Ofuji's Papuloerythroderma: Report of a New Case Responding to PUVA

Sir,

Papuloerythroderma was first described in 1984 by Ofuji et al. (1) as a pruritic eruption of widespread red-brown flat-topped papules sparing the skin folds, a feature subsequently described by Farthing et al. (2) as "the deck-chair sign". This erythroderma is associated with hematologic abnormalities such as eosinophilia and lymphopenia. Treatment by topical steroids is generally unsatisfactory, but systemic steroids usually give a good result. We present a new case with some distinctive features, particularly a severe inflammatory involvement of the scalp and an excellent response to PUVA.

CASE REPORT

A 67-year-old white woman suddenly developed a severely itching dermatosis, which spread within 2 weeks over her entire body and particularly the scalp. She complained at the same time of bilateral hypoacusis. Her past medical history included hypercholesterolemia and 6 pregnancies, and she had been taking fenofibrate for 5 years without noticeable side-effects. On initial examination, she had widespread lesions involving the lateral aspects of the chest and the abdomen, the back and the proximal parts of the limbs. These lesions consisted of brown red papules clustering in large infiltrated sheets, sparing the body folds particularly on the back and the abdomen. The face was unaffected but the scalp showed inflammatory and squamous helmet-like lesions. The palms of the soles were slightly hyperkeratotic and the nails were longitudinally striated. The physical examination was otherwise unremarkable. The hypoacusis was evaluated and a diagnosis of recent cholesteatoma complicating a long-lasting presbyacusis was established.

Histological examination of a papule of the back showed hyperkeratosis with focal parakeratosis, acanthosis and a dermal perivascular infiltration of mononucleated cells with some eosinophils. Lesions of the scalp were similar, with some additional features such as non-necrotic perifollicular epithelioid granuloma with multinucleate giant cells. Immunostaining of the dermal infiltrate showed 50% of T lymphocytes, and about 20% of protein S 100-positive cells.

Erythrocyte sedimentation rate, complete blood cell count, comprehensive chemical panel, antinuclear and anti-DNA antibodies, urinalysis, serum IgE, serological tests for HIV, HTLV1, herpes virus, adenovirus and EBV were normal, negative or non-contributing. Conversely, some abnormal laboratory findings were noted: eosinophilia (1.710 10^9/L) contrasting with lymphopenia (1.0 10^9/L) with a mildly low T4/T8 ratio (1.38; normal value: 1.5–2.5); high serum levels of aldolase (9.3 IU/l; normal value: 1–7.6), LDH (791 IU/l; normal value: 200–600), TGO (52 IU/l; normal value: 5–40), γGT (142 IU/l; normal value: 5–80) and β_2 microglobulin (4.35; normal value: 1–3). Chest X-ray, abdominal ultrasound and brain, thorax and abdominal computed tomodensitometry were normal. Parasitological investigations, bone marrow examination and gastroscopy were not performed. Patch tests and phototesting were not considered because of the extent of the eruption.

The patient was first treated with local corticosteroids, with limited results. PUVA therapy was subsequently attempted on a 3 times a week basis during 6 weeks; this schedule resulted in a fast and complete clearing of the eruption after a cumulative dose of 22 J/cm^2. Furthermore, biological abnormalities returned to normal values 1 month after the beginning of the photochemotherapy. Regarding hypoacusis, local corticosteroids were prescribed with success before surgical treatment for cholesteatoma. No relapse was noted after an 18-month follow-up.

DISCUSSION

Ofuji's papuloerythroderma does not seem to be a unique clinical entity but conversely a reactive pattern to a number of triggering events, including medication intake and non-cutaneous neoplasias (1–5). However, it cannot be regarded as a true paraneoplastic syndrome. Our patient had been taking fibrate for 5 years; raised levels of LDH and β_2 microglobulin suggest the presence of a lymphoma, but no evidence could be found to support this diagnosis.

Photochemotherapy is a possible therapeutic mean, as it decreases the number of surface markers of Langerhans' cells in the skin. PUVA has before been used in 2 cases with success (3, 4) and in 2 others with exacerbation and side-effects, respectively (5, 6). Nevertheless, PUVA should perhaps be more frequently considered, as systemic steroids, the "classical" treatment, are often associated with side-effects in elderly people.

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