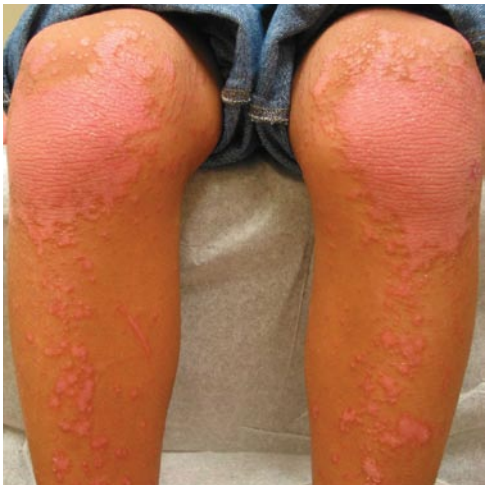
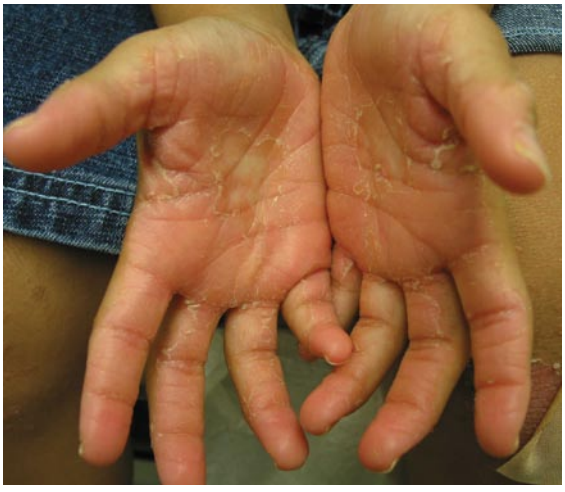
What Is Your Diagnosis?





A 4-year-old boy presented with a 2-month history of generalized, asymptomatic, scaly, expanding plaques.

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The Diagnosis: Juvenile Pityriasis Rubra Pilaris

The patient presented to the dermatology clinic after 2 months of unsuccessful treatment with topical and systemic antifungals prescribed by his primary care physician. Results of a biopsy specimen of a characteristic lesion showed irregular acanthosis, psoriasiform epidermal hyperplasia, dilated plugged follicles, perifollicular parakeratosis, and a sparse lymphohistiocytic perivascular infiltrate consistent with pityriasis rubra pilaris (PRP).

PRP is an uncommon papulosquamous disorder with an incidence of 1 in 5000 to 1 in 50,000.[1,2] It has no known gender bias. The etiology of PRP is unknown, though vitamin A deficiency, dysfunction in keratinization, and dysfunction in vitamin A metabolism have been proposed.[3] The clinical findings in PRP are well-established. Follicular hyperkeratosis with surrounding erythema is typical. PRP in adults typically begins on the scalp and spreads caudally (Figure 1); in children, PRP often begins on the lower extremities (Figure 2).[4] Other classic findings include the "nutmeg grater" appearance of the dorsal fingers caused by the rough perifollicular papules; the "islands of sparing," caused by coalescence of large plaques on the trunk, surrounding healthy skin; and a waxy orange-red palmar and plantar keratoderma (Figure 3).

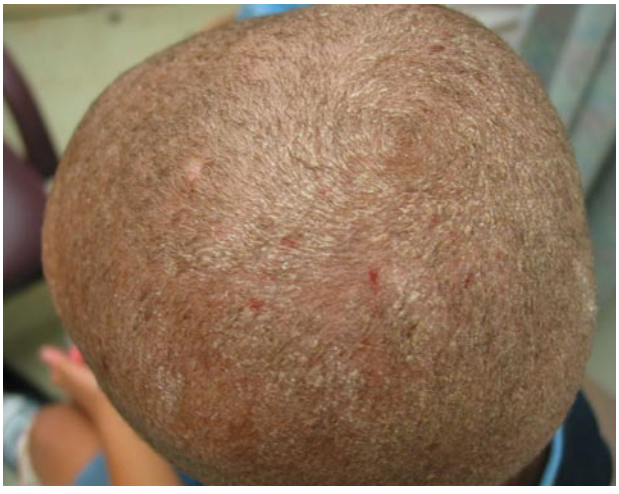

Figure 1. Confluent scaling of the scalp.

Griffiths[1] proposed a classification system to help distinguish the varied clinical presentations of PRP. Types I and II are both adult forms of PRP and together account for nearly 60% of all cases of PRP. Types III through V are juvenile forms of PRP. Type III, classic juvenile PRP, accounts for 10% of all cases of PRP and resembles type I PRP in adults. Type III is a generalized PRP that exhibits classic findings. These patients with type III PRP typically clear within 3 years of diagnosis. We believe our patient is classified as having type III PRP. Type IV, circumscribed juvenile PRP, accounts for 25% of all cases of PRP but is more focal, with an uncertain course. Type V, atypical juvenile PRP, accounts for the remaining 5% of cases of PRP, is generalized, and has a more chronic course.[3]

Biopsy specimens of PRP typically show irregular acanthosis and hyperkeratosis, with parakeratosis

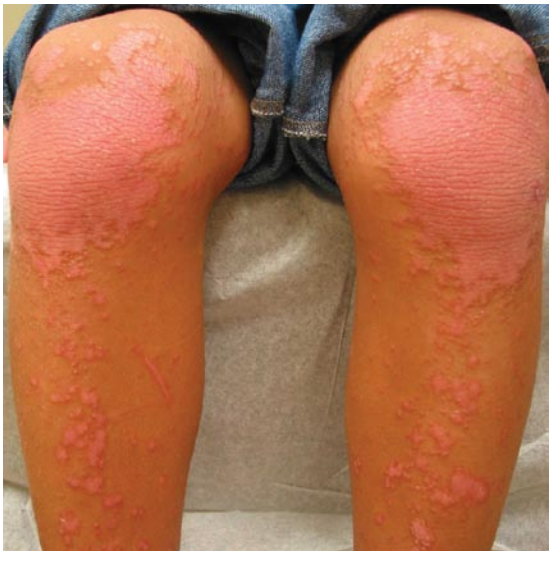

Figure 2. Hyperkeratotic papules and plaques.

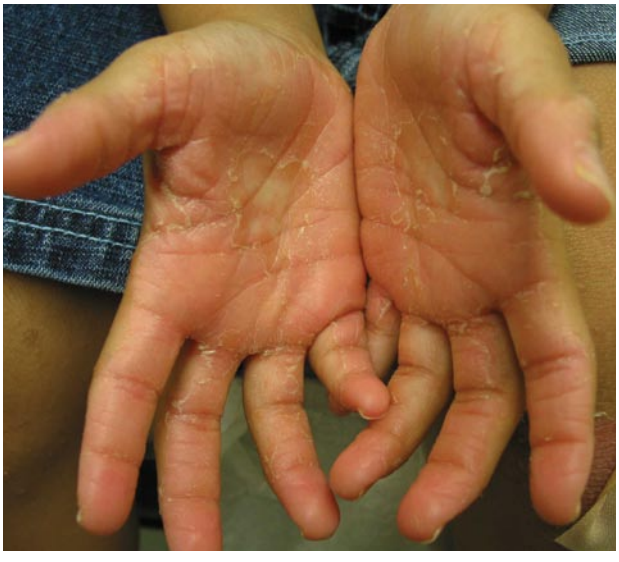

Figure 3. Palmar erythema and scaling.

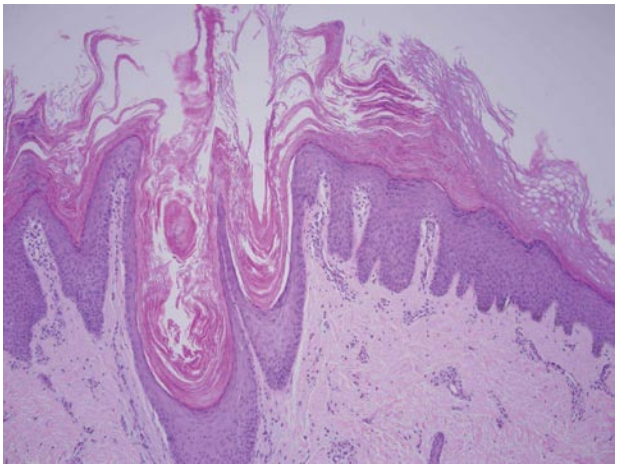

Figure 4. Hematoxylin and eosin stain of a biopsy specimen demonstrating irregular acanthosis, psoriasiform epidermal hyperplasia, dilated plugged follicles, and perifollicular parakeratosis (original magnification ×20).

around follicular openings (Figure 4). A classic histopathologic finding is the checkerboard pattern within the stratum corneum, caused by alternating vertical and horizontal parakeratosis, though this feature is not always present.[5] A sparse chronic inflammatory infiltrate often is present in the papillary dermis.

Safe effective treatment options for juvenile PRP are limited, as there is no known etiology and there are no large-scale pediatric studies. While good results have been obtained in treating adult PRP using methotrexate or isotretinoin,[6] side-effect profiles of these medications limit their use in children. Given the potential for spontaneous clearance, a conservative approach is advised. Our patient was started on topical triamcinolone acetonide ointment 0.1% and topical tretinoin cream 0.025%. At 6-month follow-up, the papules and plaques had cleared and only residual hypopigmentation remained in the affected areas.

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