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An Adolescent With Desquamation of Both Feet

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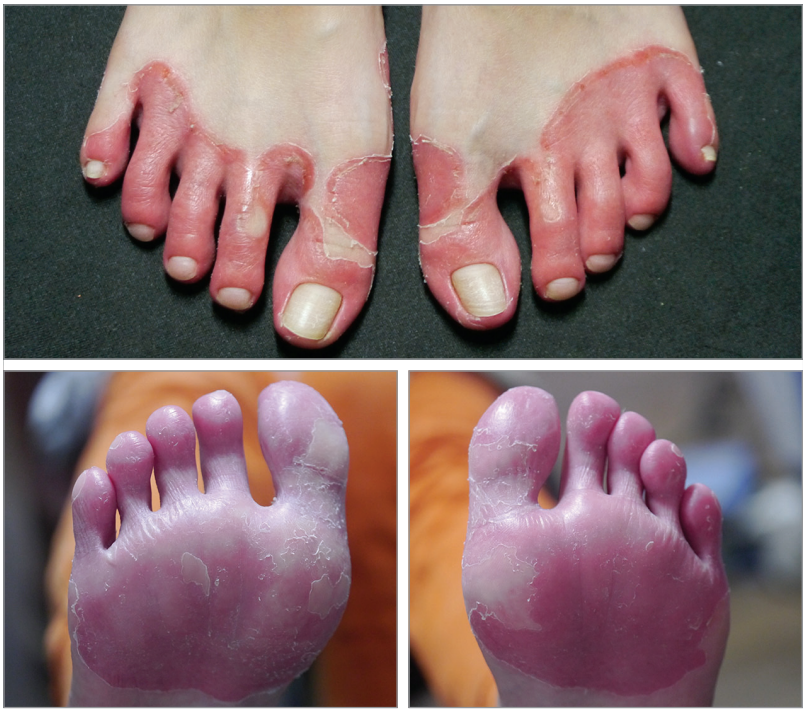

Figure 1. Well-defined hyperkeratotic desquamation on both feet, representing a sandal appearance.

A healthy adolescent girl presented with a prolonged dermatitis of both feet characterized by pruritus and accompanying desquamation (**Figure 1**). Physical examination revealed well-demarcated desquamative patches with scales on both distal feet, showing a sandal appearance. No local heat, swelling, or discharge from the skin lesions was noted. The patient's medical history was otherwise unremarkable and she had no known history of atopic dermatitis. A fungal smear skin test with potassium hydroxide preparation was performed on the lesion, which showed negative findings.

WHAT IS YOUR DIAGNOSIS?

A. Tinea pedis

B. Psoriasis

C. Pityriasis rubra pilaris

D. Juvenile plantar dermatosis

Diagnosis

C. Pityriasis rubra pilaris

Discussion

A skin biopsy was performed at the end of the stem of the lesion. Histopathologic testing revealed alternating orthokeratosis and parakeratosis in a checkerboard pattern and epidermal hyperplasia without neutrophilic infiltration. In combination with the clinical findings of localized plantar keratoderma (keratoderma is a group of disorders characterized by thickening of the skin with hyperkeratosis), pityriasis rubra pilaris (PRP) was diagnosed. Differential diagnoses, including tinea pedis and psoriasis, were ruled out based on negative fungal smear skin tests and absent neutrophilic infiltration on histopathologic examination. Juvenile plantar dermatosis was also ruled out as it frequently affects patients with atopic dermatitis and involves the weight-bearing and frictional areas of the feet, leaving toe webs spared. After treatment with oral, low-dose isotretinoin, 0.3 mg/kg/d, for 2 months, the lesions resolved significantly (**Figure 2**). No relapse occurred over 5 months with ongoing treatment.

Pityriasis rubra pilaris is a rare papulosquamous disease with an incidence of 1 in 3500 to 5000 patients in the United States.[1] Pityriasis rubra pilaris shows a bimodal age distribution, peaking in the first and fifth to sixth decades. Males and females are equally affected. Griffiths[2] originally classified PRP into 5 subtypes.[1] Type 1 (classic adult type, >50% of all cases) is the most common subtype characterized by follicular hyperkeratotic papules that spread in a cephalocaudal direction

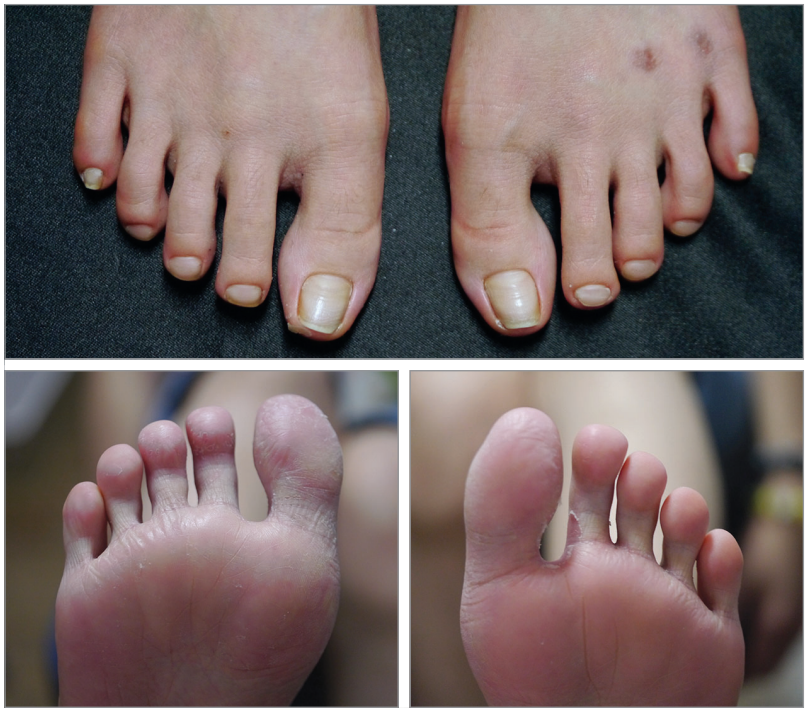

Figure 2. After treatment with oral, low-dose isotretinoin, 0.3 mg/kg/d, for 2 months, the lesions showed significant improvement.

and often progress to a generalized erythroderma with islands of normal skin. A waxy, diffuse, yellowish keratoderma of the palms and soles, the so-called sandal appearance, may occur. This form of PRP carries the best prognosis, showing a remission rate of more than 80% within 3 years. Type 2 (atypical adult type, 5%) is characterized by a duration of 20 years or more as well as by atypical morphologic features. Follicular hyperkeratosis, as well as ichthyosiform scaling, especially on the lower limbs, dominates the clinical picture. Type 3 (classic juvenile type, 10%) seems to be the counterpart of the classic adult type (type I PRP) with the age of onset the only difference, and type 3 PRP spontaneously clears within a year. Type 4 (circumscribed juvenile type, 25%) is characterized by well-demarcated hyperkeratotic erythematous plaques limited usually to the elbows and knees with palmoplantar involvement, showing a 3-year remission rate in approximately one-third of affected patients. Type 5 (atypical juvenile type, 5%) is characterized by an early age of onset and a chronic course. It is distinguished by follicular hyperkeratosis and a scleroderma-like appearance on the hands and feet. Following the initial categorization, human immunodeficiency virus–associated PRP was added as a separate type (type 6), showing clinical features similar to those of type 1.[2,3] However, the clinical spectrum of PRP could be diverse, and some cases are not neatly categorized by the classification based on age and body surface involvement.[4]

Therapeutic options for PRP include retinoids, antimetabolites, immunosuppressive agents, antibiotics, UV phototherapy, and biologic agents.[1,5] Among these treatments, systemic retinoids, such as isotretinoin, have shown the best outcomes.[1,6,7] The short-term use of oral retinoids in otherwise healthy children with PRP seems to be well tolerated and safe. In previous studies, isotretinoin, at dosages of 1 to 2 mg/kg/d for 3 to 6 months, showed favorable results.[6,7] More recently, alitretinoin, at dosages of 0.5 mg/kg/d, also yielded a convincing clinical response after 7 months.[8] In the present case, the patient responded to low-dose isotretinoin treatment of 0.3 mg/kg/d; this could be a feasible treatment option for pediatric patients without serious adverse events.

Differentiation between PRP and other diseases is important because of the difference in treatment and prognosis. The presence of follicular keratotic papules, diffuse palmoplantar keratosis, recalcitrance to topical corticosteroid treatment, and the absence of neutrophilic infiltration and alternating orthokeratosis and parakeratosis on histopathologic examination favor a diagnosis of PRP.[9] A waxy erythematous palmoplantar keratoderma can also be helpful in distinguishing psoriasis (scaly and pink-red) from PRP (waxy and pink-orange).

ARTICLE INFORMATION

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REFERENCES

1. Klein A, Landthaler M, Karrer S. Pityriasis rubra pilaris: a review of diagnosis and treatment. *Am J Clin Dermatol*. 2010;11(3):157-170.

2. Griffiths WA. Pityriasis rubra pilaris. *Clin Exp Dermatol*. 1980;5(1):105-112.

3. Misery I, Faure M, Claidy A. Pityriasis rubra pilaris and human immunodeficiency virus infection–type 6 pityriasis rubra pilaris? *Br J Dermatol*. 1996;135(6):1008-1009.

4. Piamphongsant T, Akaraphant R. Pityriasis rubra pilaris: a new proposed classification. *Clin Exp Dermatol*. 1994;19(2):134-138.

5. Dicken CH. Treatment of classic pityriasis rubra pilaris. *J Am Acad Dermatol*. 1994;31(6):997-999.

6. Dicken CH. Isotretinoin treatment of pityriasis rubra pilaris. *J Am Acad Dermatol*. 1987;16(2, pt 1):297-301.

7. Goldsmith LA, Weinrich AE, Shupack J. Pityriasis rubra pilaris response to 13-*cis*-retinoic acid (isotretinoin). *J Am Acad Dermatol*. 1982;6(4, pt 2)(suppl):710-715.

8. Yun CH, Kim JS, Ryu HR, et al. Circumscribed juvenile pityriasis rubra pilaris responsive to alitretinoin. *Dermatol Ther*. 2016;29(2):81-83.

9. Soeprono FF. Histologic criteria for the diagnosis of pityriasis rubra pilaris. *Am J Dermatopathol*. 1986;8(4):277-283.