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Papuloerythroderma of Ofuji (PEO) successfully treated with acitretin

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Introduction

Papuloerythroderma of Ofuji (PEO) is a rare skin disorder, which affects predominantly older males of Asian or Caucasian descent with a male to female ratio of 4:1. It is characterized by pruritic flat-topped erythematous papules, which might progress into erythroderma. Exanthema typically spares the skin folds, which is known as the deck-chair sign. Common laboratory findings include lymphopenia, peripheral eosinophilia, and elevated serum IgE. Histological image shows nonspecific inflammatory reaction and needs to be correlated with clinical and laboratory findings [1, 2]. PEO should remain a diagnosis of exclusion after eliminating more frequent causes of itch and eczematous rash, such as atopic dermatitis of the elderly [3].

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Case 1

An 85-year-old man was referred with a 2-year history of pruritic nonconfluent erythematous papules sparing the skin folds. No systemic underlying cause had been identified. Blood work-up showed lymphopenia, eosinophilia, and elevated serum IgE. Histology shows edematous changes of papillary dermis with mixed inflammatory infiltrate comprising multiple mast cells, eosinophils, and isolated plasma cells. The diagnosis of PEO was made based on the typical clinical and laboratory findings and compatible histopathological features. Systemic treatment with acitretin (25 mg/day) combined with topical corticosteroids led to complete resolution of the skin manifestations after 3 months of treatment.

Case 2

A 95-year-old man was referred to our dermatology clinic with a 3-month history of pruritic papular exanthema (Fig. 1a), now progressing to erythroderma sparing the skin folds (Fig. 1b). Indolent multiple myeloma has been diagnosed 4 years prior to rash onset. Histology showed nonspecific interstitial and perivascular dermatitis with multiple eosinophils (Fig. 2). Lymphopenia and eosinophilia further supported the diagnosis of PEO. Immunophenotypic blood analysis identified a monotypic TRBC1+CD4++CD3++CD5++CD7-CD26- population reaching 4.9% of lymphocytes with a blood T-cell receptor (TCR) clone. The patient was started on combination therapy with oral acitretin (10 mg/day) and topical corticosteroids with a complete skin response after 3 months (Fig. 1c). Patient continues to be monitored for a possible cutaneous T-cell lymphoma (CTCL) onset.

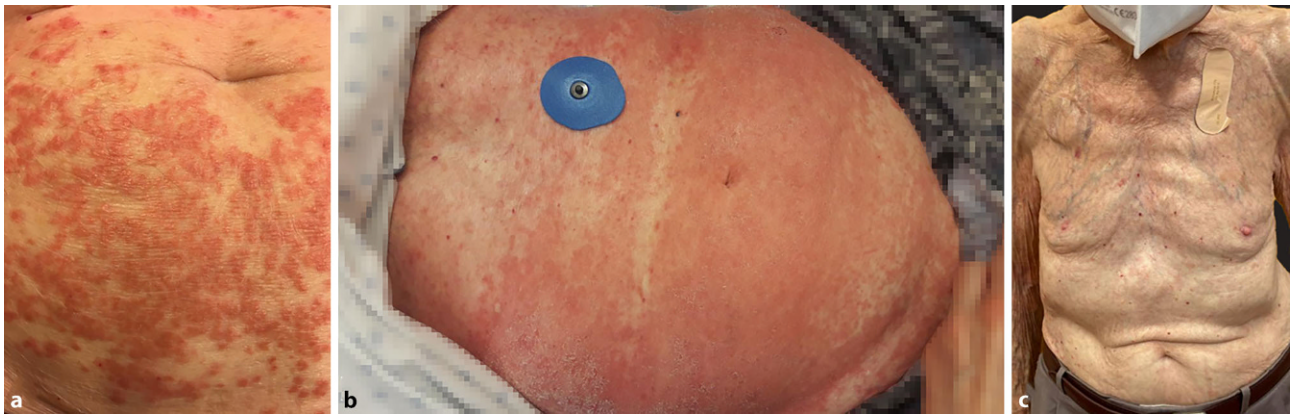

Fig. 1 Cutaneous manifestations in patient 2. Papular exanthema (**a**) progressing into skin-sparing erythroderma with deck-chair sign (**b**). Complete resolution of the skin manifestations after combination treatment with acitretin and topical corticosteroids (**c**)

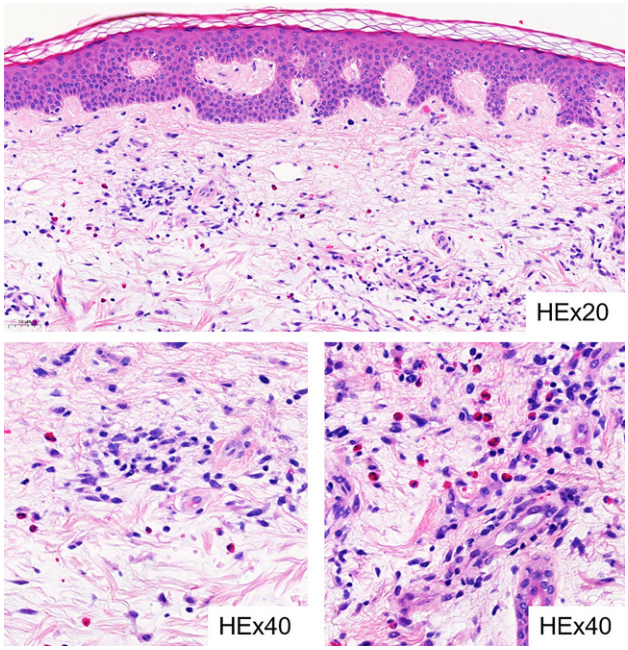

Fig. 2 Histological manifestations in patient 2. Skin biopsy showing nonspecific interstitial and perivascular dermatitis with multiple eosinophils. *HE* hematoxylin–eosin staining

Discussion

Approximately half of the reported PEO cases have been described as idiopathic. The other half has been defined as secondary PEO, associated with various systemic diseases, including malignancies, infections, and medications. Among those, cutaneous T-cell lymphoma (CTCL) has been identified as the most frequent underlying cause, found in half of the patients with secondary PEO [2]. The relationship between PEO and CTCL remains poorly understood but due to their well-established association, individuals with PEO should be closely monitored for possible CTCL onset [4]. Blood immunophenotyping and TCR clonality studies are necessary, while physicians should have a low threshold for performing repeated skin biopsies. We regarded patient 1 as an idiopathic PEO. In patient 2, we considered the PEO as a possible paraneoplastic dermatosis due to multiple myeloma although the possibility of a future CTCL emergence cannot be excluded. PEO pathogenesis is poorly elucidated and development of effective treatment guidelines thus remains challenging. As skin-homing Th2/Th22 cells seem to play a role in the pathogenesis of PEO, use of dupilumab, human monoclonal antibody against IL-4Rα, has recently been investigated [5]. Nevertheless, due to recent reports on worsening or progression of CTCL after dupilumab treatment [6], such treatment should be carefully considered in the PEO context, and only after exclusion of underlying CTCL. Herein, we report successful treatment of two PEO patients using combination therapy of oral retinoids and topical corticosteroids. Complete resolution of the dermatological manifestations was reached in both patients after 3 months of treatment. Patient 1 has subsequently continued acitretin treatment as maintenance therapy at a lower dose (10 mg/day), while patient 2 decided to stop treatment due to side effects after 3 months. Complete skin response persisted in both patients at 1 year after diagnosis. Our findings correlate well with recent reports suggesting higher success rates of oral retinoids compared to psoralen plus ultraviolet-A radiation (PUVA) therapy or oral corticosteroids, yet with a more favorable safety profile and patient tolerance [2].

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Declarations

Conflict of interest G. Blanchard and E. Guenova declare that they have no competing interests.

Ethical standards Discussed patients gave consent for their photographs and medical information to be published in print and online and with the understanding that this information may be publicly available.

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