

A Report of Two Cases of Pigmented Purpuric Dermatoses Treated with PUVA Therapy

W. K. WONG and K. V. RATNAM

National Skin Centre, Singapore

This is a report of 2 patients with pigmented purpuric dermatosis treated successfully with Psoralen photochemotherapy (PUVA). Treatment for pigmented purpuric dermatosis is generally unsatisfactory, and we believe this to be the first report of successful response of this condition to PUVA. Key words: *Purpura pigmentosa chronica*; Photochemotherapy.

(Accepted August 1, 1990.)

Acta Derm Venereol (Stockh) 1991; 71: 68–70.

W. K. Wong, National skin Centre, 1 Mandalay Road, Singapore 1130.

Pigmented purpuric dermatoses (PPD) refer to a group of conditions characterized histologically by a capillaritis of unknown cause (1). Four distinct entities have been described under various eponyms and clinically they can be distinguished from each other. Although these conditions have been known for some time, their treatment has not been entirely successful. Various modalities have been tried, with limited success. As phototherapy has been used in the treatment of pityriasis lichenoides with partial success (2), and since both conditions may share a common problem at the vascular level, phototherapy was tried successfully on 2 patients with PPD. This report provides yet another modality of treatment for this benign but cosmetically unattractive problem.

CASE REPORTS

Case 1

A 37-year-old male Chinese welder presented with asymptomatic red and brown spots on both his legs since December 1986. There was no history of an antecedent infection but he had a history of taking Valium on and off for insomnia. Examination showed patches of brown hyperpigmentation with petechial and purpuric spots present mainly on the legs. In some areas, these patches were confluent. The rest of the examination was normal. The following laboratory tests proved normal or negative: FBC including platelet, ESR, LFT, ANA, ASOT and urinalysis. A clinical diagnosis of Schamberg's disease was made as the patient refused a skin biopsy.

Because the condition had progressed at follow-up, prednisolone was added at an initial dose of 20 mg daily to the

topical betamethasone 0.05% cream that he was using. There was an initial improvement but when the dose of prednisolone was reduced to 5 mg daily there was clinical relapse.

By this time the patient agreed to a skin biopsy which showed a superficial perivascular infiltrate of lymphocytes and histiocytes with extravasation of erythrocytes and presence of siderophages. This histology was consistent with Schamberg's disease. Direct immunofluorescence studies proved negative. Patch tests to the standard battery and to textile dyes and resins were negative.

A trial of topical steroids alone was just as disappointing. So UVB was started in October 1987 and it was not found to be useful after 26 treatment sessions.

As the patient still had extensive PPD, PUVA therapy was instituted in January 1988 three times weekly with 8-methoxypsoralen at 0.6 mg/kg body weight 2 h before UVA irradiation. The starting dose of UVA was at 2 J/cm² because of the pigmentation from the previous UVB therapy and this was increased by 1 J/cm² at each treatment. The maximum UVA dose used was 12 J/cm². Topical steroids were stopped when PUVA therapy commenced. After 4 weeks of PUVA (total 99 J) the lesions cleared. At review 6 weeks later the patient was still free of lesions.

Case 2

A 55-year-old Chinese lady was first seen in July 1989 for a 5-month history of a mildly pruritic reddish rash which subsequently healed, leaving pigmentation. There was no precedent history of drugs, contactants or infections. On examination the lesions were mainly on the lower limbs. They consisted of patches of brown hyperpigmentation with purpuric/petechial spots. Some were annular in configuration and there were also some erythematous papules (Fig. 1).

Skin biopsy of a papular lesion on the leg showed a lichenoid infiltrate of lymphocytes and histiocytes with extravasation of erythrocytes. There was also orthokeratosis and spongiosis of the epidermis. Pigment-laden macrophages staining positively for iron were present in the papillary dermis. This was consistent with PPD of Gougerot and Blum type.

The patient was treated with topical betamethasone 0.05% cream. As she was not responding to the topical therapy, the patient was put on a course of PUVA, initially on her left leg only, three times weekly. She was given 8-methoxypsoralen at 0.6 mg/kg body weight 2 h before UVA irradiation (Dermaray, Eisai). The initial UVA dose was 0.5 J/cm² and this was increased by 0.5 J/cm² at each session. The maximum dose given was 6 J/cm². Topical steroids were stopped at the start of PUVA therapy.

On review at 4 weeks there was marked improvement on the treated leg but the right leg remained unchanged. With



Fig. 1. Case 2. Lower limbs before PUVA.



Fig. 2. Case 2. Lower limbs after 8 weeks of PUVA therapy.

this encouraging result, the other limb was treated similarly and clearance of the lesions on both limbs was achieved at the end of another 4 weeks (total UVA right limb 33.5 J and the left limb 96 J) (Fig. 2). On review she was free of lesions at the end of 6 weeks post-treatment.

DISCUSSION

Four distinct clinical entities have been described under this condition of pigmented purpuric dermatoses: Schamberg's disease or progressive pigmented purpuric dermatosis; pigmented purpuric lichenoid dermatosis of Gougerot and Blum; purpura annularis telangiectodes or Majocchi's disease and eczematid-like purpura of Doucas and Kapetanakis (1). A localized variety, lichen aureus is sometimes added to this group.

The basic process is thought to be a capillaritis limited to the upper dermis (3). In early lesions the endothelial cells of the capillaries may be swollen with a perivascular infiltrate of lymphocytes and histiocytes. The infiltrate may be sparse (as in Schamberg's disease) or it may be lichenoid (as in PPD of

Gougerot and Blum) (4). Extravasation of red blood cells is usually seen. In older lesions, haemosiderin and siderophages are present in varying amounts. Slight epidermal hyperplasia, spongiosis and focal parakeratosis may be present.

There are no specific laboratory findings characteristic of this group of conditions. One must exclude drugs, the most notable of which is carbromal and other sedatives, and contact allergy such as to dyes, fabric material or rubber in the clothing (5). Other causes like infections, allergy to infections and delayed hypersensitivity reactions have been suggested by various authors (6). Fishman suggested that there might be a link with alcohol consumption (7). Immunological processes may be involved, as direct immunofluorescence studies show depositions of C3 or C1q with or without immunoglobulins, and of fibrin in the papillary vessels (8).

The treatment of PPD has not been entirely satisfactory. Topical and systemic steroids have been the mainstay of therapy (6,7,9). Patient no. 1 was

treated with both topical and systemic steroids but he relapsed when the oral prednisolone was reduced to 5 mg daily. An alternative treatment was sought as his condition was progressing. UVB proved to be ineffective in his case but PUVA cleared his lesions after 4 weeks. Patient no. 2 was given PUVA initially on one limb and then both limbs were irradiated. Clearance was achieved again at about 4 weeks (right leg 12 treatments, total 33.5 J; and left leg 22 treatments, total 96 J because of the initial 4 weeks of treatment alone). Neither patient had received any other treatment (including vitamin C) when PUVA commenced.

The mechanism of action of PUVA may be one of immune modulation if the etiology of PPD is indeed due to a combination of infection and allergy. Based on recent immunohistologic findings, Aiba & Tagami in their study of 8 patients with Shamberg's disease think that the Langerhans' cells may play a role in the pathogenesis of the condition (10). Perhaps PUVA may be acting partially through its effect on the Langerhans' cells (11) but its other effects remain speculative. We believe that improvement was not due just to pigmentation of the skin alone, as UVB was unsuccessful in Case 1. Moreover, it was unlikely to be a spontaneous remission because in Case 2 the untreated leg remained unchanged whereas the treated one improved after 4 weeks of PUVA.

As far as we aware, this is the first successful use of PUVA to treat PPD.

REFERENCES

1. Rook A. Textbook of dermatology, 4th edn. Oxford: Blackwell Scientific Publications, 1986; 1116-1119.
2. Powell FC, Muller SA. Psoralens and ultraviolet. A therapy of pityriasis lichenoides. *J Am Acad Dermatol* 1984; 10: 59-64.
3. Lever WF, Schaumburg-Lever G. Histopathology of skin, 6th edn. Philadelphia: JB Lippincott Co, 1983; 173-174.
4. Ackerman AB. Histologic diagnosis of inflammatory skin diseases. Philadelphia: Lea & Febiger, 1978; 172-173, 211.
5. Fisher AA. Contact dermatitis, 3rd edn., Philadelphia: Lea & Febiger, 1986; 102-103.
6. Wong RC, et al. Pigmented purpuric lichenoid dermatitis of Gougerot-Blum mimicking Kaposi's sarcoma. *Cutis* 1983; 31 (4): 406-408, 410.
7. Fishman HC. Pigmented purpuric lichenoid dermatitis of Gougerot-Blum. *Cutis* 1982; 29 (3): 260-261, 264.
8. Iwatsuki K, et al. Immunofluorescence study in purpura pigmentosa chronica. *Acta Derm Venereol (Stockh)* 1980; 60: 341-345.
9. Samitz MH. Cutaneous disorders of the lower extremities, 2nd edn., Philadelphia: JB Lippincott, 1981; 76.
10. Aiba S, Tagami H. Immunohistologic studies in Shamberg's disease. Evidence for cellular immune reaction in lesional skin. *Arch Dermatol* 1988; 1058-1062.
11. Friedmann PS. Disappearance of epidermal Langerhans cells during PUVA therapy. *Br J Dermatol* 1981; 105: 219-221.