Pityriasis rubra pilaris–like eruption in the setting of transient acantholytic dermatosis

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INTRODUCTION

Transient acantholytic dermatosis (TAD; also known as *Grover disease*) often presents as pruritic erythematous papules on the chest or back, most frequently seen in older men. Histopathology is significant for acantholysis and dyskeratosis.[1] Pityriasis rubra pilaris (PRP) is a rare condition that classically presents with keratotic papules and perifollicular erythema that coalesce to form generalized erythroderma with islands of uninvolved skin.[2] Although alternating vertical and horizontal orthokeratosis and parakeratosis with follicular plugging are the most frequently described histopathologic findings in PRP, both focal and extensive acantholysis have also been increasingly described in this condition, leading to the notion of a potential overlap between TAD and PRP.[3-5] We present the case of a 61-year-old woman with a clinical and histologic diagnosis of TAD, who subsequently had a PRP-like eruption with corresponding histopathology.

Abbreviations used:

PRP: pityriasis rubra pilaris
TAD: transient acantholytic dermatosis

CASE REPORT

A 61-year-old woman presented to the dermatology clinic with multiple monomorphic, erythematous, flat-topped papules varying from 2 to 5 mm in size, involving the trunk and bilateral upper and lower extremities. She had a nearly 10-year history of an eruption diagnosed as TAD. The rash had begun to worsen despite topical therapy with triamcinolone 0.1% cream and lotion. She underwent 33 sessions of narrow-band ultraviolet B phototherapy, which was discontinued because of burning sensations, and her rash continued to worsen, involving her trunk, upper and lower extremities, dorsal hands, and feet. There was relative sparing of the face, scalp, palms, and soles (Fig 1). Biopsy was performed from the abdomen, which found mild spongiosis with acantholysis and dyskeratosis, consistent with TAD (Fig 2).

The patient was started on acitretin, 10 mg daily, which was discontinued because of severe pruritus. She was then switched to tretinoin 0.1% cream and minocycline, 100 mg twice daily. Her rash continued to worsen, until finally she presented with diffuse erythroderma involving her trunk and extremities, encompassing 80% of her body surface area, with rare islands of sparing (Fig 3). Repeat biopsy was performed from the left thigh, which found psoriasiform hyperplasia with alternating horizontal and vertical parakeratosis and orthokeratosis, consistent with PRP (Fig 4). The patient was then started on methotrexate, 15 mg weekly, with significant improvement, and was nearly clear after 6 months of methotrexate therapy. Her symptoms remain well controlled on low-dose methotrexate therapy at the time of publication 2 years later.

DISCUSSION

TAD is characterized by discrete, pruritic erythematous papules and papulovesicles, commonly found on the trunk of older white men.[1,6] Lesions can vary from few to several dozen and may be transient or chronic.[1] Histologically, TAD presents as acantholysis frequently associated with dyskeratosis and may

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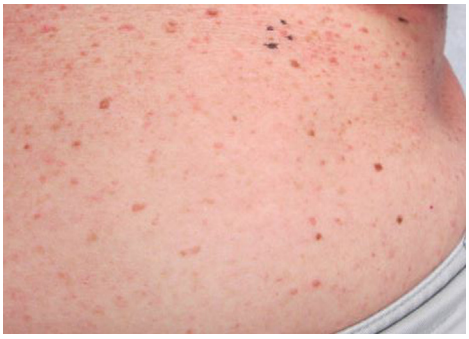

Fig 1. Initial presentation of monomorphic, erythematous papules involving the trunk, clinically consistent with transient acantholytic dermatosis.

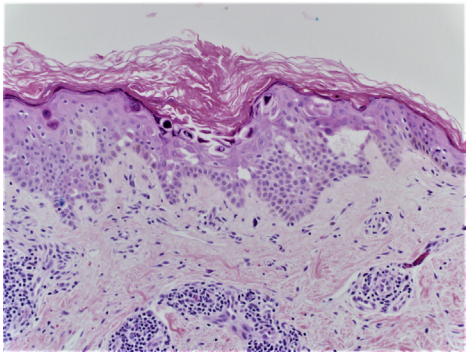

Fig 2. Histopathology of initial biopsy of the abdomen, which shows mild spongiotic dermatitis with acantholysis and dyskeratosis, consistent with transient acantholytic dermatosis.

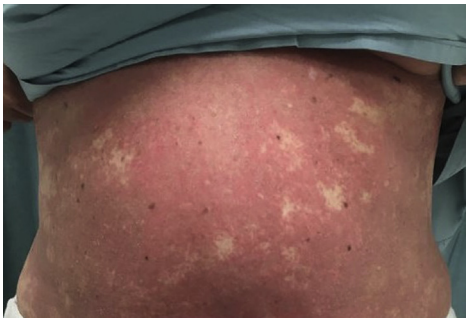

Fig 3. Subsequent presentation of diffuse erythema with islands of sparing, clinically resembling PRP.

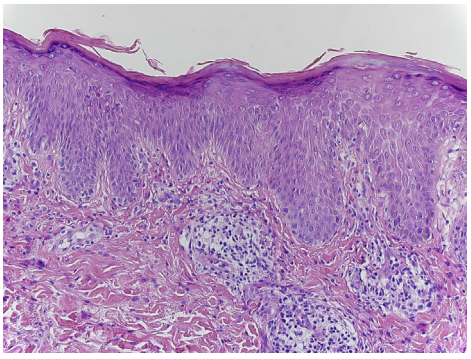

Fig 4. Histopathology of subsequent biopsy, which shows psoriasiform hyperplasia with alternating horizontal and vertical parakeratosis and orthokeratosis consistent with PRP.

mimic Darier disease, Hailey-Hailey disease, or pemphigus.[1,7] The exact etiology of TAD is unknown, but it can be associated with excessive heat, perspiration, ultraviolet exposure, and malignancy.[1]

PRP is a rare condition with similarly unknown etiology. The classic type (type 1) presents as pruritic follicular keratotic papules that coalesce into erythematous or salmon-colored plaques with intervening areas of uninvolved skin.[2] The lesions usually start on the face and scalp and progress caudally, and hyperkeratosis of the palms and soles may develop.[2] The condition frequently progresses to erythroderma.[2] Histopathologic findings of PRP often include psoriasiform hyperplasia with alternating orthokeratosis and parakeratosis in both vertical and horizontal directions and follicular plugging.[2] Acantholytic dyskeratosis in PRP was first described in 1989 by Kao and Sulica,[8] and has increasingly been reported as a potential histological feature of PRP.[3,4,8] As in TAD, variable patterns of acantholysis including suprabasal, midepidermal, subcorneal and follicular, with or without dyskeratosis, have been reported, often in cases of PRP that were originally diagnosed as a different disease.[3-5,8] Initial presentation on the trunk, rather than the head and neck, has been speculated to correlate with acantholytic cases of PRP.[3] PRP can be quite resistant to treatment; methotrexate and retinoids are recommended as first-line systemic options.[2]

Although cases of PRP with TAD-like histologic findings have been described, reports of a PRP-like eruption occurring in the setting of TAD are extremely rare, and a similar case has been described only once before by Cowen and O'Keefe in 1997.[3,9] In that case, the rash that was initially diagnosed as TAD had only been present for 2 weeks, and evolved into PRP over the course of 1 week, suggesting that what was initially diagnosed as TAD was more likely an early presentation of PRP. In our case, the patient had a longstanding history of TAD for several years before the development of a PRP-like eruption. Our case highlights potential similarities between TAD and PRP and the possibility of an overlap between these two diseases that were previously thought to be distinct.

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