A Case of Pityriasis Rubra Pilaris
Associated with Incidental Acantholysis

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Pityriasis Rubra Pilaris is a rare, chronic, mildly inflammatory disease characterized by fine acuminate follicular papules with orange-red to salmon-colored scaling of the skin and erythroderma that surround islands of normal, uninvolved skin, particular in the trunk. Most patients also develop palmpoplantar hyperkeratosis in the early course of the disease.

Focal acantholysis occurring in pityriasis rubra pilaris is an unusual and incidental histologic finding within the spectrum of histologic change of the disease. No case featuring this concomitant histologic finding has been reported in the Korean literatures up to date.

We report a case of focal acantholysis occurring in a patient with the clinical features of pityriasis rubra pilaris which may be considered as an incidental finding.

(Ann dermatol 7:4 354-357, 1995)

Key Words : Pityriasis Rubra Pilaris, Incidental Focal Acantholysis

Pityriasis rubra pilaris (PRP) is characterized by a reddish orange-colored scaling plaque, keratotic follicular papules and palmpoplantar hyperkeratosis. Their histologic appearances are parakeratosis around the follicular openings, hyperkeratosis, irregular acanthosis and mild chronic inflammatory infiltrate in the upper dermis, but focal acantholysis in PRP is an unusual histologic feature that may be considered as an incidental finding.

We describe a case of focal acantholysis occurring in a patient with the clinical feature of PRP.

REPORT OF A CASE

A 65-year-old male patient who was consulted in our Department of Dermatology for generalized mild itching sensation had been admitted to the Department of Internal Medicine for the evaluation and treatment of diabetes mellitus. On past medical history, he had suffered from diabetes mellitus for about 20 years. At admission, his laboratory findings were unremarkable, except for his blood glucose level (FBs:188, PP2hr:206) and triglyceride level (161mg/dL). He complained of generalized mild itching sensation about 3 weeks ago.

Physical examination revealed erythematous scaly patches with follicular papules and erythematous scaly patches on the upper trunk, especially around the neck (Fig. 1). The skin of the palms and soles had prominent hyperkeratosis with scaling and desquamation (Fig. 2). The finger and toe nails showed whitish thickening. He had a lesion of tinea cruris and onychomycosis confirmed by KOH mounting. Mucous membranes were normal.

Punch biopsy specimens were taken from two separate sites of the upper back and forearm with a 2-week interval and H&E staining was performed. Histologic findings of the lesion of the upper back were as follows; hyperkeratosis, parakeratosis around the follicular openings, suprabasal acantholysis extending into the stratum spinosum, and mild perivascular lymphocytic infiltration in the dermis (Fig. 3 a, b). Sections taken from a hyperkeratotic papule located in the forearm also showed
Fig. 1. Erythematous perifollicular papules with central keratotic acuminate plug in the upper trunk.

Fig. 2. Hyperkeratosis of the palms with a tendency to fissure.

Fig. 3. a) Hyperkeratosis, parakeratosis around the follicular opening, and suprabasal acantholysis in the epidermis, mild superficial perivascular infiltration in the dermis (H & E stain, ×100).

Fig. 3. b) Higher magnification of the acantholytic focus which extended into the stratum spinosum with no visible cellular infiltration in it (H & E stain, ×200).

Fig. 4. Hyperkeratosis with acanthosis in the epidermis and mild superficial perivascular infiltration in the dermis (H & E stain, ×100).

hyperkeratosis with acanthosis in the epidermis and mild perivascular lymphocytic infiltration in the dermis (Fig. 4). Immunofluorescent staining of the section taken from the forearm showed negative results. On the basis of the clinical and histopathological data, a diagnosis of PRP associated with focal acantholysis was made. Treatment was instituted with etretinate 30mg/day and topical steroid, and the lesion of tinea cruris was healed with a topical antifungal agent. The dose of etretinate was reduced to 20mg per day three weeks later. One month after the onset of treatment, erythroderma that surrounded islands of uninvolved skin was more conspicuous (Fig. 5) and desquamation was observed in the trunk, palms, and soles, but follicular papules were not present. About fifty days after the onset of treatment, the erythroderma
had disappeared and only slight desquamation remained.

**DISCUSSION**

Pityriasis rubra pilaris (PRP) is a chronic dermatitis characterized by erythematous or orange-red scaling plaques and keratotic follicular papules as well as diffuse erythroderma with areas of spared normal skin. The nails are grossly thickened and discolored distally showing splinter hemorrhage. The clinical manifestation and appearance of the lesions are often distinctive enough to allow the diagnosis of PRP. Both a juvenile and an adult type of the disease have been described. In the juvenile type, the onset is gradual; in the adult type of PRP, such as seen in the present case, the onset of the disease is more rapid.

The clinical differential diagnosis, particularly in the early phases of the disease, should include pityriasis rosea, treated psoriasis, papulosquamous drug eruption. In the later stage, the clinical features of PRP need to be distinguished from those of seborrheic dermatitis and generalized exfoliative erythroderma.

There are several therapeutic approaches for patients with PRP. Currently, retinoids and antimitabolites are the most successful therapies.

The characteristic histologic features observed in different stages of PRP have been reassessed and defined by Soerjomomo. The diagnostic histologic features include alternating hyperkeratosis and parakeratosis, hyperkeratosis of the follicular ostia and infundibula with follicular plugging, and a typical epidermal proliferation pattern i.e., thick suprapapillary plates, broad rete ridge, and narrow dermal papillae, but histologic changes vary according to the stage of the process and may therefore differ from site to site in the same patient.

The incidental finding of microscopic foci of a acantholytic dyskeratosis, reproducing the histological pattern of Darier's disease, has been described in a number of papers. The incidental finding of other types of acantholysis has been mentioned recently by Sanchez et al who studied 9,000 cutaneous biopsy specimens. In 14 of these (0.15%), a tiny acantholytic focus was found, either within the lesion or in the adjacent apparently normal skin. The histopathological diagnoses of these were basal cell carcinoma, keratoacanthoma, psoriasis, elastolytic granuloma, acral arteriovenous angioma, tinea corporis, leukocytoclastic vasculitis. The histological patterns of acantholytic foci simulated pemphigus vulgaris, superficial pemphigus, Hailey-Hailey disease, and unclassifiable acantholysis. These are incidental findings during the study of biopsy specimens of other conditions. In terms of these classification of incidental acantholysis, our case is incidental acantholysis of an unclassifiable pattern. The pathogenic mechanism of incidental acantholysis is unclear, but there has been speculation that sun or ultraviolet light exposure, viral infection, calcium metabolism, and hormonal etiology contribute to the development of these lesions. There were two cases of PRP associated with incidental acantholysis that represent the histologic pattern of Darier's disease, but no case featuring this concomitant incidental acantholysis has been reported in the Korean literatures up to date.

The incidental findings of focal acantholysis seen in various skin lesions most likely represent an unusual reaction of the epidermis to various associated conditions, rather than a specific histologic entity.

**REFERENCES**