A case of resistant pityriasis rubra pilaris responsive to combination acitretin and ustekinumab

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Abstract

Pityriasis rubra pilaris is a rare psoriasiform dermatitis. Treatment has been adopted from psoriasis protocols, with topical corticosteroids and systemic retinoids as first-line agents, followed by escalation to biologics for recalcitrant disease. We report a patient with resistant pityriasis rubra pilaris who dramatically improved with acitretin and ustekinumab, a combination not well documented in the literature. The purpose of this letter is to emphasize the potential benefit of dual therapy in patients who fail traditional pityriasis rubra pilaris treatment regimens.

Keywords: pityriasis rubra pilaris, psoriasiform dermatitis, ustekinumab

Introduction

Pityriasis rubra pilaris (PRP) is a rare psoriasiform dermatitis presenting clinically with pink-orange, follicular hyperkeratotic papules and plaques with islands of normal skin and palmoplantar keratoderma [1]. Histologic features commonly vary early in the disease process, requiring multiple biopsies to confirm the diagnosis [2]. Treatment begins with topical corticosteroids, phototherapy, and systemic retinoids and escalates to immunomodulatory agents and biologics for resistant disease [3, 4]. The role of dual therapy is not well documented in this difficult-to-treat condition, but as we will show, may provide longer-term symptom relief and minimize flares in select patients.

Case Synopsis

An 81-year-old man presented with a five-month history of a "pin-pricking" rash consisting of orange-red folliculocentric papules and plaques on his trunk, extremities, and scalp covering a body surface area (BSA) of 65%. He also had hyperkeratosis of palms, soles, and fingernails with onycholysis. Skin biopsy showed many features consistent with PRP, including 'checkerboard' parakeratosis and orthokeratosis, but lacked follicular hyperkeratosis and plugging. This non-classic histology was not surprising, especially early in the disease process when PRP can vary widely. The diagnosis was made on the basis of strong clinical presentation. The patient experienced improvement with acitretin titrated up to 50mg daily but developed a subsequent flare involving 70% BSA when the dose was lowered owing to palm and sole hypersensitivity (**Figure 1**). Acitretin 50mg daily was reinstated and phototherapy 2-3 times weekly was added to his regimen. Again, the acitretin was lowered owing to undesired symptoms including hair loss, nail dystrophy, gingival pain, and a "sticky-skin" sensation. At this time, the patient began combination therapy with low-dose acitretin 35mg daily and an injection of ustekinumab 90mg given every three months. Four months after starting this

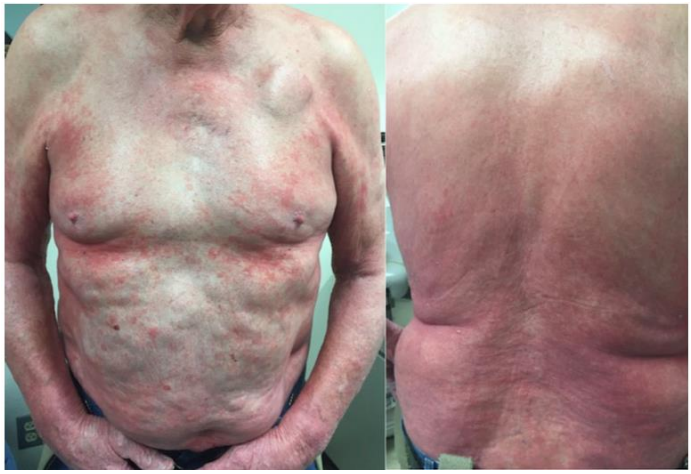

Figure 1. *Type I Pityriasis rubra pilaris rash with orange-red folliculocentric papules, coalescing into plaques with islands of spared skin covering 70% body surface area.*

regimen, the patient had improvement in nail dystrophy and palmoplantar hyperkeratosis in addition to near complete resolution of the rash with only 6% BSA remaining. His disease remains stable and he has not developed a flare for one year since beginning dual therapy (**Figure 2**).

Case Discussion

The treatment of PRP was originally adopted from psoriasis protocols, with systemic retinoids, like acitretin, as first-line agents. Ustekinumab, an IL-12/IL-23 monoclonal antibody targeting the T_H17 cell axis, is currently approved for use in plaque psoriasis [3, 4]. Recent studies have shown promising results for its use in PRP, likely related to similarities in cytokine activation seen in both PRP and psoriasis [3, 4]. A 2017 systemic review of PRP therapies found a superior response with ustekinumab (62.5%) compared to acitretin (24.7%) as well as fewer side effects reported when both agents were compared as monotherapy [5]. There are few reports suggesting benefit of acitretin and ustekinumab dual therapy [6], but because of the infrequency of PRP and the small number of published studies, guidelines for combination therapy have not been described. Our patient benefitted from combination low-dose acitretin with ustekinumab when higher-dose acitretin was not tolerated. We feel that select patients with resistant disease or intolerance to side effects of traditional medications should be considered for similar combination therapy provided that no contraindication exists. More research is needed to understand the potential benefits of combination therapy and we encourage providers to report on the uses of combination therapy in PRP.

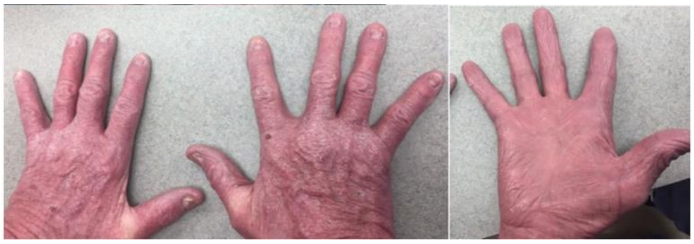

Figure 2. *Cutaneous and nail improvement after one injection of ustekinumab.*

Conclusion

Pityriasis rubra pilaris is often a challenging, resistant psoriasiform dermatitis. Clinical and histopathologic similarities in PRP and psoriasis account for the use of psoriasis treatment protocols in this condition. Dual therapy with acitretin and ustekinumab was successful in our patient who had resistant disease and who was intolerant to higher-dose acitretin monotherapy. Combination therapy should be considered for similar patients. Further research into PRP combination therapy is recommended.

Potential conflicts of interest

The authors declare no conflicts of interests.

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