Case 17

A 45-year-old Thai woman from Petchburi.

Chief complaint: Progressive rash for 5 months

Present illness: 5 month ago, She had developed erythematous rash started from face, trunk and then spread to entire body. No history of photosensitivity, no alopecia

Skin examination

-Generalized well defined erythematous scaly plaque on scalp face and all extremities with the area of sparing normal skin

-Multiple discrete erythematous hyperkeratotic follicular papules at lower trunk

-Hyperkeratosis plaque at palms and soles

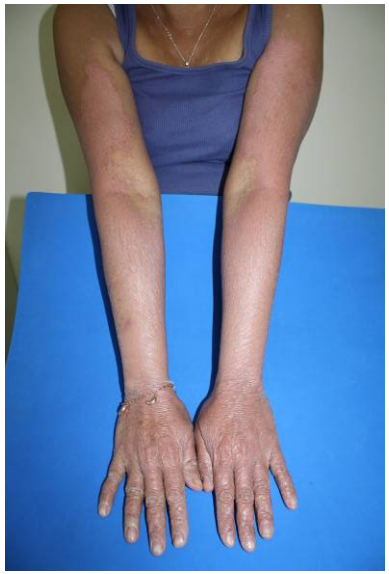
-No nail abnormality, no ectopion

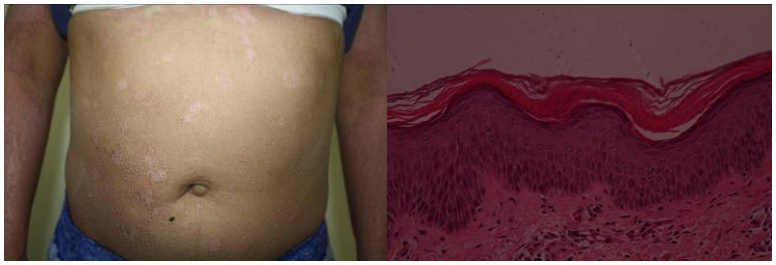
Laboratory investigation -AntiHIV -negative

Histopathology (54-0257)

Mild epidermal hyperplasia with irregular hyperkeratosis of alternating vertical and horizontal ortho- and parakeratosis

The hair follicles are dilated and filled with a keratinous plug





Diagnosis: Pityriasis rubra pilaris
(classic adult type,Type I)

Treatment: acitretin 10 mg oral OD, mineral oil apply bid, 5%urea apply bid , 0.1%TA apply bid , 2%Hydrocortisone cream apply at face bid

Presenter: Piyatida Prasertvit

Consultant: Sutthinun Wichyanrat

Discussion:

Pityriasisrubrapilaris (PRP) is a rare inflammatory dermatosis of unknown etiology,which characterized by follicular hyperkeratosis, perifollicular erythema and that may progress to generalized erythroderma, and palmoplantar hyperkeratosis. Lesions are characteristically orange or yellow, with approximately 1-cm islands of sparing.

Alphonse Devergie, a professor of dermatology in Paris,First called this rare disorder 'pityriasispilaris' in 1856.[1,2] Besnier finally classified the disease as pityriasisrubrapilaris (PRP) when describing several cases in 1889. Diagnosis can only be confirmedby clinical presentation and histologic examination; no typicalserologic markers existed. And The disease issubclassified into six types including both hereditary andacquired form according to Griffiths[3,4] as in table1

However Piamphongsant and Akaraphant[5] in a study of 168 PRP patients of Thai origin proposed a new classification of four types based only on physical (morphologic) findings. But nowadays Griffiths' classification into five or six well recognized groups remains the standard form. Not every case of PRP can be assigned with certainty to a specific type because of the possibility of intermediate forms and transitions from one type to another. Shahidullah and Aldridge[6] reported a child presenting with type III classicaljuvenile-onset PRP, who later developed type IV PRP.

Histologic findings for the diagnosis: alternating vertical and horizontal parakeratosis('checkerboard pattern') and irregular hyperkeratosis, epidermal acanthosis,follicular plugging, a thin stratum granulosum, and lymphohistiocytic perivascularinfiltrate in the underlying dermis. The stratum granulosum was partlythickened and always present in PRP but absent in psoriasis. inflammatory dermalinfiltrates, which, in PRP, exclusively consisted of lymphocytesand monocytes. In contrast to psoriasis, polymorphonuclearleukocytes and Munro microabscesses were always absent inPRP.

For the treatment, due to relatively rare of PRP no randomized, controlled trials on PRP have been published, only case reports and case series have seen. Anoverview of possible topical and systemic treatments in juvenileand adult PRP is given in tableII[7]. Nowadays no standardized therapy has been established. Topical corticosteroids, keratolytics, vitamin D3 analogs, andemollients are important for supporting systemic treatment ormay even be sufficient for treating localized forms of PRP. Retinoids remain the first-line systemic therapy. By the way Abbott and Griffiths[8] treated an adult patient with type I PRP with varying dosages of etretinate (0.45–0.75 mg/kg/day) during 9 years with only modest improvement. After a period without any treatment besides emollients, his skin symptoms resolved spontaneously after 20 years. Another study for the new drug retinoids, a 74-year-oldwoman who had not experienced stable remission of her skin symptoms during priortreatments including topical and systemic corticosteroids, phototherapy, orallyadministered acitretin, cyclosporine, methotrexate and adalimumab.Treatment with oral alitretinoin 30 mg daily has a good improvement only in weeks[9, 10].

Further studies are needed to be investigated. As PRP and psoriasis share clinical and histologic features, another modals in treatment for psoriasis have been tried in patients with PRP includingsPUVA, NB-UVB, cyclosporine, methotrexate, and combined therapy. Some cases have shown good response but some case not.

However in retractable PRP another choice can be TNFalphaInhibitors[11].the roleof TNFalpha in the pathophysiology of PRP remains unclear.

Type	Age at onset (%)	Clinical manifestations	Prognosis
I (classical adult)	Adult (55)	Red-orange plaques, confluent, with islands of sparing, beginning on the head and neck then spreading caudally. Perifollicular papules withkeratotic plugs. Diffuse, waxy palmoplantarkeratoderma	Spontaneous remission in 80% within 3 y
II (atypical adult)	Adult (5)	Ichthyosiform scales and areas of eczematous dermatitis and areas of alopecia. Palmoplantarkeratoderma with lamellated scales	Chronic course
III (classical juvenile)	5–10 y (10)	Similar to type I	Often remission after 1 y
IV (circumscribed juvenile)	3–10 y (25)	Sharply demarcated areas of follicular hyperkeratosis, erythema of knees and elbows; prepubertal children	Unclear
V (atypical juvenile)	0–4 y (5)	Mainly follicular hyperkeratosis; begins in first few years of life. Familial PRP	Chronic course
VI (HIV-associated)	Variable	Similar to typeI. Prominent follicular plugging with formation of spicules. Associated with acne conglobata, hidradenitis suppurativa, and lichen spinulosus	Variable[a]

Table II. Treatment of pityriasis rubra pilaris (PRP)

Topical treatment

Emollients
Urea 5–10%
Salicylic acid 1–3%
Corticosteroids[8,79]
Topical retinoids 0.05–0.1%[44]
Calcipotriene (calcipotriol)[80]
Pimecrolimus 1%[81]

Systemic treatment in children (off-label use)

Acitretin 0.5 mg/kg/day (3–5 mo) ± UVB 311[82,83]
Isotretinoin 0.5–1 mg/kg/day (>12 y)[8,82]
Only in severe recalcitrant cases: methotrexate, cyclosporine (ciclosporin), azathioprine (>10 y),[8,84] or TNFα inhibitors[85-87]

Systemic treatment in adults

First-line treatment
Retinoids (isotretinoin 1 mg/kg/day; etretinate 0.5 mg/kg/day)[9,79,88-90]
Methotrexate (5–30 mg/wk)[9,90]
Second-line treatment
Cyclosporine (<5 mg/kg/day)[91,92]
Acitretin + UVA1[93]
Re-PUVA[94]
ECP[95,96]
Fumaric acid[6]
Intravenous immunoglobulins (2 g/kg over 3 days)[97]
Antiretroviral therapy (type VI PRP)[31]
TNFα inhibitors[98-106]
Stanozolol (2 mg/day)[10]
Azathioprine (50–200 mg/day)[10]

ECP = extracorporeal photochemotherapy; **Re-PUVA** = retinoid and psoralen plus UVA; **TNF** = tumor necrosis factor.

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