Case 28

Salmon-colored rash in a teenager

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Patient: A 19-year-old Thai man from Chachoengsao

Chief complaint: Generalized orange rash for 19 years

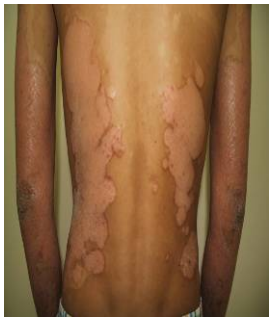
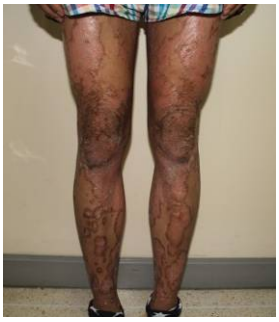
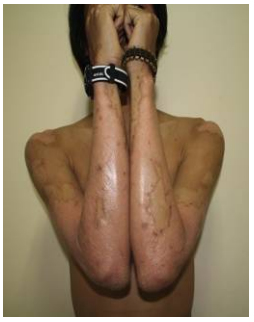
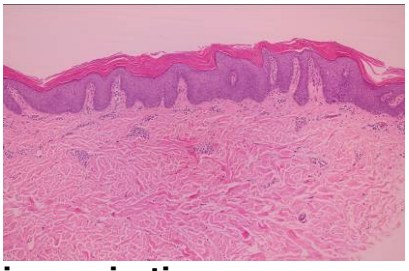
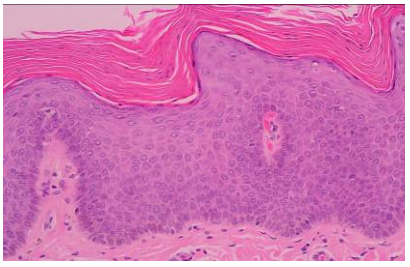
Present illness:

The patient has developed dry and cracking orange rashes since he was a nine-month-old. The rashes started from the left hand then spreaded to other parts of the body, predominantly on extremities. Some rashes faded and some developed in new areas.

Past history: No underlying disease

Family history:

His mother developed the similar rashes with less severity.

Skin examination:

- Symmetrical salmon-colored plaques with fine scales on face trunk and extremities with scalp, nail, palm and sole sparing

Histopathology: (S14-17782, back)

- Alternating ortho-parakeratosis with mild papillated epidermal hyperplasia
- Superficial perivascular inflammatory-cell infiltrate

Diagnosis: Pityriasis rubra pilaris (Type V, atypical juvenile)

Treatment:

- Advice about the natural history and prognosis of disease
- Oral isotetrinoin 10 mg/day
- Emollient for face and body

Discussion:

Pityriasis rubra pilaris (PRP) is a name given to a group of rare skin disorder of uncertain etiology. It was first described in 1835 as a variant of psoriasis. Later, this condition was recognized as a separate entity and named it "pityriasis rubra pilaris" in 1889.

PRP affects both men and women equally. The diagnosis of PRP is often difficult. It is commonly confused with other papulosquamous and erythrodermic disorders, particularly psoriasis. Diagnosis can only be confirmed by clinical presentation and histologic examination; no typical serologic markers exist for PRP[1].

Clinically, The classic presentation of PRP is follicular hyperkeratosis as the elementary lesion. They can coalesce to form large salmon-colored to orange-red plaques with islands of sparing in different areas of the body. The plaque can then progress to an erythrodermic appearance. In Thailand, Piamphongsant and Akaraphant[2] found only 14 of 168 PRP Thai patients presenting with erythroderma.

The clinical course of PRP is highly variable. In 1980, Griffiths[3] classify PRP into 5 types based on age, duration, and type of cutaneous involvement. The generalized eruption is seen in all types except type IV. Type I (50%), classic adult, the most common type, show a classic clinical presentation. It tends to clear spontaneously in 80 percent of patients within 3 years. Type II (5%), atypical adult, differs form type I by palmoplantar keratoderma with coarse lamellated scale, a more icthyosiform scaling. It tends to be more chronic and presents with areas of alopecia, eczematous patches. Type III (10%), classic juvenile, has the same clinical presentation as type I but occurs in children. It spontaneously clears within 3 years. Type IV (25%), circumscribed juvenile, the most common of juvenile type, affects prepubertal children and young adults. It presents with circumscribed follicular hyperkeratosis and erythema of the elbows and knees. Type V (5%), atypical juvenile, occur in the first few years of life and is very persistent. It has features similar to type II with more icthyosiform scaling. Some patients develop scleroderma-like changes on the palms and soles. It is generally inherited in an autosomal dominant fashion with variable expression. Other forms of inheritance, such as autosomal recessive and X-linked, have also been reported[4]. Most cases of familial PRP belong to this type. In 1995 Miralles et al[5] proposed a new type VI for PRP patients with HIV infection. In Thai study, the presentation of Thai PRP patients may be different from Caucasian patients. Piamphongsant and Akaraphant[2] posed a new four-type classification based on physical finding. More than 50% of the patients belonged to the type that presents with palmoplantar keratoderma, circumscribed scaly erythematous patches on elbows and knees, and follicular plugging in the patches when first seen.

Histopathologic features of PRP show psoriasiform hyperplasia with with alternating orthokeratosis and parakeratosis in both vertical and horizontal directions ('checkerboard pattern'). Hair follicles are dilated, and filled with keratinous plug. An occasional distinctive change is 'shoulder parakeratosis,' in which parakeratosis is present adjacent to both sides of the follicle. Focal or confluent hypergranulosis as well as thick, shortened rete pegs are other helpful

diagnostic features. Superficial perivascular and perifollicular infiltrate of lymphohistiocyte is often seen in the underlying dermis[1].

Due to the rarity of the disease, there are only case reports and case series. Large-scale, randomized controlled trials are not available. There are several treatments for PRP both topical and systemic therapies studying on the limited number of the patients. The efficiency of the treatment is difficult to assess.

There are some small-scale studies with some responses on topical drugs such as topical corticosteroids[6], topical calcipotriol[7], 1% pimecrolimus cream[8], 0.1% tazarotene gel[9], Goeckerman regimen[6, 10].

Systemic therapy tends to be more effective than topical therapies. To date, Systemic retinoids seem to be most effective drugs, so retinoid remain the first-line therapy for most patients[1]. Re-PUVA may be beneficial in some patients[11]. But phototest should be done before treatment because there is some reports of photoaggravated PRP. Due to some efficacy of methotrexate on PRP[3], it can be used as a second-line therapy when the retinoid is not responsive[1]. The combination between methotrexate and oral retinoid may be beneficial[12]. However this combination may lead to increase hepatotoxicity. The biologic drug may be effective in some patients[13]. Treatment with cyclosporine, azathioprine, stanozolol and PUVA has been reported with varying degrees of success. Systemic corticosteroids are not useful in PRP due to exacerbation after discontinuation[14].

For the rare type V juvenile PRP, there is a case report of dramatic improvement (almost complete clear) of fifteen-year-old boy with recalcitrant PRP after 5 week of treatment with fumaric acid[15].

For symptomatic relief, emollients, keratolytics, and systemic antihistamine therapy are beneficial. Some patients with severe pruritus resistant to anti-histamine and potent topical steroid may relieve with topical capsaicin solution (0.03%)[11]

The clinical and histopathological findings recover the diagnosis of PRP. Our patient is 19; he has had the persistent rashes since he was nine months old. His family has a history of similar rashes. From his clinical history, it matches the diagnosis of a rare type V PRP, although he did not have sclerodermoid changes of hands and feet. He was treated with systemic isotretinoin and emollient and we look forward to seeing the outcome.

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