Acta Derm Venereol (Stockh) 1995; 75: 72–74

Photosensitivity Dermatitis/Actinic Reticuloid Syndrome in an Irish Population: A Review and Some Unusual Features

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Nine patients with photosensitivity dermatitis and actinic reticuloid syndrome are reported. The rash affected light-exposed skin only in 7 patients and extended to covered sites in 2. Seven were sensitive to UVB, UVA and visible light, one to UVA and one to visible light only. Two patients had significant clinical improvement in spite of both having suffered severe disease previously. Another patient remains exquisitely light-sensitive after 20 years and has developed two squamous cell carcinomas, one of which metastasized to local lymph nodes, and two keratoacanthomas on light-exposed skin.

(Accepted August 12, 1994.)

Acta Derm Venereol (Stockh) 1995; 75: 72–74.

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Photosensitivity dermatitis and actinic reticuloid syndrome (PD/AR), also referred to as chronic actinic dermatitis, is a chronic disabling disease which mainly affects middle-aged men with an outdoor occupation (1–3). The severity of the condition is proportional to the degree of abnormal response to ultraviolet (UV) radiation and visible light (1). The rash may vary from a localised dermatitis of exposed skin to a more generalised eruption. In patients with AR the clinical features and histology resemble a reticulosis (4). PD/AR is uncommon and is estimated to affect 1 in 6,000 of the population in the Tayside region of Scotland (2). Although it is a chronic disease the majority of patients do well with treatment (5). Over the past 10 years we have seen 9 patients with PD/AR in our department. This paper comments on this group with particular regard to their outcome and some unusual clinical features.

MATERIALS AND METHODS

Only patients fulfilling the following criteria were included in this review: (1) persistent photosensitivity with an eruption on sun-exposed areas; (2) histological criteria consistent with PD/AR varying from a mild perivascular to a dense dermal lympho-histiocytic infiltrate with atypical lymphocytes; and (3) positive phototest results. Patients with the first two criteria but with a normal phototesting pattern were not included. The information was obtained from case notes, patient consultations and results of patch testing and phototesting. Nine patients were studied. All were male and aged between 35 and 68 years, median 61. The history of symptoms at presentation ranged from 7 months to 27 years and, with follow-up, total disease duration was 2–28 years, median 12. None of the patients were taking photosensitising medication. Five patients had an outdoor occupation, and the other 4 with an indoor occupation had an outdoor hobby. Seven had regular contact with plants and 2 with cement.

In 5 patients (cases 1, 3, 4, 6 & 9) the rash initially had a seasonal variation with summer onset and winter remission but became persistent after 3 to 25 years. In the remaining 4 patients the rash was persistent from the outset. It began on light-exposed skin in all 9 and extended to non-light-exposed skin, causing a generalised rash in 2 (cases 8 & 6) after 2 and 10 years. Typically, the rash was pruritic or burning and consisted of erythematous papules, vesicles, indurated plaques and occasional nodules. Two patients had lymphadenopathy. In one (case 6), this was dermatopathic; in the other (case 3) there were two cervical nodes secondary to a squamous cell carcinoma (SCC) on the neck. No patient had hepatomegaly or splenomegaly.

Patch tests were carried out to the European standard battery, compositae oleoresins, fragrances and allied compounds in all cases, and patients who were thought to be clinically allergic to sunscreens were patch-tested to a sunscreen battery. Photopatch testing was not performed. Minimal erythema dose (MED) phototesting was conducted in 8 patients at the Photobiology Unit in Ninewells Hospital, Dundee. Seven of these 8 (cases 1–7) had diffraction grating monochromator testing with wavebands 305 ± 5, 335 ± 30, 365 ± 30, 400 ± 30, and 430 ± 30 nm. Four patients (cases 1, 4, 5 & 8) had solar simulator testing; 3 of these were in addition to monochromator testing. Results were read at 24 h. The remaining patient (case 9) had monochromator testing in another unit with wavebands 300 ± 5, 320 ± 10, 370 ± 25, 400 ± 25 nm. Results were read at 24 h.

RESULTS

Histology showed hyperkeratosis, focal parakeratosis and a lymphohistiocytic infiltrate in the papillary and upper reticular dermis in all cases. The infiltrate was predominantly perivascular in 6 patients, and eosinophils were scattered throughout the dermis also in 6. Variation in the shape and size of nuclei within the lymphoid cells was present in 3. Full blood count, serum biochemistry and porphyrin screen were normal in all 9. Antinuclear factor was negative in all cases. With the exception of one patient, in whom it was transiently elevated at 71 mm/h, the erythrocyte sedimentation rate was normal. Patch testing was positive to one or more allergens in all 9 patients, predominantly to potassium dichromate, compositae and fragrances (Table I). The results of initial monochromator testing on 7 patients are shown in Table II. The other patient who had monochromator testing in another unit (case 9) was sensitive to UVB, UVA and visible light. The patient who had solar simulator testing alone (case 8) was sensitive to the whole spectrum and whole spectrum minus UVB. Overall, 7 showed an abnormal response to UVB, UVA and visible light, one (case 7) to UVA only and one (case 5) to visible light only.

All patients were treated with antigen and light avoidance, sunscreens, emollients and topical steroids. Five required intermittent courses of oral steroids during the early phase of the disease. Antihistamines and antibiotics were administered when necessary. Beta-carotene was used unsuccessfully in 2 cases. Overall, 7 patients have done well on treatment: 4 are well controlled on topical steroids (cases 2, 5, 6 & 9), one on azathioprine (case 7) and 2 have undergone complete clinical remission in spite of both having suffered severe disease previously; however, their phototesting remains abnormal (cases 1 and 4). Of the remaining 2, one was poorly controlled over a

 ISSN 0001-5555

Table I. *Clinico-pathological features and patch test results in patients with PD/AR*

Mod: moderate, +: positive patch test result defined as an erythematous/oedematous response at the site of application at 48, 72 or 96 h., 0: negative result, −: not tested

	Case No.								
	1	2	3	4	5	6	7	8	9
Age at onset (years)	62	68	41	49	36	51	35	59	37
Clinical severity at presentation	Severe	Mod	Severe	Severe	Mod	Mod	Severe	Mod	Mod
Histology	PD	AR	PD	PD	PD	PD	AR	PD	AR
Regular contact with plants	Yes	Yes	Yes	No	Yes	Yes	Yes	No	Yes
Regular contact with cement	Yes	No	No	No	No	No	Yes	No	No
Potassium dichromate	+	0	+	+	0	+	0	0	+
Compositae	0	+	+	+	+	0	+	+	0
Colophony	+	0	0	0	+	0	+	0	+
Rubber	+	0	+	0	+	+	0	0	0
Nickle	+	0	0	+	0	+	0	0	0
Turpentine	+	0	+	0	0	0	+	0	0
Sunscreens	0	−	+	+	+	−	−	−	−
Fragrances	+	0	+	0	+	0	+	+	0
Balsam of pine	+	0	+	0	0	0	+	0	0
Balsam of spruce	+	0	+	0	0	0	+	0	0
Preservatives	0	0	+	+	0	0	+	0	0
Wood tars	0	−	+	0	0	−	+	+	−
Topical steroids	+	−	−	−	−	−	−	−	−

3-year period until his death of unrelated causes (case 8). The last patient (case 3) remains exquisitely light-sensitive after 20 years, and phototesting continues to demonstrate persistent and severe reactions to UVB, UVA and visible light. Although he has adhered to a strict light avoidance programme, he has developed 2 rapidly growing SCCs (Fig. 1) and 2 keratoacanthomas (KAs) on light-exposed skin during the past 2 years. UVR mutation and DNA repair studies on fibroblast cultures in this case are normal.

DISCUSSION

Our group of PD/AR patients is similar to previous reports with regard to the sex and age distribution: all were middle-aged or elderly males and had an outdoor occupation or hobby (1, 2, 4–7). It has been shown that patients with PD/AR often have contact allergic sensitivities to compositae, fragrances and allied compounds and potassium dichromate (8–10), which are frequently encountered in outdoor work. Although the initial insult in PD/AR may be of a photoallergic or allergic contact nature, it is unclear whether this is so in most cases (8, 9, 11). While all our patients had positive patch tests to one or more allergens, those with allergies to a greater number of substances tended to have more severe clinical disease initially. However, there was no relationship between the number of substances that the patient tested positive to and the presence of AR on histology, or the outcome/course of the disease with therapy.

Initial phototesting with monochromator and/or solar simulator showed that most of our patients were sensitive to UVB, UVA and visible light. Approximately 10% of cases of PD/AR may have an action spectrum which does not enter the shorter UVB wavelengths (5), but isolated responses involving a narrow band of UVA or visible light, as occurred in 2 of our cases, seem a rare event. The width of the spectrum of photosensitivity does not appear to correlate with the degree of lymphoma-like histological features characteristic of AR (13), and our group were

Table II. *Monochromator testing results in 7 patients with PD/AR**

Wavelength (nm)	Lowest normal population value	Minimal erythema response (mJ/cm²)						
		Case No.						
		1	2	3	4	5	6	7
305	22	5.6	10	1.8	5.6	39	2.7	47
335	1800	180	1000	180	1200	1800	390	1000
365	8200	2200	3900	560	3300	10000	1800	12000
400	47000	3900	12000	10000	33000	47000	18000	47000
430	82000	82000	Not tested	39000	82000	56000	10000	82000

* Monochromator testing on these 7 patients was carried out in Ninewells Hospital, Dundee. Case 8 had solar simulator testing alone and was sensitive to the whole spectrum. Case 9 had monochromator at another institution to wavebands 300 ± 5, 320 ± 10, 370 ± 25 and 400 ± 25 nm and gave abnormal responses to all these wavebands.

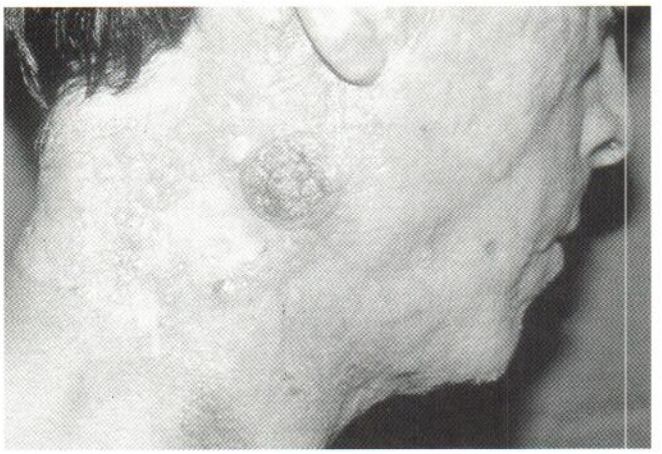

Fig. 1. Large squamous cell carcinoma on the neck of a patient with photosensitivity dermatitis/actinic reticuloid, which had metastasized to local lymph nodes.

similar in this regard. Phototesting remained abnormal in each of the 4 patients who had it repeated over periods of one to 17 years, including the 2 who underwent clinical remission. This observation has been noted previously and raises the possibility of an exposed site desensitization process (2). Of particular interest is the gentleman who after 20 years with PD/AR continues to demonstrate persistent and severe reactions to UVB, UVA and visible light.

While the outcome varies between patients with PD/AR, the majority do well on treatment (5). Our cases were similar in this regard in spite of the fact that the limited choice of adequate UVA and visible wavelength sunscreens poses problems for cases whose sensitivity extends into this part of the spectrum (13). Our 2 patients in clinical remission had previously suffered several years of poor control and initially had more severe disease than others in whom the PD/AR continues to be active. Azathioprine is known to be an effective therapy for PD/AR (14) and gave an excellent result in one of our patients. In most of our cases, however, apart from initial courses of oral steroids, systemic treatment has not been necessary. Overall, the improvement in the majority can be attributed to regular outpatient follow-up, early hospital admissions into light-protected rooms and patient advice and education.

There have been no previous reports of SCC arising in association with PD/AR. The development of SCCs and KAs on light-exposed skin in one patient after 20 years of PD/AR is very unusual. This man's light sensitivity is so severe that he rarely ventures out in daylight hours. The reason for the rapid emergence of these tumours in our patient is unclear: UV mutation and DNA repair studies on cultured fibroblasts are normal, and the patient has never been exposed to azathioprine, cyclosporin or PUVA, all of which have been used for PD/AR and which may be carcinogenic (14–16). Although five cases of lymphoma have been reported in association with this disease (4, 17 for ref.), a flow cytometric study of actinic reticuloid produced no evidence that it is a premalignant condition (18). One possible explanation for the tumours in our patient is that the persistent inflammation of light-exposed skin might allow these to develop in a way which is analogous to the pathogenesis of SCCs in lupus erythematosus, chronic ulcers and osteomyelitis sinuses (19, 20).

ACKNOWLEDGEMENTS

We wish to thank Dr. J. Ferguson Ninewells Hospital Dundee, for advice, reviewing the manuscript and phototesting cases 1–8. Our thanks also to Dr. Gillian Murphy, Mater Hospital, Dublin, for phototesting case 9 and to Dr. C. Arlett, MRC Cell Mutation Unit Brighton, for performing UV mutation and DNA repair studies.

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