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Case Letter

Pityriasis rubra pilaris in skin of color

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Dear Editor,

Pityriasis rubra pilaris (PRP) is a rare inflammatory disorder of unknown etiology (Roenneberg and Biedermann, 2018). Herein, we present a diagnostically challenging case of a patient with extensive cutaneous PRP.

A 28-year-old black woman presented with a history of eczema and a 1-year history of numerous fine, pruritic bumps that began on the hands and progressed to involve much of the body surface. A variety of over-the-counter topical treatments were unsuccessful. On physical examination, widespread monomorphic follicular papules were identified on the trunk and extremities with accentuation in areas of friction on the shoulders. Palmoplantar xerosis with hyperkeratosis was also noted. The patient was diagnosed with follicular eczema and lost to follow-up.

Three years later, the patient presented with more severe disease. Scalp flaking and pruritus were present. Diffuse monomorphic follicular keratotic papules on the trunk and extremities were still observed (Fig. 1). Perioral hyperkeratosis and hyperpigmentation with sharply demarcated sparing of the vermilion border were noted. Well-demarcated, symmetrical, mildly erythematous patches with peripheral scaling were identified adjacent to areas of uninvolved skin in the inframammary and inguinal folds (Fig. 2). The hands and feet exhibited xerosis with hyperkeratosis, fissuring, and a few scattered punctate keratotic papules. Nail dystrophy was present.

The diagnosis of PRP was made after bilateral knee punch biopsies demonstrated psoriasiform acanthosis, mild spongiosis, follicular plugging, and prominent hyperkeratosis with alternating parakeratosis and orthokeratosis. Shortly after diagnosis, the patient became pregnant, and topical corticosteroids were prescribed with the intent to begin biologic therapy after delivery. Little improvement was noted at her 4-month follow-up visit.

PRP often begins with reddish-brown, hyperkeratotic follicular papules that coalesce into salmon-colored, scaly patches and plaques adjacent to normal, uninvolved skin (islands of sparing). Scaly erythema of the scalp and prominent orange-yellow waxy palmoplantar keratoderma are also characteristic (Roenneberg and Biedermann, 2018; Ross et al., 2016). However, clinical presentations may vary widely, ranging from extremity-limited disease to diffuse erythroderma (Roenneberg and Biedermann, 2018). Furthermore, cutaneous findings at early stages may be especially variable and only later evolve into more classic features of PRP, frequently leading to initial misdiagnoses and multiple biopsies (Engelmann et al., 2019). Due to shared features with eczematous and other papulosquamous disorders, such as psoriasis, diagnostic confirmation is challenging and requires the presence of both clinical and classic histopathologic features (Engelmann et al., 2019; Roenneberg and Biedermann, 2018; Ross et al., 2016).

Hyperkeratotic follicular papules, palmoplantar disease, and scaly patches with islands of sparing were observed in our patient. However, the clinical diagnosis remained unclear because the classic coloring was absent throughout her disease course. This altered coloration may be attributed to this patient's dark skin, a feature shared by other dermatologic conditions in this patient population (Lester et al., 2019). These patients are also more likely to exhibit follicular variants of eczematous eruptions and other disorders on the differential (Silverberg, 2017), which further contributes to the difficult and initially missed diagnosis of follicular eczema in our patient.

Classic coloration and patterns play a central role in the timely recognition of dermatologic disease (Adelekun et al., 2021; Lester et al., 2019). Providers must be aware that dermatologic conditions may manifest uncharacteristically in skin of color, thereby exacerbating the diagnostic difficulty of already rare and challenging diseases. Increased representation of this population in the dermatologic literature and textbooks, as well as additional teaching and clinical exposure throughout residency, may facilitate improved diagnostic accuracy among clinicians (Adelekun et al., 2021; Lester et al., 2019), leading to reduced misdiagnoses and improved outcomes.

Conflicts of interest

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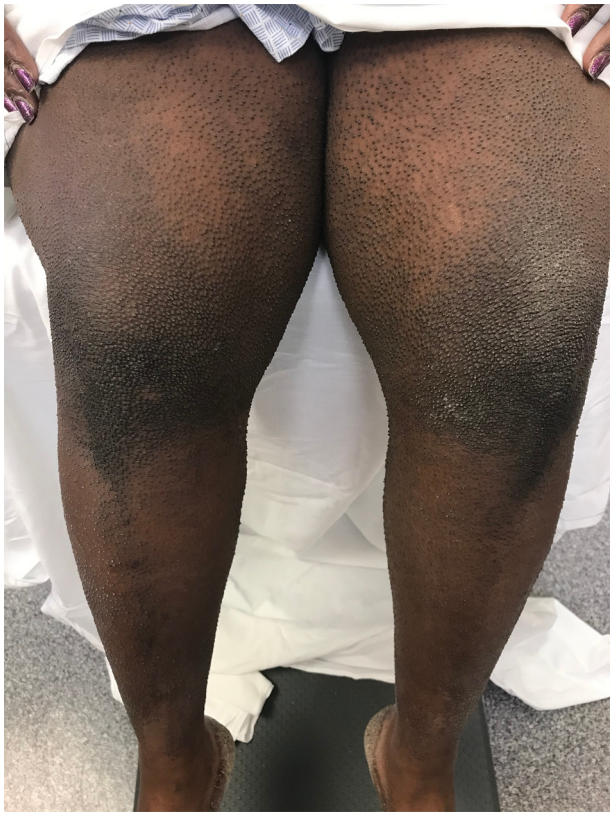

Fig. 1. Monomorphic follicular papules on the lower extremities.

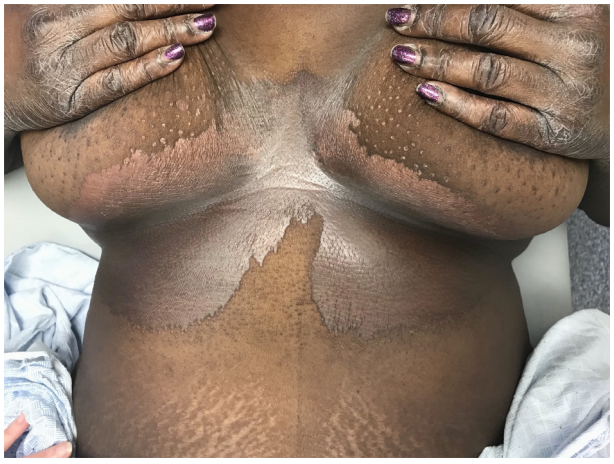

Fig. 2. Well-demarcated, mildly erythematous patches in the inframammary folds with islands of sparing.

Study approval

The author(s) confirm that any aspect of the work covered in this manuscript that has involved human patients has been conducted with the ethical approval of all relevant bodies.

References

Adelekun A, Onyekaba G, Lipoff JB. Skin color in dermatology textbooks: an updated evaluation and analysis. J Am Acad Dermatol 2021;84(1):194–6.
Engelmann C, Elsner P, Miguel D. Treatment of pityriasis rubra pilaris type I: A systematic review. Eur J Dermatol 2019;29(5):524–37.
Lester JC, Taylor SC, Chren MM. Under-representation of skin of colour in dermatology images: not just an educational issue. Br J Dermatol 2019;180(6):1521–2.
Roenneberg S, Biedermann T. Pityriasis rubra pilaris: algorithms for diagnosis and treatment. J Eur Acad Dermatol Venereol 2018;32(6):889–98.
Ross NA, Chung HJ, Li Q, Andrews JP, Keller MS, Uitto J. Epidemiologic, clinicopathologic, diagnostic, and management challenges of pityriasis rubra pilaris: a case series of 100 patients. JAMA Dermatol 2016;152(6):670–5.
Silverberg NB. Typical and atypical clinical appearance of atopic dermatitis. Clin Dermatol 2017;35(4):354–9.