



University of Dundee

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Macfarlane, L; Hawkey, S; Naasan, H; Ibbotson, S

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Characteristics of actinic prurigo in Scotland: 24 cases seen between 2001 –2015

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Dear Editor, Actinic prurigo (AP) is a chronic photosensitivity disease, which is mainly described in Amerindians and rarely in Caucasians¹⁻⁶. The pathogenesis of AP is unclear, although familial occurrence is often seen, with a strong association with HLA DR4 and in particular HLA DRB1*0407, which is present only rarely in the general population⁷.

Classically AP is reported to develop in childhood and be characterised by perennial symptoms, involvement of distal nose, lower eyelid, conjunctivae, lips and covered sites and be associated with abnormal phototesting^{1,8}. The condition is considered to spontaneously resolve in adolescence^{1,2,8} although late-onset and persistent AP may occur^{2,4,5,8,9,10}.

The Scottish Photobiology Service (SPS), based in Dundee, investigates patients with suspected photosensitivity across Scotland. In order to increase our understanding of this rare condition in Caucasian populations, we undertook a retrospective study with Caldicott guardian approval. We included all patients diagnosed and/or investigated with AP in the SPS between April 2001 and March 2015. Information on patient and disease characteristics, investigation findings and management were retrieved from the SPS database and from written case records.

Twenty-four patients were identified with AP (0.7% of 3463 patients seen in the SPS during the fourteen-year period). Eighteen (75%) were female and 6 (25%) male. Most (20 of 21 (95%); one patient was Asian and three were unknown) were Caucasian. Eighteen (75%) patients had

disease onset at <10 years, with a median age of onset of five (range 1.5-41) years. The median time between onset and diagnosis was six (<1-52) years; with broad age distribution at time of diagnosis. Fourteen (58%) patients had an atopic history and four (17%) had a history of contact allergies from previous patch testing. Eleven (46%) patients had a positive family history of photosensitivity (of unknown type) and four (17%) had a family history of atopy.

Most patients (n=18; 75%) were aware of an association with sunlight and (21; 88%) had perennial disease activity. Of those with perennial symptoms, 14 (67%) had summertime worsening. Of the patients who were aware of the association with sunlight, 11 (61%) noticed provocation of rash with less than 10 minutes of sun exposure. Fourteen (78%) patients described worsening of symptoms with light through window glass, eight (44%) with light through clothing and three (17%) with artificial lighting.

Most patients (n=23; 96%) had facial involvement. The nasal tip was affected in 14 (58%), lips in 11 (46%), ears in eight (33%) and conjunctivae in five (21%). Other affected sites included dorsal hands and forearms (15; 63%), neck (7; 29%) and legs (3; 13%). The main clinical features were itch (20; 83%), papules (15; 63%), erythema (11; 46%), vesicles (9; 38%) and scarring (8; 33%) (Figure 1). Ten (42%) patients had involvement of covered sites. Dermatology life quality index questionnaire (DLQI) scores were available in 9 patients; median DLQI 12 (8-27) and 6 considered that their condition had a 'very large adverse effect on their life'.

Monochromator phototesting showed a normal action spectrum in nine (38%) cases (Table 1). Nine (38%) had abnormal sensitivity to UVA, UVB and visible wavebands and all the remainder showed at least UVA sensitivity. One patient had normal monochromator phototesting on six occasions over five years before most recent testing in 2015 showed broadband UVB and UVA sensitivity (Figure 1). UVA provocation testing in 21 cases showed papular reactions in 13 (62%)

and abnormal erythema in six (29%). The median narrowband UVB (TL-01) MED was 0.128 (range <0.025 - >0.39) J/cm²; with abnormally low MEDs (<0.025 J/cm²) in only four cases. HLA typing was undertaken in 20 patients: 18 (90%) were HLA DR4 positive; of whom 12 (67%) were DRB1 *0407 positive.

Seventeen patients were patch tested, with 10 (58%) being positive to a range of common allergens. Fifteen patients were photopatch tested, identifying two (13%) with sunscreen allergy. IgE levels were raised (>100 kU/L) in eight of 11 (73%). Histopathology, lupus and porphyria screening were non-specific and negative respectively.

General measures of photoprotection, including sunscreens, topical emollients and corticosteroid application and sedative anti-histamine use for symptomatic relief of pruritus were advised. Treatment responses and data on prognosis were limited as most patients were followed up in their local dermatology centre. Thirteen (65%) described significant benefit with high SPF sunscreens; three (15%) slight benefit and four (20%) no improvement in symptoms (data missing for four patients). Five patients required oral corticosteroids; three received azathioprine (two of whom obtained no benefit). Three patients were offered thalidomide but all declined. Ten underwent narrowband UVB desensitisation, with efficacy in four. One of three patients found UVA desensitisation to be of benefit. Three received PUVA, with efficacy in two. Over time, five had stable disease, five symptomatically improved and 5 subjectively worsened. Of 10 patients who were followed up objectively by phototesting, 6 (60%) showed improvement in photosensitivity.

This is the largest recent report of British Caucasian patients with AP, who represented only 0.7% of all patients seen in a specialised photodiagnostic unit. This contrasts with previous studies of Amerindian populations where the prevalence was much higher. In Asian and Mediterranean

studies, 2-5% of patients investigated in photodiagnostic units had AP¹¹⁻¹³. Thus, our data highlight the rarity of this disease in a British Caucasian population.

Our study confirms the female predominance of AP^{1,2,4} although contrasts with Asian populations where it occurs more commonly in males, with late-onset, persistent disease^{9,10,12}. We highlight the relatively young age of onset for most Caucasian patients with AP, consistent with previous observations in Caucasian and Amerindian patients with AP^{1,8}. However, we also show the potential for the disease to first manifest in adulthood, highlighting the importance of considering this diagnosis in older patients. Other Caucasian case series showed a mean age of onset of 14 years^{3,4}. However, our case-series highlights an additional difference from AP in Asian populations, where onset in middle age is typical^{9,10,12}.

The median time between disease onset and diagnosis was six years. This delay could be due to several factors: disease rarity; the perennial nature of photosensitivity, making it more difficult to identify a link with sun exposure; as well as heterogeneity in disease presentation. For example, 88% of patients had perennial disease and 25% were unaware of an association with sunlight exposure. Although facial involvement occurred in 96%, classical sites of lip, conjunctival and distal nose involvement were only seen in 58%, 46% and 21% of patients respectively. Additionally, 42% of patients had covered site involvement. Thus, the diagnosis may be challenging. Diagnostic delay was also reported in an Australian study, where mean age of disease onset was 14 years, but mean age at diagnosis was 25 years⁴. This highlights the importance of awareness of this rare condition in order to ensure prompt referral and investigation.

Although we need to be guarded with respect to data interpretation, 58% of patients had a history of atopic disease, 73% had elevated total IgE levels and 58% had positive patch testing, suggestive of an association with atopy^{1,3} and contact allergy, which needs further investigation.

Monochromator phototesting showed abnormal photosensitivity in most cases. However, 38% of patients had a normal action spectrum. This appears higher than the report of Addo *et al.*, where 16% had normal responses¹, although is in keeping with Crouch *et al.*, where 40% had normal responses⁴. This highlights a further diagnostic challenge, illustrated in particular by one patient who had normal monochromator phototesting on six occasions over five years before abnormal broadband UVA and UVB photosensitivity was ultimately observed (Figure 1). Hence, repeat phototesting is indicated if there is strong suspicion of AP. Most patients with abnormal phototesting demonstrated broadband photosensitivity, with UVA wavelengths implicated in all cases. Iterative broadband UVA provocation testing was abnormal in 90%, proving helpful in diagnosis, including the patient in whom monochromator phototesting was initially normal. Interestingly narrowband UVB MEDs were normal in most patients, supporting the use of phototherapy.

Only 46% of patients reported a family history of photosensitivity, which is consistent with reports in Caucasian patients but lower than that seen in Amerindian populations^{1,8}. Our findings confirm the diagnostic utility of HLA typing in Caucasian patients, as 90% of patients were HLA DR4 positive and HLA DRB1*0407 was seen in 60% of those who were HLA tested (67% of those who were HLA DR4 positive)¹⁴.

Although AP is typically considered to improve/resolve by early adulthood^{1,2,8}, persistent chronic disease is reported, particularly in Asian patients, but also in Australian cases^{4,8-10,12}. Our Caucasian patients showed disease persistence in 10 of 15 (66%) cases where follow-up data were available, with six of 10 patients followed up with phototesting showing objective improvement in photosensitivity. Furthermore, 42% of our cohort were not diagnosed until >21 years, with 25% being >40 years old. Thus, AP may persist into mid-late adulthood, and may not

resolve in teenage years in most Caucasian patients. However, follow-up data were limited and should not be over-interpreted.

Most (80%) patients reported benefit with high SPF sunscreens, reinforcing the importance of perennial photoprotection. Fourteen patients reported abnormal sensitivity to light through window glass and advice on the use of UV-protective film can be useful. Three patients were aware of deterioration after artificial lighting exposure and advice regarding light bulb choices may be beneficial. Narrowband UVB desensitisation was beneficial in 40% and may be an important treatment option. Very few patients required systemic immunosuppression or thalidomide⁴ as most were managed by conservative approaches and phototherapy, which is safer in young patients.

For patients in whom DLQI scores were available, a median score of 12 indicates the serious adverse impact of this condition on quality of life, with a quarter of patients describing 'a very large negative effect on their life'. The psychological impact of chronic photosensitivity is increasingly recognised and our findings are in keeping with this significant level of adverse impact of disease¹⁵.

In summary, we have presented the characteristics of the largest recent case-series of AP, seen in the Scottish Photobiology Service over a 14-year period. We highlight the rarity of this disease in Caucasians and that it can present at any age, may not have the typical disease characteristics and may persist, leading to delays in diagnosis. We highlight the importance of considering a diagnosis of AP, even if photosensitivity is not immediately apparent. This should facilitate earlier diagnosis and appropriate management of a condition that can otherwise have a profound negative impact on quality of life.

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L Macfarlane, S Hawkey*, H Naasan, S Ibbotson

Photobiology Unit, Department of Dermatology,
University of Dundee, Ninewells Hospital & Medical School,
Dundee, United Kingdom, DD1 9SY

*5th year medical student, School of Medicine, University of Dundee

Correspondence:

Dr Sally Ibbotson

Tel: 01382 383499

Email: s.h.ibbotson@dundee.ac.uk

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