Prurigo Pigmentosa : A Report of 5 Cases with a Review of the Korean Literature

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Prurigo pigmentosa(PP) is a rare dermatosis with an unknown cause characterized by intensely pruritic papules that resolve with reticulate pigmentation. It is a disease entitity commonly reported in Japan but seemingly rare in other parts of the world. We report 5 cases that clinically and histopathologically fulfilled the characterisitics of PP and were treated with dapsone resulting in significant clinical improvement. (Ann Dermatol 10:(2) 132~137, 1998)

Key Words : Prurigo pigmentosa

Prurigo pigmentosa was first described by Nagashima[1] in 1971 and is characterized by a pruritic erythematous papular eruption usually involving the trunk and nape which resolves leaving a reticulated pigmentation. It tends to occur in spring or summer and young adolescent females are mainly affected. Although over 200 cases have been reported in Japan[2], it seems to be rare elsewhere. In Korea, Hong[4] first reported PP in 1988, and since then a few more cases have been reported[19-20]. Herein we review the literature and report 5 cases of PP that responded to dapsone therapy.

CASE REPORTS

Case 1

A 25-year-old woman presented with pruritic papules on the anterior chest and upper back of 15 days duration. She complained of having the same kind of lesions in an on-and-off manner for 6 years. Her medical status was good and she did not recall taking any medications or being particularly sun-exposed. A physical examination revealed edematous erythematous papules and plaques on the upper back, nape, and anterior chest area with scattered focuses of brown pigmentation.

Her family and past medical history were not significant and the results of the laboratory tests including a complete blood count, urinalysis, serum electrolytes, and liver function tests were within normal limits and antinuclear antibodies were negative. A fungus study performed on the upper back was negative.

On histopathological examination, the epidermis showed spongiosis, spongiotic vesicles, focal hydropic changes, and RBC trapping. The dermis showed a mild infiltration of mononuclear cells around the blood vessels and extravasation of RBC was observed in the papillary dermis.

Dapsone therapy at a dosage of 50 mg daily was started and after two weeks the erythematous papules disappeared but the pigmentation remained. The dosage was reduced to 25 mg daily and was continued for two more weeks which resulted in lightening of the pigmentation.

Case 2

An 18-year-old female visited our dermatology department because of a pruritic erythematous eruption involving her back area. The lesions had persisted in an intermittent manner for 6 months. She claimed to have had recurrences from April

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Table 1. Summary of 5 reported cases of prurigo pigmentosa in Korea compared with our present 5 cases

Case	Sex/Age	Localization	Duration	Therapy
1[4]	F/27	upper back chest	1 year	dapsone
2[4]	F/20	back chest	1 month	antihistamines topical steroid
3[19]	M/20	back	1	month dapsone 50mg/day for 2 weeks
4[20]	F/21	nape back chest	remote-3 recur each summer	years dapsone 100mg/day for 2 weeks
5[20]	F/23	back	1	month dapsone 50mg/day for 1 week dapsone 100mg/day for 1 week
6*	F/25	upper back nape chest	remote-6 years recent-15 days	dapsone 50mg/day for 2 weeks
7*	F/18	back	remote-3 years recent-6 months	dapsone 50mg/day for 1 month dapsone 25mg/day for 1 month
8*	F/20	back chest	remote-3 recur each summer	years dapsone 50mg/day for 1 month
9*	F/37	left chest upper back	1 month	dapsone 50mg/day for 1 week
10*	M/20	chest back	remote-2 years recent-2 months	no treatment

* Our present cases

through to early September for three years. Her family and medical history were non-contributory and routine laboratory evaluations revealed no abnormalities. A physical examination showed pinhead to rice-sized erythematous papules and reticulated brownish pigmentation on the back.

Biopsy specimens showed mild epidermal hyperplasia and mild hyperkeratosis, with focal dermal chronic inflammation.

Dapsone therapy was initiated using 50 mg daily and improvement in the lesions and symptoms were observed after two weeks. After one month, dapsone was tapered to 25 mg daily for one month. The medication was discontinued but 2 months later new pruritic lesions appeared on the same site and dapsone 25 mg daily was started again. After two weeks of medication, the lesions resolved. In June of the following year, the patient returned with the same problem which disappeared after three weeks' treatment of dapsone at 25 mg daily.

Case 3

A 20-year-old female presented with itchy papules on her back and anterior chest which she had had for three years with exacerbations each summer and improvement in autumn and winter. Upon skin examination, there were multiple pea to egg-sized dark erythematous papules, some of which had coalesced into plaques and patches of brown pigmentation. Her past medical history and family history were insignificant and the results of routine laboratory investigations were normal. A histopathological examination of a specimen ob-

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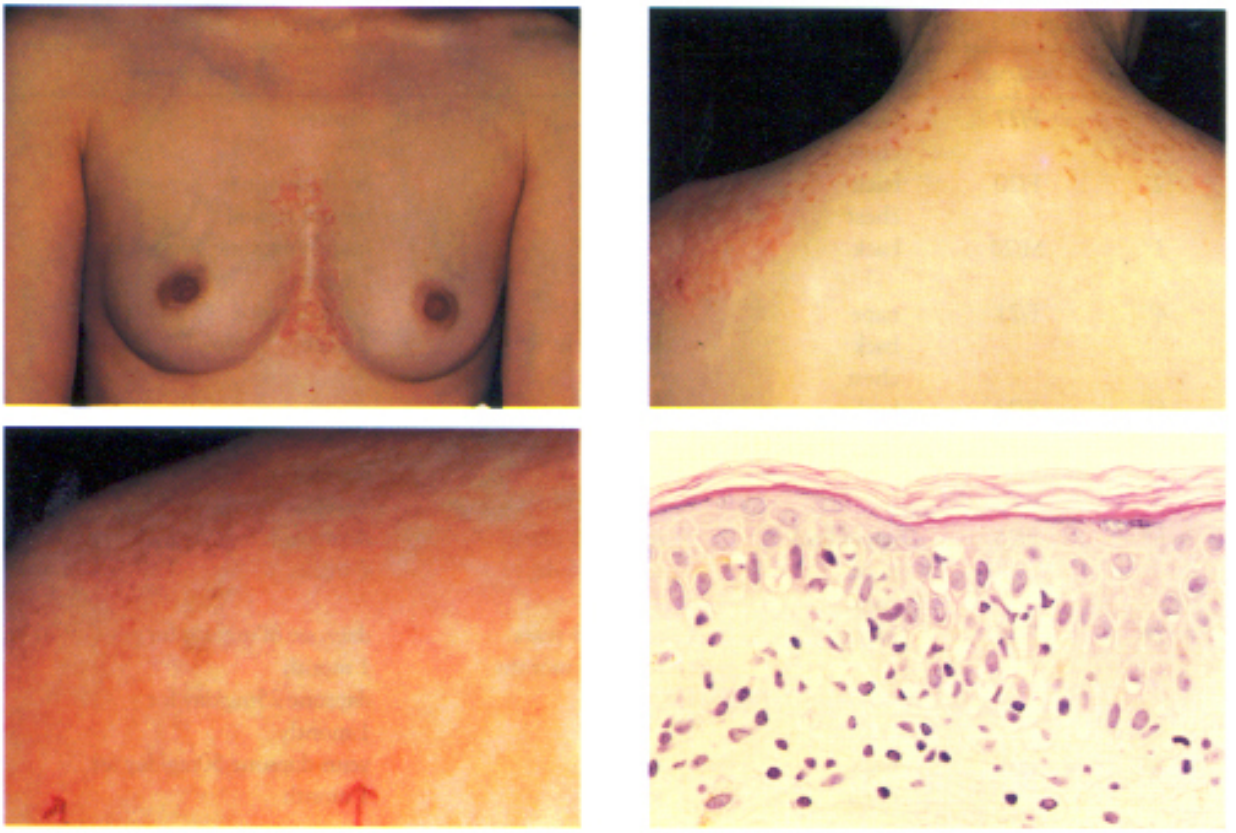


Fig. 1. Grouped erythematous papules on the anterior chest (a) and the left upper back (b), and brownish reticulated hyperpigmentation on the right upper back (b) ; close-up view of the grouped erythematous papules with reticulated distributions on the left upper back (c) ; a superficial perivenular lymphocytic infiltration with multifocal basal cell damage and exocytosis (d).

tained from an erythematous papule on the back revealed focal hydropic degeneration of the epidermis and mild perivascular infiltration of mononuclear cells in the dermis. In addition, findings of a specimen obtained from the reticulated pigmented area showed an increase in dermal melanophages. Dapsone 50 mg daily was started and after two weeks no new lesions appeared although there was not much improvement in the symptoms.

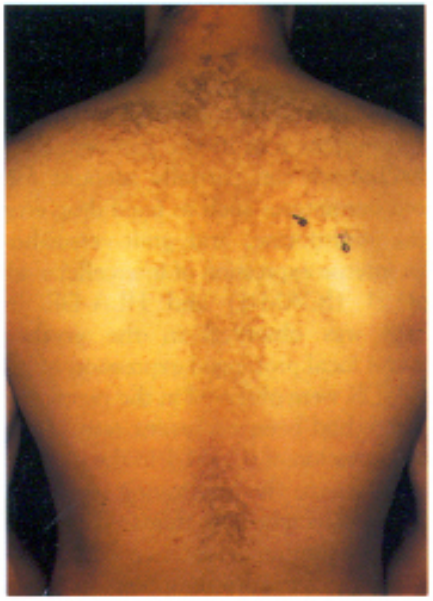
One month's treatment of dapsone resulted in almost total clearance of the papules but some of the pigmentation still remained. The medication was continued for two more months. There have been no recurrences for 18 months.

Case 4

A 37-year-old woman had had erythematous papules on the anterior chest and upper back and shoulders for 1 month. The patient complained of marked itching and noticed that the lesions were aggravated after alcohol consumption. On physical examination, there were grouped erythematous papules with areas of brownish reticulated hyperpigmentation on the anterior chest (Fig. 1a) and upper back (Fig. 1b, 1c). She recalled having the same problem on the corresponding site the previous year. The family and past medical history were non-contributory and routine laboratory tests were within normal limits. Antinuclear antibodies and VDRL test results were negative. A KOH smear of the upper back was also negative.

A skin biopsy revealed a superficial perivenular lymphocytic infiltration with multifocal basal cell damage and exocytosis(Fig. 1d).

On the basis of the above findings, the diagnosis of prurigo pigmentosa was established and treatment with dapsone 50 mg daily was started. After just 1



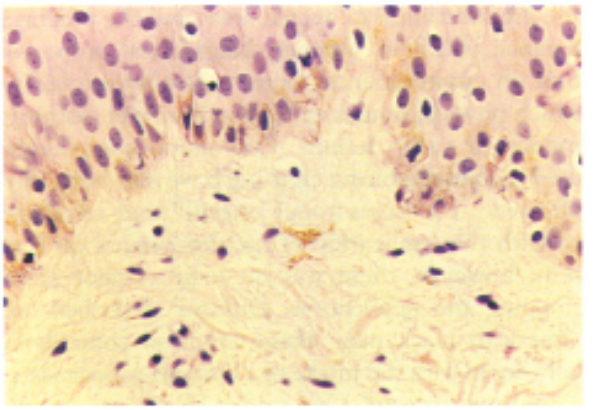


Fig. 2. A brownish reticulated pigmentation on the back (a) ; an increase in basal pigmentation and upper dermal melanin (b).

week, the papules disappeared and the pruritus subsided.

Case 5

A 20-year-old man presented with brownish reticulated pigmentation on the chest and back area. He had no symptoms but recalled having severely pruritic erythematous papules in the same regions 2 years ago which spontaneously resolved, leaving a brownish reticulated pigmentation (Fig. 2a). The same kind of erythematous papules developed 1 month ago but had disappeared at the time of his hospital visit with evidence of only the characteristic residual hyperpigmentation. The patient had no significant medical or family history and routine laboratory tests were within normal limits. A skin biopsy at the site of pigmentation showed an increase in basal pigmentation and upper dermal melanin (Fig. 2b). The patient recieved no specific treatment.

DISCUSSION

Prurigo pigmentosa(PP) is an intensely pruritic eruption that typically involves the trunk and resolves leaving a characteristic reticulate hyperpigmentation. It was first recognized by Nagashima[1] in 1971 and was described as " A peculiar pruriginous dermatosis with gross reticular pigmentation". Since this original report, more than 200 cases have been described from Japan[2] but reports from other parts of the world are quite rare[5-9]. In Korea, Hong[4] first reported 2 cases of PP in 1988 and since then, a few more cases have been reported (Table 1).

PP is characterized by the sudden onset of pruritic crops of erythematous papules that often coalesce. They may evolve into urticarial plaques and may have scales. Lesions resolve within several days or weeks leaving a reticulated, marble-like post-inflammatory hyperpigmentation for many months or years. The total duration may be 6 months to 8 years[3] and despite the fact that PP follows a chronic course, lichenification of the lesions are surprisingly rare. Recurrences and exacerbations are frequent with the recurrence occuring mainly in the pigmented areas, the other areas being normal. The distribution of the lesions typically involves the trunk and nape[3] although there have been other reports affecting the clavicular regions, chest, antecubital fossa, lumbosacral region, limbs, and forehead[2-3,5-6,9-11].The mucous membranes are spared. The lesions usually occur in spring and summer and are more common in adolescent females, but occasionally males and elderly persons are affected. Systemic findings have not been correlated and no laboratory abnormalities have been detected except for a few cases of peripheral blood eosinophilia

The pathogenesis of PP is still unclear although several hypotheses have been suggested. Considering

the predilection for covered areas and the preponderance of PP in summer, Nagashima[3] proposed that an environmental factor may play a role, in which physical trauma or friction from wet clothing induces the skin lesions. Para-amino compounds used in manufacturing clothing have also been implicated[13]. Two cases from Cotterill[14] were associated with the application of trichlorophenol on the skin; however, there was no improvement after stopping the application. Dijkstra[5] described an American black man whose eruption coincidentally began with the intake of a bismuth-subsalicylate containing antacid[5]. One case of PP was associated with the use of acupuncture needles for 3 years in a Japanese man[12] which was presumably caused by an allergic reaction to the chromiun component of the needles. Other authors think that PP might be a type of photodermatitis[6].

Histological features of PP are relatively nonspecific. Joyce[9] summarized the histological findings of the erythematous papules as a combination of a lichenoid tissue reaction with variable psoriasiform hyperplasia. Other characterisitics of the papular lesions are elongation of the rete ridges, intercellular and intracellular edema, exocytosis, basal cell liquefaction degeneration, papillary dermal edema, superficial blood vessel dilatation, and a mild perivascular lymphocytic infiltrate. The areas of reticulate hyperpigmentation show pigmentary incontinence and variable perivascular lymphohistiocytic infiltration. Pinkus[17] believed from a histopathological point of view, that pigmented dermatoses, such as erythema dyschromicum perstans, lichen pigmentosus, pigmented contact dermatitis, and PP, which all show lichenoid tissue infiltrations, were due to the reaction of the skin of predisposed individuals to an unidentifed environmental contaminant. Electron microscopic observation shows injury to the basal cells and severe damage to the lower epidermal cells suggesting that PP is a tissue reaction similar to lichen planus[18].

Dijkstra[5] reported granular deposition of C3 at the dermoepidermal junction and around papillary dermal vessels with direct immunofluorescence; however, most other reports have found no deposition of immunoglobulin or complement.

Immunohistochemical studies on the papular eruptions by Teraki[2] revealed a predominance of CD4+ cells in the dermal infiltrate, whereas those lymphocytes in the epidermis turned out to be mainly CD8+ cells. Moreover, some lymphocytes were still observed adhering to the epidermis in the pigmented areas. These findings support the view that CD8+ cells adhering to the epidermis play a prominent role in the pathogenesis of PP. This mechanism can partially account for other lichenoid dermatoses such as fixed drug eruptions, erythema multiforme, and graft-versus-host disease. An intense expression of intercellular adhesion molecule 1 (ICAM-1) and HLA-DR by keratinocytes was observed in the erythematous papules and focal prolonged expression of ICAM-1 and HLA-DR were observed in the pigmented areas where there had been no recent rash. The persistence of expression of ICAM-1 in the pigmented areas may be due to the continuing secretion of cytokines from T lymphocytes that are adherent to the epidermis. A similar persistence of ICAM-1 has been reported in fixed drug eruptions and this can be connected to the site-specific tendency of the eruption. Likewise, persistence of ICAM-1 in the pigmented areas of PP could account for the recurrent rash localized to the typical sites.

The treatment of choice for PP is dapsone and in 1973, Sugawara[15] first reported dramatic improvement following dapsone therapy of 25 mg to 100 mg daily. Following reports confirmed the efficacy of dapsone therapy. In 1979, Tashiro[21] first reported sucessful treatment with minocycline using 100 mg to 200 mg daily which resulted in rapid clearing and prevention of recurrences. In comparison, the response to topical or oral corticosteriods or oral antihistamines is poor. Miyachi[11] suggested that oxygen intermediates generated by the perivascular inflammatory cells in the dermis damages the tissue giving rise to the lichenoid tissue reaction. He proposed that dapsone and other sulfonamides might exert its anti-inflammatory effects by suppressing these oxygen intermediates induced by an unidentified allergen which ultimately protects the tissue from injury by the hydroxy radicals. Aso[16] reported good results with treatment with minocycline 100-200 mg daily for over I week in 5 PP patients. In particular, two patients showed recurrences after cessation of dapsone therapy and treatment with minocycline resulted in persistent clearance of lesions for over one year after minocycline therapy. Minocycline is thought to have the same anti-inflammatory properties through inhibition of oxygen intermediates.

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