

PhotoDermDiagnosis

Erythematous acrally prominent plaques with palmoplantar keratoderma

Section Editor

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A 30-year-old-male truck driver presented to us with an itchy erythematous eruption on his extremities of 6 months duration. The lesions first appeared on his upper limbs without any previous history of trauma or infection and soon spread to involve the lower limbs below the knees. Symptomatic treatment with keratolytics and emollients failed to cause resolution of the lesions. Cutaneous examination revealed grouped erythematous hyperkeratotic follicular papules on the upper and lower extremities bilaterally. The lesions had coalesced to form scaly erythematous plaques on the elbows, knees, dorsa of hands and feet with islands of normal skin (**Figures 1**). The rest of the body was not involved. Palms (**Figure 2**) and soles showed waxy hyperkeratosis with fissuring at places and



Figure 1



Figure 2

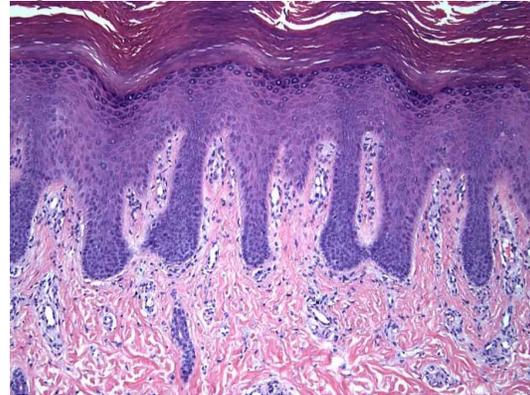


Figure 3

there was onychodystrophy with subungual hyperkeratosis affecting all the toe nails. All the routine haematological and biochemical investigations were normal. ELISA for HIV was negative. Skin biopsy from the erythematous hyperkeratotic follicular papule is shown (**Figure 3**).

What is your diagnosis?

Diagnosis

Pityriasis rubra pilaris (type 2)

Discussion

Pityriasis rubra pilaris (PRP) is a chronic papulosquamous disorder of unclear etiology. The first description of PRP is credited to Claudius Tarral¹ in the year 1835, but it was Besnier in 1889 who named it as PRP.² The incidence is bimodal with peak in the first and fifth decade. Most cases are acquired; however, a familial form of the disease exists with an Autosomal dominant mode of inheritance.³ The classic form of PRP has a cephalocaudal distribution, and is characterised by hyperkeratotic follicular papules coalescing to large scaly erythematous plaques that may progress to erythroderma with palmoplantar keratoderma. Lesions are typically orange in colour with 1cm islands of sparing within areas of erythroderma. 20% of patients complain of pruritus or burning sensation. Acuminate follicular papules resembling a 'nutmeg grater' are seen on the dorsal phalanges and extensor aspect of wrists. Nail involvement manifests mostly as yellow brown discolouration and thickening of the nail plate, subungual hyperkeratosis and splinter haemorrhages.⁴ PRP has been classified into five types by Griffiths based on the age and pattern of onset, as well as prognosis.⁵ Type 1 is classic adult onset PRP with clinical features as described above. Type 2 is atypical adult onset PRP seen in only 5% cases, which differs from type 1 based on its longer duration and atypical morphological features. Type 1 PRP accounts for 50% cases and has the best prognosis with 80% resolving within 3

years. Types 3-5 are the juvenile onset forms of PRP. Type 3 PRP is the classical juvenile onset PRP which differs from type 1, only by its onset in childhood. Type 4 PRP is the circumscribed juvenile onset PRP, the most common type of PRP seen in children, characterised by well-demarcated symmetrically distributed plaques on the knees, elbows and ankles. Type 5 PRP is the atypical juvenile PRP which like type 2 is chronic and has ichthyosiform features. Recently, type 6 PRP has been described in patients of HIV characterised by a poor prognosis and refractoriness to treatment.⁶

The pathogenesis of PRP is unknown; however, theories such as dysfunctional keratinisation or abnormal vitamin A metabolism, physical triggers such as trauma, an autoimmune phenomenon and a superantigen-mediated process have been proposed.⁷ Histologically, PRP is characterised by an acanthotic epidermis with broad and short rete ridges and alternating ortho- and parakeratosis in both vertical and horizontal directions (checker board pattern). Follicular plugging with perifollicular areas of parakeratosis may be evident with a superficial perivascular infiltrate of lymphocytes in the dermis. Additional features that may help in differentiation from psoriasis include hypergranulosis, thick supra papillary plates and acantholysis seen in the former.⁸

PRP must be differentiated from psoriasis, follicular eczema, follicular psoriasis, seborrheic dermatitis, follicular ichthyosis and erythroderma variabilis. Distinguishing features of PRP include "islands of spared skin" within generalised erythroderma, follicular keratotic papules

Table 1 Classification of pityriasis rubra pilaris [5].

Clinical type	% affected	Age at onset	Distribution	Skin findings	Course
I	50%	Peaks in the 6 th decade	Generalised beginning in cephalocaudal Direction	Orangish red plaques with islands of sparing, perifollicular keratotic papules, waxy PPK	Often resolves in 3 years
II	5%	Adults of various ages	Generalised	Areas of eczematous dermatitis, Ichthyosiform scaling on the limbs	Chronic
III	10%	Peaks in first few years then in teens	Generalised	Orangish red plaques with islands of sparing, Less than 50% have PPK	Chronic but often resolves in 3 years
IV	25%	Pre Pubertal	Focal with involvement of elbows and knees	Well demarcated erythematous plaques, perifollicular papules with follicular plugging, diffuse waxy PPK	Variable
V	5%	First few years of life	Generalised	Ichthyosiform dermatitis, perifollicular papules with keratotic plugging, scleroderma like appearance on hands and feet, accounts for most of the familial cases of PRP	Chronic
VI	NA	Variable	Generalised	Similar to Type I, associated with acne conglobata, hidradenitis suppurativa and lichen spinulosus like lesions	May respond to ART

and an orangish hue to the involved skin. Because of the relative rarity of PRP, randomized, double-blind, placebo-controlled trials assessing treatment options are scanty. Nevertheless treatments that have been tried for PRP include topical steroids, topical and oral vitamin A, topical vitamin D analogues, oral retinoids, methotrexate, azathioprine, UV phototherapy. Current evidence states that initiation of therapy with oral retinoids with low dose weekly methotrexate seems to offer the best improvement.⁹

References

1. Tarral C. General psoriasis. Desquamation from the parts covered with hair. In: Rayer PA, ed. *Theoretical and Practical Treatise on the Skin, 2nd edn.* London: Bailliere; 1835. p. 648-9.
2. Besnier E. Observations pour servir a l'histoire clinique du Pityriasis rubra pilaire. *Ann Dermatol Syphiligr (Paris)* 1889; **10**: 253-87.
3. Vanderhooft SL, Francis JS, Holbrook KA. Familial pityriasis rubra pilaris. *Arch Dermatol* 1995; **131**: 448-53.
4. Albert M, Mackod B. Pityriasis ribra pilaris. *Int J Dermatol* 1999; **38**: 1-11.
5. Griffiths WA. Pityriasis ribra pilaris. *Clin Exp Dermatol* 1980; **5**: 105-12.
6. Miralles ES, Nunez M, Delas Heras ME. Pityriasis ribra pilaris and human

- immunodeficiency virus. *Br J Dermatol* 1995; **133**: 990-3.
7. Bragg J, Witkiewicz A, Orlow SJ, Schaffer JV. Pityriasis rubra pilaris, type 4. *Dermatol Online J* 2005; **11**: 14.
 8. Magro CM, Crowson AN. The clinical and histomorphological features of PRP: a comparative analysis with psoriasis. *J Cutan Pathol* 1997; **24**: 416-24.
 9. Clayton BD, Jorizzo JL, Hitchcock MG, Fleischer AB Jr. Adult Pityriasis rubra pilaris: a 10 year case series. *J Am Acad Dermatol* 1997; **36**: 959-64.

**26th PAD and 5th SARAD Conference of Dermatology,
November 15-18, 2007**

Lahore, Pakistan

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