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Successful treatment of a child's pityriasis rubra pilaris (PRP) with ustekinumab and acitretin

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Abstract

Pityriasis rubra pilaris (PRP) is a rare inflammatory skin disease that occurs with phenotypic variability in adults of all ages as well as in children. Data on the treatment of PRP is limited. Here, we report a 5-year-old girl with widespread skin involvement and prominent palmoplantar hyperkeratosis who was initially treated for psoriasis. After reevaluation, a diagnosis of PRP was made, and the patient had an excellent therapeutic response to ustekinumab and acitretin.

KEYWORDS
children, pityriasis rubra pilaris, retinoids, successful treatment, ustekinumab

1 | CASE REPORT

A 5-year-old female presented with koebnerizing psoriasiform skin lesions, palmoplantar hyperkeratosis, and fatigue that had occurred after a respiratory tract infection. Based on morphology, histology revealing psoriasiform dermatitis, and increased anti-streptolysin levels, the patient was diagnosed with infection-triggered psoriasis and treated with topical glucocorticoids, systemic antibiotics, and adalimumab (40 mg loading dose, followed by 20 mg subcutaneously every other week), without improvement. Palmoplantar keratoderma worsened and blood tests revealed marked thrombo- and leukocytosis. After six injections, the patient had multiple follicular, hyperkeratotic papules (whose appearance resembled the round, sharp grid array on the surface of a nutmeg, known as the "nutmeg-grater sign"), coalescing into scaly, orange-red plaques on her scalp, trunk, and extremities. Skin involvement showed islands of sparing ("nappes claires") in the midst of affected skin areas (Figure 1). Based on the clinical appearance and the negative family history of psoriasis, the diagnosis of classical juvenile (type III) PRP was established, although *CARD14*-gene sequencing was normal.

We treated her with ustekinumab (0.75 mg/kg) and after 8 weeks of anti-IL-12/23 therapy, skin involvement had substantially improved, and her hair started to regrow. Considering the excellent tolerability and efficacy, the ustekinumab dosage was increased to 1.5 mg/kg and combined with acitretin 10 mg two times a week. This regimen further improved the symptoms without adverse effects (Figure 2).

2 | DISCUSSION

PRP is a rare inflammatory skin disease with phenotypic variability and overlapping features with psoriasis. Pathophysiology may involve *CARD 14*-mutations activating the Th17 pathway as well as triggers such as viral or bacterial infections, autoimmune diseases, and neoplasia.[1,2] Treatment of PRP is challenging and evidence from randomized, controlled trials is lacking.[1,2] For both adults and children with insufficient response to topical interventions (emollients, urea, topical steroids), systemic therapy with retinoids is a common first-line approach.[1,2] In contrast to adult patients, there is no clear dose recommendation for

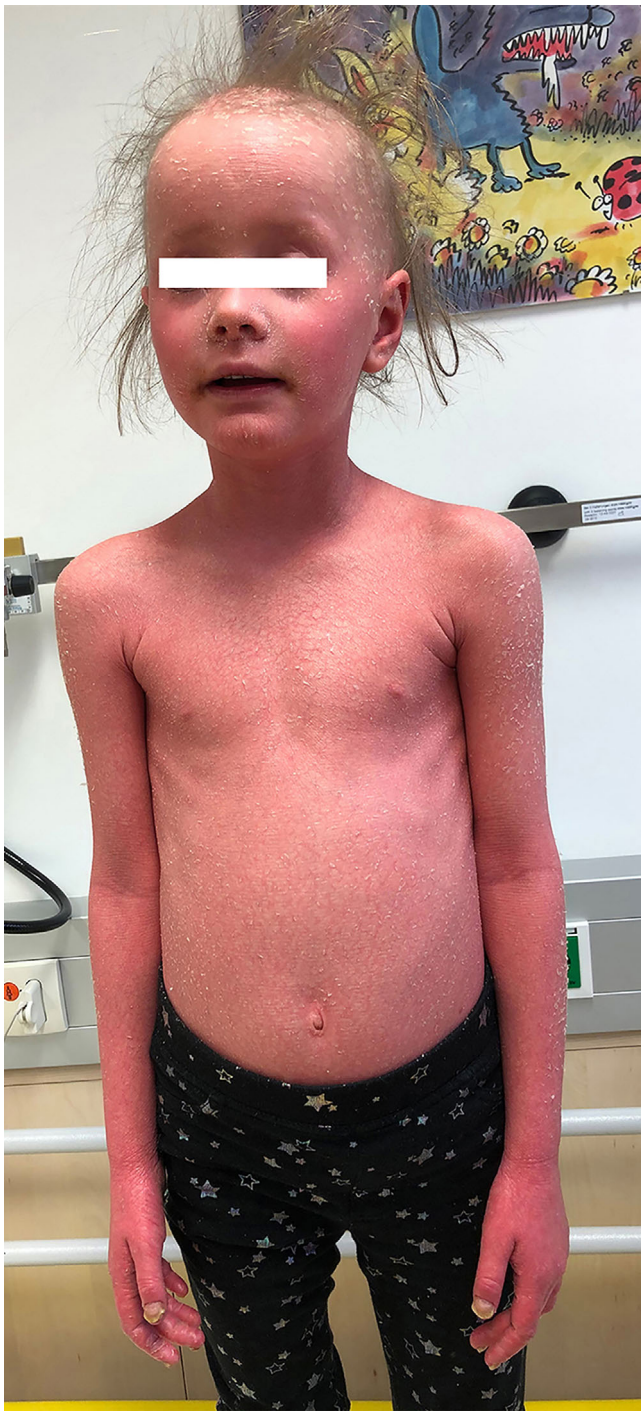

FIGURE 1 Symptomatic exacerbation in the course of the disease with generalized scaly erythroderma and alopecia of scalp, eyebrows, and eyelashes (A). All fingers and toenails presented with onycholysis and discoloration of the nail plate as well as subungual changes. Diffuse palmoplantar hyperkeratosis resulted in painful fissures

younger patients. Sharing molecular, histologic, and clinical characteristics with psoriasis,[1] biologic agents have been used off-label in various subtypes of PRP, including familial and pediatric presentations.[2,3]

Ustekinumab has been used in children suffering from inflammatory bowel disease and *CARD14*-associated papulosquamous eruption (CAPE).[4] We present the use of an anti-interleukin 12/23 monoclonal

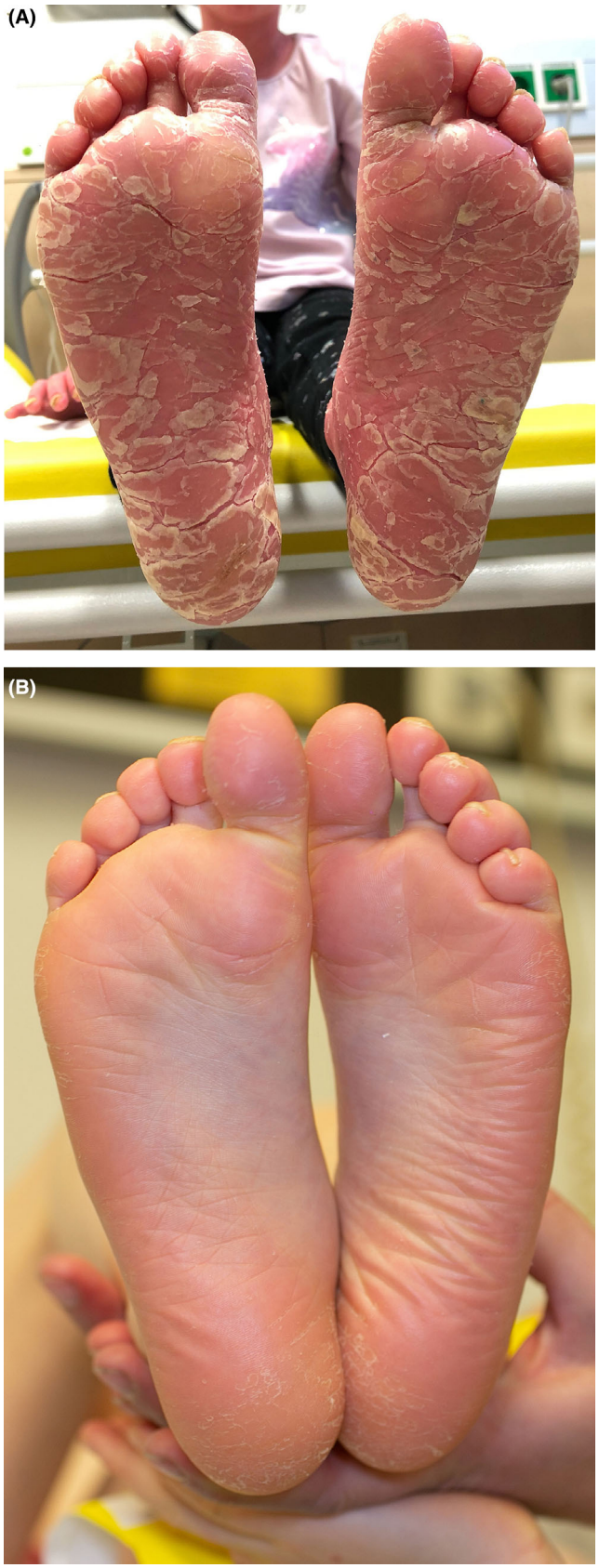

FIGURE 2 Combination therapy of ustekinumab and acitretin was associated with hair regrowth, clinical improvement of skin lesions, as well as regressive palmoplantar hyperkeratosis and nail involvement (before treatment (A) vs. after treatment (B))

antibody therapy in combination with acitretin for the effective treatment of pediatric PRP. In line with other anecdotal reports, ustekinumab proved effective and tolerable in the treatment of refractory PRP.[3,5] Combination with acitretin did not adversely affect the safety profile and may be considered to increase the therapeutic impact, especially on hyperkeratotic lesions, though it is possible that improvement in this patient was from ustekinumab dose or duration alone. Controlled studies are warranted to corroborate our observation.

AUTHOR CONTRIBUTIONS

Katharina Medek, MD (Department of Dermatology, University Hospital of the Paracelsus Medical University Salzburg, 5020 Salzburg, Austria): design and conduct of the study, preparation, drafting of the manuscript, review, and approval of the manuscript, collection, management, analysis, and interpretation of the data; decision to submit the manuscript for publication. **Sylvia Selhofer, MD** (Department of Dermatology, University Hospital of the Paracelsus Medical University Salzburg, 5020 Salzburg, Austria): design and conduct of the study, preparation, drafting of the manuscript, review, and approval of the manuscript, collection, management, analysis, and interpretation of the data; decision to submit the manuscript for publication. **Matthias Buchner, MD** (Department of Pediatrics, University Hospital of the Paracelsus Medical University Salzburg, 5020 Salzburg, Austria): design and conduct of the study, preparation, drafting of the manuscript, review, and approval of the manuscript, collection, management, analysis, and interpretation of the data; decision to submit the manuscript for publication. **Mrowietz Ulrich, MD** (Psoriasis-Center at the Department of Dermatology, University Medical Center Schleswig-Holstein, Kiel, Germany): review and approval of the manuscript, collection, management, analysis, and interpretation of the data; decision to submit the manuscript for publication. **Laimer Martin, MD** (Department of Dermatology, University Hospital of the Paracelsus Medical University Salzburg, 5020 Salzburg, Austria): review and approval of the manuscript, collection, management, analysis, and interpretation of the data; decision to submit the manuscript for publication.

DATA AVAILABILITY STATEMENT

Data sharing not applicable to this article as no datasets were generated or analysed during the current study

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