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LETTERS TO THE EDITOR

Lack of Effect of Cyclosporin A in Pityriasis Rubra Pilaris

Sir,

We treated a patient with longstanding erythroderma due to a therapy resistant Pityriasis Rubra Pilaris (PRP) type I (classical adult onset type) with cyclosporin A (CyA) (1).

PRP was diagnosed in this male patient at the age of 22. The skin disease started at the age of 21. From the age of 24 the patient was erythrodermic, apart from some islands of normal skin, with palmoplantar and subungual hyperkeratosis. Therapeutic attempts with photo-therapy (UVA+UVB, oral PUVA), methotrexate, danazol, topical aminonicotinamide and topical steroids were unsuccessful. The therapeutic effect of high-dose oral vitamin A treatment and long-term treatment with retinoic acid, etretinate and isotretinoin was limited (2).

In 1986, at the age of 37, treatment was started with CyA, 10 mg/kg body weight/day, administered orally in 2 daily doses. One day earlier etretinate (dosage 0.7 mg/kg/day) treatment was stopped. Blood pressure, hematologic values and renal and hepatic functions were carefully monitored. After one week the CyA dose was lowered to 8 mg/kg/day and after the second week to 6 mg/kg/day. CyA was given during a total of 18 weeks. The blood levels of CyA varied during this period between 350 and 920 ng/ml (Radio Immuno Assay Sandoz Kit Non Specific; advised therapeutic blood levels 400–800 ng/ml).

In the beginning there was no worsening as one might expect after withdrawal of etretinate, but a limited improvement: the erythema was reduced, especially on the legs, but the skin became drier. During the 16 weeks of CyA dosage of 6 mg/kg/day no further improvement was noted. On the contrary: the erythematous area on the legs increased again and there was a recurrence of the palmoplantar hyperkeratosis, which had been reduced by retinoid treatment. The ungual abnormalities did not change. During the last four weeks CyA was combined with etretinate (0.7 mg/kg/day). This resulted in limited improvement, i.e. desquamation of the palmoplantar hyperkeratosis and less dryness of the skin. Withdrawal of CyA did not change the clinical picture. Side-effects of the CyA treatment were limited to paraesthesias of hands and feet and initial abdominal discomfort.

There have been reports of the beneficial effect of CyA in a variety of skin diseases with an inflammatory component (3–5). Recently it has been reported that CyA is able to inhibit keratinocyte proliferation in vitro, which might be relevant for PRP, as epidermal cell kinetics are altered in PRP (6, 7). The absence of effect of CyA treatment in our patient with a very persistent erythroderma should not deter others from using CyA in PRP, especially in cases with a partial response to retinoid treatment and phototherapy.

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