Papuloerythroderma of Ofuji: First Case Report in Portugal

Papuloeritrodermia de Ofuji: Primeiro Caso em Portugal

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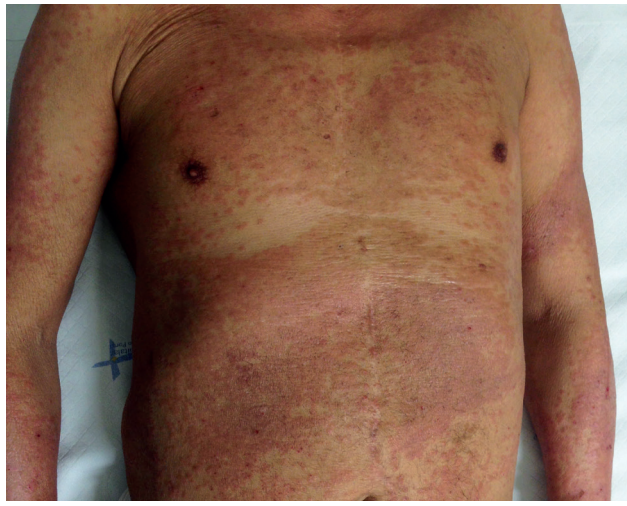

Figure 1 – Clinical aspect of the lesions. Cutaneous eruption sparing all skin folds – deck-chair sign.

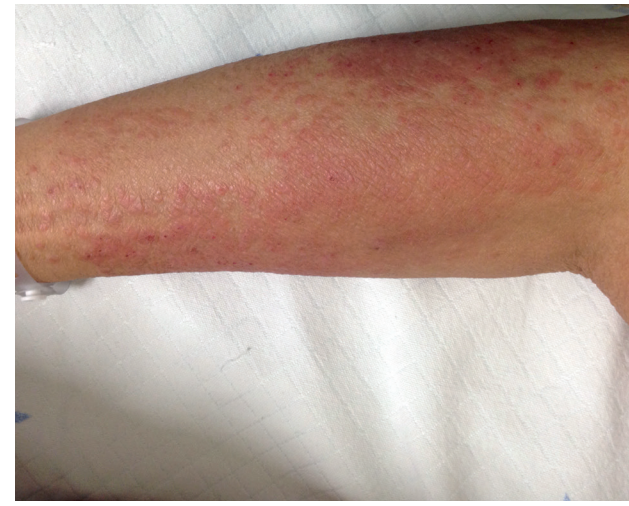

Figure 2 – Cobblestone-like pattern of the cutaneous lesions

A 65-year-old man presented with a pruritic, erythrodermic cutaneous eruption consisting of multiple flat-topped red-brown papules and plaques with a cobblestone appearance, strikingly sparing all skin folds (Figs. 1 and 2). Laboratory results showed peripheral eosinophilia (1.96 x 10^3/uL), lymphopenia (0.83 x 10^3/uL) and elevated total IgE (3951KU/L). Histopathology exhibited perivascular dermal infiltrate of lymphocytes and eosinophils. Cutaneous and peripheral blood cytometry, infectious and age-appropriate cancer screening revealed no changes. No atopy or drug history was identified.

Clinical and laboratory data fulfilled diagnostic criteria of primary papuloerythroderma of Ofuji.[1] The patient initiated PUVA phototherapy, with complete clinical response.

Papuloerythroderma of Ofuji represents a strikingly rare cause of erythroderma. Cutaneous lesions with a characteristic appearance (cobblestone-like) and distribution pattern (uninvolved skin folds – 'deck-chair sign') represent the hallmark of the dermatosis,[1,2] alongside the frequent association with peripheral eosinophilia, lymphopenia and increased IgE.[1,2]

Most cases have no underlying cause, while exclusion of secondary causes (neoplasms, infections, atopy or drugs) is of prognostic importance.[3,4]

PROTECTION OF HUMANS AND ANIMALS: The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the Helsinki Declaration of the World Medical Association.
DATA CONFIDENTIALITY: The authors declare having followed the protocols in use at their working center regarding patients' data publication.
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