Widespread erythematous skin eruption

The patient presented with a salmon-colored rash from head to toe. The distinctive clinical presentation and a biopsy pointed to an uncommon diagnosis.

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A 48-YEAR-OLD WOMAN sought care for a widespread pruritic skin eruption that began on her upper back and spread to her arms, lower trunk, and lower legs. She'd had the rash for approximately 2 months and didn't have any systemic symptoms. A course of prednisone prior to her presentation failed to improve the rash. She denied a personal or family history of rheumatologic or dermatologic disease and reported no new medications or exposures.

On physical exam, she was afebrile and her vital signs were normal. The rash had red-to-salmon–colored scaling patches with discrete and coalescing follicular papules. There were prominent islands of sparing (**FIGURE 1**).

The patient's palms were waxy and erythematous and her feet had hyperkeratosis. A complete blood count, comprehensive metabolic panel, and lipid panel were normal. A skin biopsy demonstrated psoriasiform dermatitis with alternating areas of orthokeratosis and parakeratosis (the presence of keratinocyte nuclei within the stratum corneum where nuclei typically aren't found).

- WHAT IS YOUR DIAGNOSIS?
- HOW WOULD YOU TREAT THIS PATIENT?

FIGURE 1
A cephalocaudal rash with prominent islands of sparing

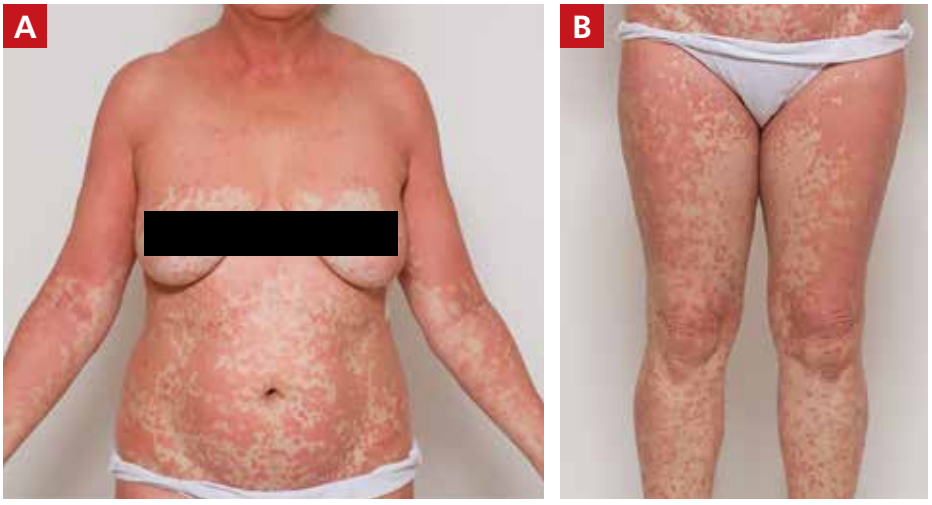

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TABLE

Types of pityriasis rubra pilaris[1,4]

Type	Population	Typical course
I	Adult	Spontaneous remission within 3 years
II	Adult	Chronic course
III	Pediatric	Spontaneous remission within 1 year
IV	Pediatric	Relapsing and remitting
V	Pediatric	Chronic course
VI	HIV-infected	Variable course

HIV, human immunodeficiency virus.

In rare cases, pityriasis rubra pilaris has been associated with internal malignancy and human immunodeficiency virus infection.

Diagnosis: Pityriasis rubra pilaris

The patient was given a diagnosis of pityriasis rubra pilaris (PRP) based on her distinctive clinical presentation. This included the presence of prominent islands of sparing, the red-to-salmon scaling patches with follicular papules, the waxy erythema of her palms, and the cephalocaudal progression of her rash. The patient's skin biopsy findings (in particular, the alternating orthokeratosis/parakeratosis) were also supportive of the diagnosis and helpful to exclude other potential causes of erythroderma (described below).

PRP most often affects middle-aged individuals with an equal sex distribution. The etiology and pathogenesis of PRP are not well understood. In rare cases, it has been associated with internal malignancy and human immunodeficiency virus (HIV) infection.[1,2] PRP may stem from a combination of a dysfunction in vitamin A metabolism, genetic factors, and immune dysregulation.[3] Six types of PRP have been identified; they differ in the way they present and the populations affected (**TABLE**).[1,4]

PRP can be confused with other causes of erythroderma

PRP can cause erythroderma (also known as exfoliative dermatitis), which is the term applied to an erythematous eruption with scaling that covers ≥90% of the body's surface area. Akhyani et al found that PRP is responsible for approximately 8% of all erythrodermas;[5] the other causes of erythroderma are manifestations of numerous conditions, including psoriasis, dermatitis, drug eruptions, and malignancy. The course and prognosis of the erythroderma varies with the underlying condition causing it.[6]

Psoriasis is a common cause of exfoliative dermatitis in adults. Erythroderma may occur in patients with underlying psoriasis after discontinuing, or rapidly tapering, systemic corticosteroids.[7] Because PRP is a papulosquamous eruption, it is often confused with psoriasis.[1,3]

Dermatitis. Several subtypes of dermatitis can be associated with erythroderma. These include atopic, seborrheic, allergic contact, airborne, and photosensitivity dermatitis.

Drug eruptions. Numerous pharmacologic agents have been associated with the development of widespread drug-induced skin eruptions. These eruptions include the severe reaction of toxic epidermal necrolysis, which always involves sloughing of skin.

Malignancy. Both cutaneous T-cell lymphoma (including mycosis fungoides) and internal malignancies can lead to erythroderma.[6,8,9]

PRP has several distinguishing features from other causes of erythroderma. The natural course of classic adult PRP (type 1) is variable, but is typically self-resolving within 3 years of onset. Clinical findings include red-to-salmon–colored follicular hyperkeratosis that forms papules or plaques with scales. There are often prominent islands of central sparing. Palmoplantar keratoderma is also commonly observed. The disease typically spreads in a cephalocaudal fashion and may progress to generalized erythroderma.[1]

Treatment includes oral retinoids

In the initial evaluation of most cases of erythroderma, it is important to perform a skin biopsy (a 4-mm punch is often best) with a request for a rush reading to avoid missing a possibly severe and life-threatening diagnosis. Skin biopsy is often not diagnostic, but may show alternating parakeratosis and orthokeratosis (as in this case). Careful correlation of the histopathologic findings with the clinical presentation is what usually leads to the diagnosis. Obtaining 2 punch biopsies may be helpful if there are multiple morphologies present or if mycosis fungoides is suspected. If the patient is not physiologically stable, hospitalization is warranted.

Oral retinoids (eg, acitretin) are the first-line treatment for PRP. PRP is a rare disease, so the best treatment data available include studies involving small case series. Other treatments include methotrexate and phototherapy, but results are mixed and patient-dependent.[1,3] In fact, some patients have experienced flare-ups when treated with phototherapy; therefore, it is not a commonly used treatment for PRP.

Tumor necrosis factor (TNF)-alpha inhibitors, including infliximab, adalimumab, and etanercept, have been used increasingly with varying degrees of success.[10-12] TNF-alpha inhibitors have a relatively good safety profile and should be considered in refractory cases. If there are associated conditions, such as HIV, treating these may also result in remission.[2]

Our patient was treated with oral acitretin 70 mg/d. At a 3-month follow-up visit, her skin showed signs of partial improvement. The patient was lost to follow-up. JFP

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Cases of classic adult (type 1) pityriasis rubra pilaris typically self-resolve within 3 years of onset.