PITYRIASIS RUBRA PILARIS
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ABSTRACT
Pityriasis rubra pilaris (PRP) is an uncommon skin condition that was described greater than 150 years ago. It was initially thought to be a variant of psoriasis due to sharing some clinical and histopathologic characteristics. With time PRP has been well described and differentiated as a separate condition with various morphologic and pathologic features to aid the physician in diagnosis. Making the diagnosis of PRP can be challenging and treatment can likewise have its difficulties. The exact etiology of PRP has yet to be unfolded. While many mechanisms have been proposed, none of them has been fully substantiated. This case described the clinical course of a patient who presented to the dermatology clinic complaining of a generalized pruritic rash that had been present intermittently over a year’s time. Following clinical and histopathologic correlation, the patient was diagnosed with PRP. Herein we present a rare case while highlighting the key features necessary to distinguish PRP from psoriasis and offer a brief overview of treatment options.

INTRODUCTION
Pityriasis rubra pilaris (PRP) was first introduced by Claudius Tarral in 1835 listed under “general psoriasis.” It was fully described in 1857 by Deverge who named it Pityriasis pilaris leading to the eponym Devergie’s disease.1 In 1889 it received the full name PRP which has since been used to reference the condition.2 PRP is an uncommon inflammatory papulosquamous skin disorder characterized by follicular hyperkeratotic papules on an erythematous base which coalesce into large scaly plaques. Lesions often progress into generalized erythroderma along with palmoplantar hyperkeratosis. The characteristic orange-red color and waxy nature of the palmar lesions help clinical suspicion for the condition. Differential diagnoses include psoriasis, hypersensitivity reaction, seborrhoeic dermatitis, lichen planopilaris, follicular eczema, cutaneous T-cell lymphoma, subcutaneous lupus erythematosus, and erythroderma progressiva symmetrica.4 Since its discovery, it has been particularly challenging for the physician to differentiate PRP from psoriasis clinically. Features that assist in distinguishing PRP from psoriasis include “islands” of sparing within the erythroderma, follicular keratotic plugs, and an orange hue of the involved skin areas.2

CASE PRESENTATION
49 year old male with a past medical history of hypertension presents to the dermatology clinic complaining of an itchy, burning full body rash that has come and gone intermittently over the last year. Associated symptoms include swollen and painful hands. Oral prednisone helps relieve the rash temporarily but upon cessation the rash returns immediately. He has been to multiple dermatologists in the past for this same rash as well as an allergic patch testing done previously was only positive for cat litter. The patient cannot recall if a biopsy has ever been taken. Denies any new clothing, detergents, medications, fragrances, dyes, or other skin products in recent months.

PHYSICAL EXAM
Vitals: T 98.6 P 70 bpm RR 14 BP 130/80 mmHg BMI 29
Skin: Generalized erythema with superficial scaling, and follicular hyperkeratotic papules diffusely on trunk and extremities with 3-5cm islands of sparing scattered throughout. Hyperkeratotic palms and soles with fissuring. (see photos 1 and 2)

DISCUSSION
Despite having been described more than 150 years ago, the etiology of PRP remains unknown. This is attributed to the low prevalence of the disease and hence minimal clinical studies. Multiple potential mechanisms have been proposed but none have yet to be substantiated. Historically among these are abnormal vitamin A metabolism of the skin, autoimmunity, association with internal malignancies, and infections.2 A recent study identified a possible role of the IL-23-T17 axis in PRP which provides some rationale in targeting this pathway, though more data is needed to further substantiate the evidence.2 Incidence is estimated to be 1 in 400,000 and the exact prevalence is unknown.3 PRP occurs equally in both genders with a bimodal distribution with peaks in the first and then sixth to seventh decades.2 Historically, PRP has been described as a self-limiting disease with spontaneous remission in 2-3 years from onset. A recent study found the mean length of skin manifestation to be 84 months with a range of 4 to 516 months.5 These findings demonstrate a wide range of variability in the course of the disease and that the condition can easily persist beyond the commonly reported 2-3 year self remission time.

Diagnosis of PRP is best made with clinical and histopathologic correlation.3 Histologic findings include an acanthotic epidermis with alternating orthokeratotic and parakeratotic keratinocytes in both horizontal and vertical directions (checkerboard pattern). Other features include irregular hyperkeratosis, broad rete ridges, narrow dermal papillae, follicular plugging, a thin stratum granulosum, and lymphohistocytic perivascular infiltrate in the underlying dermis.2 Griffiths originally classified PRP into five different types based on clinical features, age of onset, and prognosis(see table 1). A sixth type has since been added. Making the diagnosis of PRP can be challenging, and unfortunately the treatment is no less of an issue.6 A myriad of topical and systemic treatments have been used with varying levels of success. While no standardized approach has been established, systemic retinoids are considered to be first line.

Table 1
<table>
<thead>
<tr>
<th>Type</th>
<th>Age of onset</th>
<th>% of cases</th>
<th>Manifestations</th>
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<tbody>
<tr>
<td>I (classical adult)</td>
<td>Adult</td>
<td>55</td>
<td>Red-orange plaques with islands of sparing, begins on head and neck then spread</td>
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<tr>
<td>II (atypical)</td>
<td>Adult</td>
<td>5</td>
<td>5-50 y/o</td>
</tr>
<tr>
<td>III (classical juvenile)</td>
<td>3-12 y/o</td>
<td>25</td>
<td>Erythema and follicular papules primarily affecting elbows and knees</td>
</tr>
<tr>
<td>IV (pruritic juvenile)</td>
<td>0-4 y/o</td>
<td>5</td>
<td>Predominantly follicular hyperkeratosis, accounts for most familial cases of PRP</td>
</tr>
<tr>
<td>V (HIV associated)</td>
<td>Variable</td>
<td>Variable</td>
<td>Similar to type 1</td>
</tr>
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REFERENCES

CASE CONCLUSION
Patient is currently undergoing treatment with acitretin 25mg twice a day by mouth and cyclosporine modified 150mg twice a day by mouth. Overall his condition has greatly improved but he continues to have occasional relapses. He continues to follow up with the dermatology clinic on a regular basis for frequent lab monitoring due to the potential medication adverse reactions.

TAKE HOME POINTS
1. PRP is an uncommon inflammatory condition with an unknown etiology.
2. Diagnosis includes clinical and histopathologic correlation.
3. Must be differentiated from psoriasis.

ACKNOWLEDGEMENTS
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