ABSTRACT

Introduction: Pityriasis rubra pilaris (PRP) is an inflammatory eruption of unknown origin; rare cases of COVID-19 vaccine-induced PRP have been reported. Here, we present a case of COVID-19 vaccine-induced pityriasis rubra pilaris inadequately managed with acitretin, successfully treated with the IL-4/IL-13 inhibitor dupilumab.

Case Report: A 76-year-old male presented to an outpatient dermatology clinic with a 2-month history of a profoundly pruritic worsening rash characterized by large, orange-salmon-colored follicular papules with scale coalescing into plaques covering approximately 80% of the body surface area. The plaques had well-defined borders and multiple islands of sparing characteristic of PRP. Multiple therapies were trialed with no improvement, including oral prednisone, mycophenolate, topical corticosteroids, antiparasitics, antifungals, doxepin, and UVB treatments. A trial of oral acitretin resulted in improvement of the skin plaques and keratoderma, but the patient remained uncontrollably pruritic. Dupilumab was initiated which provided rapid relief, and the patient has remained clear for several months.

Conclusion: Clinicians should be aware of dupilumab’s potential for effective treatment of PRP with recalcitrant pruritus.

INTRODUCTION

Pityriasis rubra pilaris (PRP) is a rare, inflammatory, papulosquamous eruption of unknown origin. While its etiology remains unclear, medications, viral infections, autoimmune mechanisms, vitamin A deficiency, and genetic factors have all been proposed as potential triggers. PRP can involve the entire body but commonly affects palms and soles. PRP affects all races, ages, and genders equally. Since the COVID-19 pandemic and widespread disbursement of COVID-19 vaccinations, multiple cases of COVID-19 vaccination-induced pityriasis rubra pilaris have been reported. Here, we present a case of COVID-19 vaccine-induced pityriasis rubra pilaris inadequately managed with acitretin, successfully treated with the IL-4/IL-13 inhibitor dupilumab.

CASE REPORT

A 76-year-old male presented to an outpatient dermatology clinic with a 2-month history of a profoundly pruritic worsening rash that began on the upper back and progressed to involve the entire body. He presented with...
large, orange-salmon-colored follicular papules with scale coalescing into plaques covering approximately 80% of the body surface area (Figures 1-3). The plaques had well-defined borders and multiple islands of sparing. The patient also had prominent palmoplantar keratoderma. The patient reported severe generalized pruritus that prevented him from sleeping at night, to the point that he was tearful and expressed suicidal ideation. His past medical history was not significant for any chronic conditions, and he did not take any medications. Of note, he had received one dose of the Pfizer-BioNTech COVID-19 vaccination three weeks prior to the eruption and he had no other recent travels or exposures. One outside biopsy was performed which showed spongiosis with eosinophils, favoring an eczematous process. Two weeks later, at our institution, two punch biopsies were taken from the different areas on the trunk, which demonstrated psoriasiform spongiotic dermatitis. All laboratory testing came back within normal limits, including complete blood count, complete metabolic panel, QuantiFERON gold tuberculosis testing, hepatitis B, hepatitis C, and HIV serologies. Multiple therapies were trialed with no improvement, including oral prednisone, mycophenolate, topical corticosteroids, antiparasitics, antifungals, doxepin, and UVB treatments. Two UVB treatments resulted in significant worsening in skin pain, pruritus, and significant bilateral pedal edema requiring two weeks of oral furosemide. Given the distinct clinical appearance of the skin findings, the slides were reviewed and the dermatopathologist agreed that they showed findings compatible with a spongiotic pityriasis rubra pilaris. A trial of oral acitretin resulted in improvement of the skin plaques and keratoderma, but the patient remained uncontrollably pruritic. Dupilumab was initiated which provided rapid relief, and the patient has remained clear for several months.

**DISCUSSION**

To our knowledge, this is the first report of Covid vaccine-induced pityriasis rubra pilaris that was successfully treated with dupilumab (Dupixent) for recalcitrant pruritus. The mechanism underlying Covid vaccine-induced PRP is unclear. Dupilumab is FDA-approved in dermatology for the treatment of atopic dermatitis, chronic spontaneous urticaria, and prurigo nodularis. It has also been reported off-label for alopecia areata, hand dermatitis, and allergic contact dermatitis. Dupilumab is a human monoclonal antibody that selectively blocks interleukin-4 (IL-4) and interleukin-13 (IL-13) receptors, which inhibits downstream signaling of the JAK/STAT pathway. Dupilumab treats atopic dermatitis by effectively inhibiting Th2 cytokine differentiation and IgE class switching. Dupilumab has an overall excellent safety profile and few adverse effects due to its targeted mechanism. Another benefit of this medication is that it does not require routine lab monitoring.

The exact pathophysiology of pityriasis rubra pilaris is unknown, making it difficult to ascertain why dupilumab was effective in this patient at a molecular level. Dupilumab was chosen as therapy due to chief complaint of pruritus and histopathologic findings of spongiotic dermatitis. IL-4 and IL-13 are key cytokine drivers of pruritus; although the exact mechanism is not completely understood, both IL-4 and IL-13 levels have been found to be elevated in skin and serum of patients with atopic dermatitis. Injections of IL-4 and IL-13 have also been shown to increase itching in mice. Therefore,
Figure 1. Orange-salmon-colored follicular papules with scale coalescing into plaques along the bilateral shins and dorsal feet

Figure 2. Orange-red plaques and bright yellow palmar keratoderma
dupilumab, a dual IL-4/IL-13 blocker, was a logical treatment choice for our patient with severe pruritus. Further studies are needed to investigate the utility of dupilumab for pityriasis rubra pilaris and accompanying pruritus.

CONCLUSION

We report a rare case of COVID-19 vaccine-induced pityriasis rubra pilaris inadequately treated with acitretin and successfully treated with dupilumab, an IL-4/IL-13 inhibitor. Clinicians should be aware of dupilumab’s potential for effective treatment of PRP with recalcitrant pruritus.

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