this patient led to a strong cell destruction with consequent release of large amounts of uric acid. This cause of uraemia is well known in the treatment of patients with leukaemia, but seems not to have been described before in a patient with Mycosis fungoides. Hence if patients with Mycosis fungoides present with a pronounced skin affection, then caution should be taken during therapy to avoid this complication.

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Normalized Light Reactions in Mycosis Fungoides Patients after Complete Remission of Skin Lesions

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Abstract: Eleven mycosis fungoides patients were phototested on apparently normal skin before treatment with mechlorethamine, topically in 8 patients and with methoxsypsoralen followed by longwave ultraviolet light (PUVA) in 3 patients. Abnormal photosensitivity to UVB was seen before treatment in 4, to UVA in 4, and to visible light in one patient. The abnormal photosensitivity to all wavelength regions was normalized after complete remission of the cutaneous lesions. Two of the PUVA-treated patients demonstrated that special care should be taken during the initial phase of this treatment because of the light sensitivity, especially on lesional skin.

Key words: Mechlorethamine; Mycosis fungoides; Photochemotherapy; Photosensitivity

Abnormal photosensitivity is most pronounced on lesional skin in mycosis fungoides patients. Clinical evidence for this assertion is given by the observation that diseased skin in these patients very easily burned during photochemotherapy with psoralen and subsequent exposure to long-wave ultraviolet light (PUVA) (1, 3–6). Experimentally it is shown that the erythemal threshold in apparently normal skin is lowest close to cutaneous lesions (10). These observations are important, considering the increasing use of PUVA treatment in mycosis fungoides patients.

The purpose of the present study was to explore the possibility of normalized light reactions in mycosis fungoides patients after complete remission of skin lesions induced by mechlorethamine in 8 patients and by PUVA in 3 patients.

MATERIALS AND METHODS

The material consisted of 11 mycosis fungoides patients, 5 women and 6 men, age range 45–72 years. Five of them are reported in a previous paper (10). All patients had clinical cutaneous lesions of mycosis fungoides and the diagnosis was confirmed histologically, before treatment was started. Eight were classified as stage II and three as stage III.

Table I. *Doses corresponding to the MED before treatment and after complete clearance of all mycosis fungoides skin lesions*

Doses in mJ/cm²
HN_2 = mechlorethamine, CR = complete remission, – = not tested

Patient no.	Treatment	UVB		UVA		Visible light	
		Before	CR	Before	CR	Before	CR
1	HN_2	30	70	14 400	>15 600	>91 200	–
2	PUVA	30	>100	>16 400	>16 200	>51 000	>46 800
3	HN_2	70	110	>19 200	>18 000	>54 000	>48 000
4	HN_2	70	120	>31 200	>32 000	>49 200	>46 800
5	HN_2	55	90	>13 600	>9 900	>48 000	–
6	HN_2	60	110	15 300	>21 600	>48 200	>49 200
7	HN_2	33	120	12 600	>21 000	>80 800	>46 800
8	HN_2	60	200	>12 600	–	>46 800	–
9	PUVA	36	>120	>14 400	>18 000	>93 200	–
10	PUVA	60	>200	12 600	>18 400	25 200	>31 200
11	HN_2	50	80	>21 600	–	>49 200	–

All patients were phototested on apparently normal dorsal skin to determine the patient's individual minimal erythemal dose (MED). Light tests were performed before the treatment was started and after achieving complete remission of all skin lesions. The light testing procedure and the filters used are recorded elsewhere (10).

Eight patients were treated with mechlorethamine topically, as described elsewhere (8). Three were treated with photochemotherapy consisting of oral 8-methoxsalen, approximately in doses of 0.6 mg/kg body weight, 2 hours before irradiation with long-wave ultraviolet light (PUVA). The PUVA therapy was given according to our standard procedure for treating psoriatic patients. This treatment procedure is also discussed in detail elsewhere (7). One patient (no. 2) developed erythema following the first exposures to an UVA dose as low as 1.0 J/cm². Especially lesional skin was burned. The UVA exposure dose was therefore lowered to 0.5 J/cm² and then cautiously increased up to 4.0 J/cm². This rather low UVA dose, which was considered to be just under the erythemal threshold, was sufficient to clear all his skin lesions. Patient no. 10 also got a burning sensation and soreness on lesional skin during the first 2 weeks of treatment following UVA doses in the range 1.0–2.0 J/cm². However, it was possible to increase the UVA dose up to 6.0 J/cm² within 2 months. This dose was then well tolerated by the patient without any troublesome side effects.

RESULTS

The results of phototesting in 11 mycosis fungoides patients on apparently normal skin on the back before and after treatment are summarized in Table I. Before topical treatment with mechlorethamine or PUVA therapy, the erythemal threshold to UVB (filter 310) was under the lower limit for healthy volunteers for 4 patients (nos. 1, 2, 7, and 9); the doses being respectively 30, 30, 33 and 36 J/cm², while the lower limit for healthy volunteers in our laboratory is 50 mJ/cm². Irradiation though the 310 filter showed reactions within the normal control range after complete remission of all skin lesions.

Four patients (nos. 1, 6, 7 and 10) showed abnormal photosensitivity to UVA (filter 360) following doses of 12.6–15.3 J/cm² before treatment was started. None of the controls reacted to the tested doses 17–26 J/cm². After clearance of the mycosis fungoides skin lesions, none showed any reaction to the tested doses, as shown in Table I.

One patient (no. 10) reacted to visible light with erythema before treatment, whereas none of the normal controls did so. After healing of the skin lesions, no reaction was seen after approximately the same dose.

In conclusion, we noticed that the abnormal photosensitivity to UVB, UVA and visible light as demonstrated before whole body treatment, was normalized after clearance of all skin lesions.

DISCUSSION

The erythemal threshold is lowered in apparently normal skin of some mycosis fungoides patients over a wide wavelength range, including UVB, UVA and in a few cases also extending into the visible regions (9). The photosensitive reactions are most abnormal on the border of skin lesions (10). A practical consequence of the low erythemal threshold is that caution must be taken when treat-

ing these patients with photochemotherapy, and the UVA doses used for PUVA therapy should for quite a number of mycosis fungoides patients be lower than for psoriatics. However, remission can well be induced with low UVA doses, as demonstrated by 2 of the patients in this study, without troublesome side effects.

By treating cutaneous mycosis fungoides lesions to complete remission, the most photosensitive areas are thus removed. However, even in apparently normal skin which is treated simultaneously the light reactions are normalized. Following PUVA therapy this could be ascribed to the melanin protection and increased thickness of the epidermis, especially of stratum corneum. This photoprotective mechanism has already been shown to be effective in the treatment of a photodermatosis such as polymorphic light eruption (2). Hyperpigmentation occurs commonly following topical application of mechlorethamine (8). It occurs in the absence of any sign of cutaneous irritation or inflammation. The mechanism of the tanned skin following use of topical mechlorethamine is thus unknown. Nevertheless, the tan induced by mechlorethamine could certainly contribute to the normalized light reactions.

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Association between Psoriasis and the α_1-Antitrypsin Deficiency Gene Z

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Abstract. In a series of 72 psoriatic patients the frequency of individuals carrying the α_1-AT deficiency gene Z was found to be significantly ($p<0.001$) increased.

Key words: Psoriasis; Alpha-1-antitrypsin types; Disease association

More than two dozen electrophoretically distinct variants of α_1-antitrypsin (α_1-AT), controlled by a series of autosomal alleles at a locus Pi have been described (6). By isoelectric focusing it has been possible to distinguish subtypes of the common variant M, viz. M_1, M_2 (9) and M_3 (8). Three rare genes, Z, 0 (null) and Duarte, are associated with a very low level of α_1-AT in serum.

Individuals homozygous for the Z gene have 10–15% of the normal serum level. Homozygotes for the null allele probably lack α_1-AT altogether. Homozygosity for the Duarte gene results in about 10% of the normal serum level of α_1-AT. The S gene is associated with a moderate decrease of α_1-AT. Homozygotes for this gene have 60% of the normal serum level.

A decreased level of α_1-AT has been found to be associated with chronic obstructive lung disease (9), juvenile liver cirrhosis (10), respiratory distress syndromes (5), malignant hepatoma (4) and non-atopic asthma in children (1).