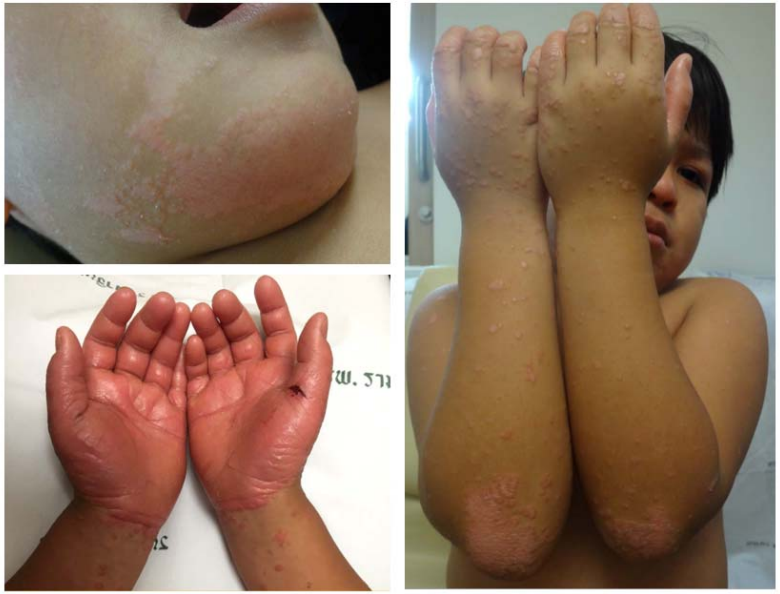
Case 12
A 5 year-old Thai girl from Bangkok
Chief complaint: Non-pruritic rash on her body for 3 weeks



Present Illness:

Six weeks PTA, she developed rash on both palms and soles then became cracked and painful. Initially, she was treated with cold cream and urea cream.

Three weeks PTA, the non-pruritic rash extended to the knees followed by the face, trunk and both extremities. They were treated with triamcinolone cream without improvement. She had no lesions on the mucosa or nails. Her mother denied history of recent fever, URI or diarrhea.

Past history:
Her mother denied underlying disease. She had normal growth and development.

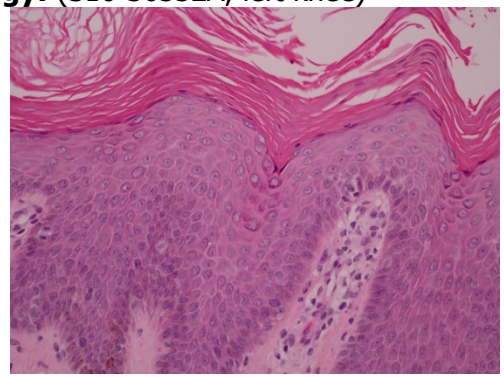
Family history: None of her family member has similar symptoms.

Skin examination:

- Multiple follicular scaly hyperkeratotic skin to salmon-colored papules coalescing into well-circumscribed plaques on forehead, chin, trunk and all extremities with predominantly on both elbows and knees.
- Bilateral symmetrical hyperkeratotic scaly salmon-colored to erythematous plaques on both palms and soles with some desquamations and few fissures.
- No mucosal involvement.

Physical examination:
Systemic examination other than skin lesions revealed no abnormality.

Histopathology: (S16-36532A, left knee)



- Alternating orthokeratosis and parakeratosis, psoriasiform epidermal hyperplasia
- Sparse superficial perivascular lymphocytic infiltration

Diagnosis: Pityriasis rubra pilaris type III

Treatment:
- Emoillient and topical steroid (0.1% triamcinolone cream) were applied on the lesions.
- Follow-up session was appointed to monitor the progression of disease.

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Discussion:

Pityriasis rubra pilaris (PRP) is a rare, chronic, inflammatory dermatosis with poorly understood etiology. The typical features of PRP include follicular hyperkeratosis and orange-red scaling plaques with island of normal skin. PRP affects both sexes similarly. The age distribution is bimodal with peak incidences in the first and fifth decades of life. An estimated incidence is about 1 in 5,000 new dermatology patients. Autoimmune, infections, internal malignancies and abnormal vitamin A metabolism are possible triggers.[1]

The clinical classification of PRP has been proposed by Griffiths'[2], based on clinical characteristics and course, which divided PRP into 5 types. Types I and II represent the classic and atypical forms of adult PRP, respectively, whereas types III, IV, and V are seen in juveniles. (Table 1.)

Table 1.

Type	Typical features	Other distinguishing features
I (>50%) Classic adult	Erythroderma with islands of normal skin, follicular hyperkeratosis, waxy diffused palmoplantar keratoderma	Cephalocaudal spread
II (5%) Atypical adult	Follicular hyperkeratosis and ichthyosiform lesions	Sparse scalp hair
III (10%) Classic juvenile*	Generalized coalescent hyperkeratotic follicular papules and plaques with islands of normal skin	Cephalocaudal spread
IV (25%) Circumscribed juvenile	Focal hyperkeratotic follicular papules and plaques on the elbows/knees/ palms/soles	Palmoplantar hyperkeratosis; disseminated small papules or plaques on face,trunk, or extremities
V (5%) Atypical juvenile	Generalized ichthyosiform dermatitis	Scleroderma-like changes of hands and feet

* Juvenile acute pityriasis rubra pilaris (JAPRP) considered as a subtype of type III, which differs from other PRPs from its acute

onset and self-limiting course. A possible trigger mechanism is mediation of superantigens from preceding streptococcal infections.[4] JAPRP is initially seen as scarlatiniform erythema, followed by the appearance of follicular papules that may generalize. A desquamation period of 2–3 months is followed by complete resolution. Recurrences with nearly identical skin lesions may be provoked by new infections with the same bacteria.[1,4]

A type VI variant related with HIV infection has been purposed. It was characterized by a distinctive triad of acne conglobata, lichen spinulosus, and a PRP-like eruption.

The nail changes of juvenile PRP include distal yellow–brown discoloration, subungual hyperkeratosis, thickening of the nail plates, and splinter hemorrhages. The involvement of the nail plate was observed in one-third of a PRP series.[5] The hair and teeth are usually unaffected. Mucous membrane involvement is occasionally presented with a diffuse whitish patch on buccal mucosa.[4]

Clinically, there are some difference between PRP in children and adults as shown in the table 2.

Type III form usually occurs between the ages of 5 and 10 years and runs a benign course with spontaneous clearing in 1-2 years.[4,7] Type IV, however, affects prepubertal children and young adults and has an uncertain prognosis, as a proportion of cases clear by the late teens. In contrary, type V is is characterized by an early age of onset and a chronic course. Most cases of familial PRP belong to type V.[1]

PRP may be clinically difficult to distinguish from psoriasis. Therefore, histopathology is an essential diagnostic criterion. PRP shows hyperkeratosis, acanthosis with broad short rete ridges and alternating orthokeratosis and parakeratosis oriented in both horizontal and vertical directions. Sparse superficial, perivascular lymphocytic infiltrate in the underlying dermis usually presents.

Table 2

	Children	Adults
▪ Male:Female	3:2[4]	1:1[1]
▪ Initial affected location	Lower part of the body[4]	Upper part of the body[1]
▪ Mucosal involvement	Occasionally seen[4]	Uncommon
▪ Most common type	IV[5]	I[1]
▪ Tendency to relapse	More	Less
▪ Prognosis	Better	Poorer

Keratinous plugs as well as shoulder parakeratosis may also be present.[1,4,7] The important features that help to distinguish from psoriasis are a prominent granular layer and dilated but not tortuous capillaries.

In our patient, we confirmed the diagnosis of Pityriasis rubra pilaris by histopathology. We initially diagnosed her as circumscribed juvenile (type IV) due to the predominated sites at elbows and knees. In the follow-up period the rash gradually extended to her trunk and face. Therefore, we finally made a final diagnosis of this case as classic juvenile (type III).

Topical treatment is often used in combination with systemic therapy to reduce the adverse effects of the latter. Topical therapy used in children includes topical corticosteroids, keratolytics, emollients, tar, calcipotriene (calcipotriol), and topical tretinoin cream. Even though it is off-label use, many case reports showed

significant improvement of isotretinoinin in juvenile PRP including an Asian patient.[6,7] Combined UVA1 radiation and acitretin therapy may be an alternative treatment.[8] In severe and recalcitrant PRP, methotrexate[1], cyclosporine[9], azathioprine[1], TNFα inhibitors such as Infliximab[10] and adalimumab[11] may be promising treatment options.

References

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