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CASES FOR DIAGNOSIS

Erythroderma That Spares the Major Flexion Creases

M Fernández-Guarino,[a] R Carrillo-Gijón,[b] and E Muñoz-Zatoa

[a]Servicio de Dermatología and [b]Servicio de Anatomía Patológica, Hospital Ramón y Cajal, Madrid, Spain

Patient History

An 80-year-old man with no relevant history except for chronic gastritis presented with progressive brownish hyperpigmentation over the entire skin surface that had appeared 4 months earlier and was accompanied by severe pruritus. The patient had been assessed 2 years previously for lesions in the form of erythematous papules and plaques with well-defined borders on the arms, back, and groin. At that time, the histological study showed advanced lichenoid dermatitis with no other specific findings.

Physical Examination

The examination revealed generalized brownish hyperpigmentation, slightly infiltrated on palpation, in which the abdominal folds, skin folds on the thorax, and inguinal folds were unaffected, with generalized furfuraceous desquamation (Figures 2 and 3). Palmoplantar hyperkeratosis was also observed. The general examination was normal except for the presence of symmetrical enlarged axillary and inguinal lymph nodes of approximately 0.5 cm in diameter. The enlarged nodes had an elastic consistency, were freely movable, and were not painful.

Additional Examinations

The laboratory workup showed 11% eosinophilia, but the remaining differential white cell count and total leukocytes were normal. Lactate dehydrogenase and γ-glutamyl transferase were elevated. Hemostasis, immunoglobulin assays, computed tomography of the body, and abdominal ultrasound were normal. Human immunodeficiency virus testing was negative. Immunophenotyping in peripheral blood was normal and no Sézary cells were found in blood smears.

Correspondence:
Montse Fernández Guarino
Hospital Ramón y Cajal
Carretera de Colmenar km 9,100, 28034 Madrid, Spain
montsefdez@msn.com

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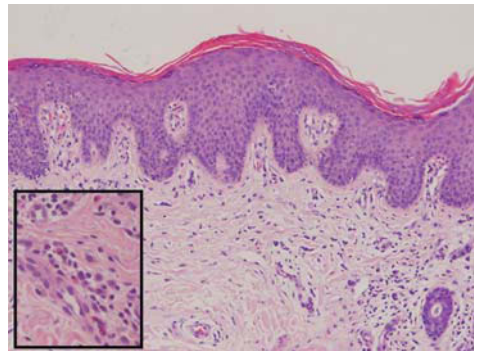

Figure 1. Hematoxylin-eosin, ×30

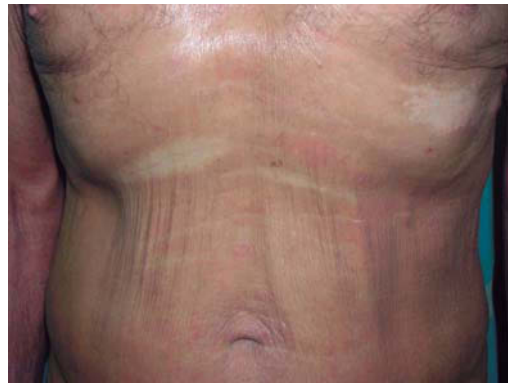

Figure 2.

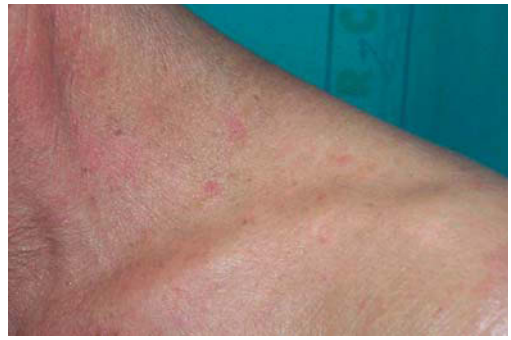

Figure 3.

Histopathology

Skin biopsy showed mild epidermal hyperplasia with areas of spongiosis and exudative phenomena in the horny layer, as well as parakeratosis. There was a mild lymphocytic inflammatory infiltrate with abundant eosinophils in the superficial dermis and some sclerosis of the papillary dermis.

What is your diagnosis?

Diagnosis

Papuloerythroderma of Ofuji

Treatment and Clinical Course

Oral prednisone was prescribed at a dose of 20 mg/d, with good control of the clinical symptoms and pruritus.

Comment

Papuloerythroderma of Ofuji was described as a distinct clinical entity in 1984 by Ofuji et al.[1] It is a disease of unknown etiology characterized by the onset of disseminated brownish erythematous papules that converge to produce erythroderma that typically spares the major skin folds (deck chair sign).[2]

The condition affects men more often than women, with a mean age of 60 years and approximate prevalence of 1.5 per million. Apart from cutaneous involvement, there may be other abnormalities, both dermatologically (palmoplantar keratoderma and nail-fold infarcts) and in the laboratory workup. The most characteristic laboratory finding is peripheral eosinophilia. High levels of immunoglobulin E and lymphopenia may also be detected. The results of liver function tests may be abnormal, with elevated alkaline phosphatase and γ-glutamyl transferase.[2]

In most cases, papuloerythroderma of Ofuji is idiopathic. It has been suggested that the condition is the expression of common chronic diseases of the skin, such as eczema or psoriasis, in elderly patients.[4] However, most patients have no prior history of skin disease or problems. Progression to cutaneous T-cell lymphoma has been described in a large number of cases, suggesting that it may be a prelymphomatous condition.[3] Therefore, such patients should undergo close clinical and histological follow-up.

Papuloerythroderma of Ofuji has also been reported in association with visceral neoplasms, Hodgkin lymphoma, acute myeloid leukemia, hypereosinophilic syndrome, AIDS, drug hypersensitivity, and biliary sepsis after cholecystectomy.[4,5]

The histology is nonspecific. The epidermis is usually normal, but may show acanthosis, spongiosis, parakeratosis, and occasionally exocytosis.[4]

The diagnosis is based on the clinical symptoms, laboratory workup, and pathology.

Although the cause of papular dermatosis is often unknown, the associations described in the literature make analysis and follow-up of the patients essential.

This requires an individualized approach to the management of this uncommon dermatosis, which should be considered a form of erythroderma that may be due to one of several etiologies. The term papuloerythroderma of Ofuji should be used for cases such as that of our patient, in whom the etiology of the dermatosis is unknown. In contrast, the term papuloerythroderma would be limited to cases with a well-defined etiology.

Various therapeutic options have been described in the literature. Improvement is obtained with topical and systemic corticosteroids at low doses, plus antihistamines. Psoralen-UV-A (PUVA), UV-B, retinoid-PUVA, and other treatments such as cyclosporine, etretinate, azathioprine, and interferon α have also been used successfully.[6]

Conflicts of Interest
The authors declare no conflicts of interest.

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