodal disease, although extranodal involvement is common. Only 20% of PTCLs show skin involvement [5]. These are extremely rare cases with bad prognosis and rapid fatal outcome, which must always be ruled out from all cases of mycosis fungoides (MF) by complete work-up and accurate clinical history [4].

### Case report

A 24-year-old Caucasian female presented with 2-year history of erythematous patches, papules and plaques on her trunk and extremities. Skin lesions were unpainful, slightly itchy, and showed tendency to resolve spontaneously. The first lesion reportedly appeared on the left arm, with the most recent ones developing on the face and eyebrows. The patient claimed episodes of pruritus and dry patches on her hands during childhood that usually disappeared under sun. She had 10-time elevated total IgE, which was first

detected two years ago and persisted to date. Her brother had severe atopic dermatitis presented by refracting erythematous patches and diffuse xerosis.

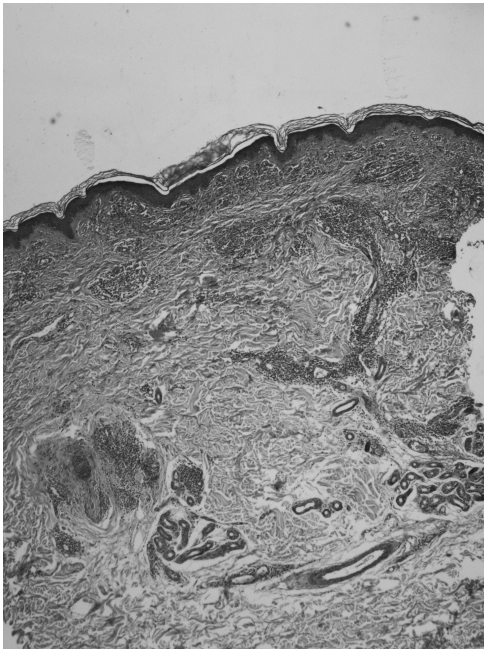
Examination revealed multiple uniform firm erythematous papules with a diameter of 1 cm on her upper and lower extremities (**Figs. 1, 2**). Multiple skin biopsies were subjected to histological examination showing parakeratosis, irregular acanthosis, mild to moderate medium and large atypical lymphoid cells (**Figs. 3, 4**). There were some



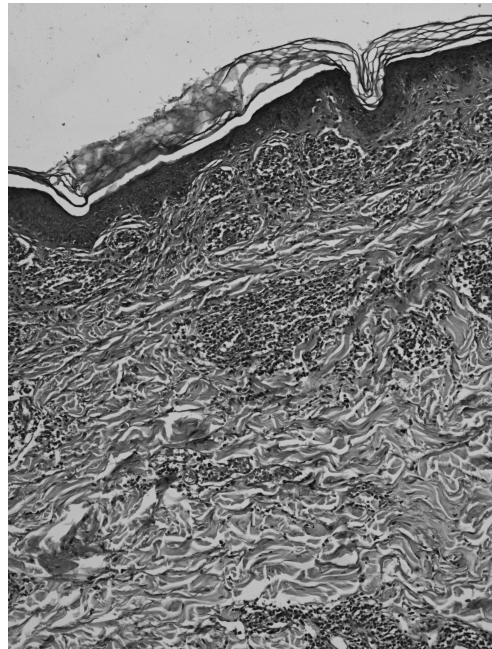
**Fig. 1.** An erythematous patch of uniform firm papules on the left elbow



**Fig. 2.** An erythematous plaque of the right cubital fold



**Fig. 3.** Parakeratosis, acanthosis, mild to moderate perivascular and periadnexal infiltrate of atypical medium to large lymphoid cells in the dermis. HE  $\times$  100



**Fig. 4.** Medium to large atypical lymphoid cells, focal perivascular necrosis. HE  $\times$  400

areas of peri-follicular necrosis. Immunophenotyping revealed CD2+, CD3+, CD5+ and partly CD4+, PD-1+, Bcl-6+, CXCL-13+,  $\beta$ F1+. TCR-gamma, TIA-1, Granzyme B, CD8 were negative. Only few cells stained positive for CD56 and CD30. Flow cytometry did not find any peripheral aberrant lymphoid proliferation.

Abdominal CT scan and ultrasound ruled out systemic involvement. PET scan showed hypermetabolic activity in a solitary right inguinal lymph node, which upon histology turned out to be free of specific involvement. Narrow band UV B therapy was introduced with good clinical outcome, therefore, 5000 mJ/sq. cm was recommended on a weekly basis.

## Discussion

Skin lymphomas usually show T-cell origin. Mycosis fungoides and Sezary syndrome comprise the majority of these cases. The primary skin lesions encompass co-existent patches, plaques or nodules, which may or may not ulcerate spontaneously. Erythroderma is typical for Sezary syndrome. Clinical picture corresponds to atypical lymphoid cells that show specific immunophenotypical characteristics, according to which the complicated classification of the various types of peripheral T-cell lymphomas is structured [6].

Interestingly, our case did not meet the histopathological or immunophenotypical criteria for any of the other well-defined subtypes of peripheral T-cell lymphoma. According to these criteria, it has to be classified as PTCL/NOS [1, 3].

Less than 100 patients were described with PTCL/NOS to date [2, 7]. In these limited reports, patients tend to present with generalized lymphadenopathy and gastrointestinal involvement. Cutaneous disease is usually represented with disseminated nodules. One fifth of the patients observed had concurrent nodal involvement at the time of presentation of the first skin lesions.

In contrast, our patient did not show any of the clinical features described. She had rather indolent course of the disease presented by erythematous papules and patches that resolved spontaneously and did not ulcerate. The lack of systemic involvement and peripheral aberrant lymphoid clone, together with favorable response to phototherapy, suggest more of an atypical immunohistochemical variant of MF.

## Cocclusions

We ruled out the negative prognostic indicators – multifocal skin involvement with ulceration, concurrent extracutaneous disease, lack of spontaneous improvement, age older than 70 years and large cell morphology, to favor our diagnosis. A close clinical, histological and laboratorial follow-up is, however, needed to confirm our suggestion.

## References

1. **Bekkenk, M. W., M. H. Vermeer, P. M. Jansen, A. M. van Marion, M. R. Canninga-van Dijk, P. M. Kluin et al.** Peripheral T-cell lymphomas unspecified presenting in the skin: Analysis of prognostic factors in a group of 82 patients. – *Blood*, **102**(6), 2003, 2213-2219.
2. **Friedman, B. J., C. McHargue, M. Nauss.** Peripheral T-cell lymphoma, not otherwise specified with prominent cutaneous involvement. – *Ind. J. Dermatol. Venereol.*, **81**(5), 2015, 535-537.

3. **Kempf, W., S. Rozati, K. Kerl, R. Dummer.** Cutaneous peripheral T cell lymphomas, unspecified /NOS and rare subtypes: A heterogeneous group of challenging cutaneous lymphomas. – *G. Ital. Dermatol. Venereol.*, **147**(6) 2012; 553-562.
4. **Schmitz, N., L. Trümper, M. Ziepert, M. Nickelsen, A. D. Ho, B. Metzner et al.** Treatment and prognosis of mature T-cell and NK-cell lymphoma: An analysis of patients with T-cell lymphoma treated in studies of the German High-Grade Non-Hodgkin Lymphoma Study Group. – *Blood*, **116**(18), 2010, 3418-3425.
5. **Swerdlow, S. H., E. Campo, N. L. Harris, E. S. Jaffe, S. A. Pileri, H. Stein et al.** WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues. Lyon, France: IARC Press; 2008.
6. **Wallett, A., J. S. Ibbetson, D. Kearney, K. Newland, S. Sidhu.** Cutaneous manifestations of peripheral T-cell lymphoma, not otherwise specified: a case series highlighting the diagnostic challenges for this heterogeneous group. – *Australasian J. Dermatol.*, **56**(3), 2015, 197-201.
7. **Weisenburger, D. D., K. J. Savage, N. L. Harris, R. D. Gascoyne, E. S. Jaffe, K. A. MacLennan et al.** Peripheral T-cell lymphoma, not otherwise specified: A report of 340 cases from the International Peripheral T-cell Lymphoma Project. – *Blood*, **117**(12), 2011, 3402-3408.