A case of recurrent and paraneoplastic pityriasis rubra pilaris

Olivia Lamberg, BS,[a] Severine Cao, MD,[b] Stephanie Sabater-Geib, PA-C,[c] Lori Lowe, MD,[b,d] and James Elder, MD, PhD[b,e]
Ann Arbor and Portage, Michigan

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INTRODUCTION

Pityriasis rubra pilaris (PRP) is an idiopathic papulosquamous disorder characterized by follicularly-based papules that coalesce into large confluent plaques, often progressing to erythroderma with distinctive areas of spared skin.[1] Six subtypes of PRP have been described. Type I presents with classic features and runs a self-limited course, with 80% experiencing spontaneous remission within 1 to3 years. Type II is characterized by ichthyosiform dermatitis and a more chronic course of 20 years or more, with less than 20% experiencing remission within 3 years.[1,2] With either type, recurrences have not been reported. While typically idiopathic, rare cases of paraneoplastic PRP have been reported occurring predominantly in the setting of solid organ malignancy.[3] We report a case of a 72-year-old man with recurrent PRP in the setting of lung adenocarcinoma.

Abbreviation used:

PRP: Pityriasis rubra pilaris

CASE REPORT

A 65-year-old man presented with a 2-week history of worsening rash that began on his posterior neck and spread caudally to involve the back, chest, and extremities (Fig 1). Examination revealed confluent erythematous and scaly plaques involving approximately 90% of his total body surface area with islands of sparing on the proximal upper extremities. There was waxy keratoderma of the palms and soles. Punch biopsy demonstrated subacute spongiotic dermatitis with parakeratotic scale and superficial perivascular lymphocytic inflammation, compatible with PRP. He was started on acitretin 25 mg daily with complete resolution after 4 years, at the age of 69. Acitretin was discontinued and at a follow-up 6 months after he remained rash-free.

At age 72, the patient presented with a 2-month history of rash that had started on his neck and spread caudally. Prior to this and for the intervening 3 years, the patient reports his skin had been clear. Examination again revealed confluent erythematous scaly plaques affecting 90% of his total body surface area with islands of sparing and waxy palmoplantar keratoderma (Figs 2 and 3). Repeat skin biopsy demonstrated chronic dermatitis with alternating or "checkerboard" parakeratosis and focal follicular hyperkeratosis (Fig 4). Given the clinical and histologic findings, the patient was diagnosed with recurrent PRP. He was prescribed acitretin 25 mg daily but has not been compliant with this regimen.

On review of his history, it was noted that 6 years after his first episode of PRP and 1 year prior to his PRP recurrence, at age 71, he had been diagnosed with lung adenocarcinoma in the setting of a 45-pack year history of smoking and underwent resection via video-assisted thoracoscopic surgery. Due to poor baseline pulmonary function, complete resection was not possible. He received combination radio- and chemotherapy with good response. However, 7 months later and 3 months

From the University of Michigan Medical School, Ann Arbor[a]; Department of Dermatology, University of Michigan Medical School, Ann Arbor[b]; Southwest Michigan Dermatology, Portage[c]; Department of Pathology, and University of Michigan Medical School, Ann Arbor[d]; Veterans Affairs Ann Arbor Healthcare System.[e]

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Correspondence to: James T. Elder, MD, PhD, University of Michigan, Department of Dermatology, 1910 Taubman Center, 1500 East Medical Center Drive, Ann Arbor, MI 48109-5314. E-mail: jelder@med.umich.edu.

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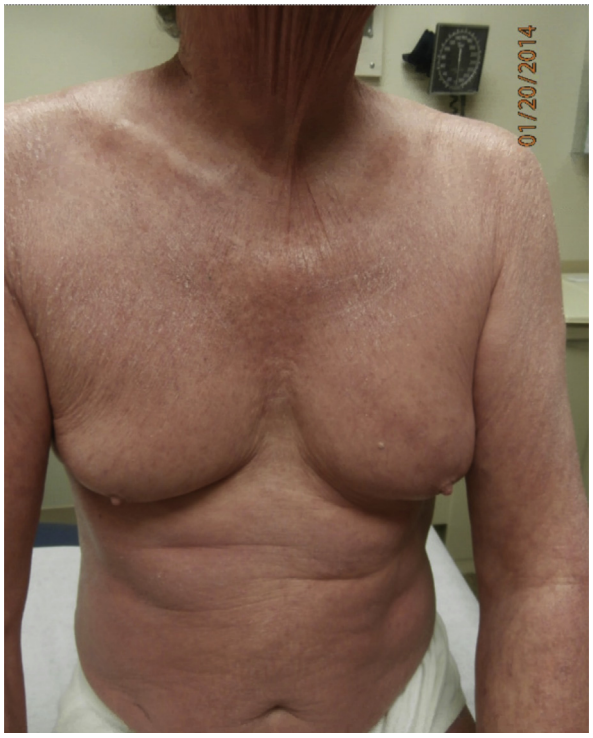

Fig 1. Clinical photo on initial presentation showing confluent erythematous and scaly thin plaques encompassing the chest, trunk, and upper extremities.

prior to the PRP recurrence, he endorsed a 50-pound weight loss at an oncology follow-up visit and positron emission tomography identified uptake in the left lung concerning for recurrence. The patient elected to monitor his disease and is undergoing serial computed tomography scans which demonstrate stability at the time of this writing. His PRP persists.

DISCUSSION

The vast majority of cases of PRP are sporadic, do not have a familial history, and have an unknown etiology.[2,4] The classic adult form, type I, is the most common and presents with erythroderma and palmoplantar keratoderma, resolving in 1-3 years in 80% of cases.[2] Our patient followed the presentation and course of type I PRP on his initial presentation, based on the characteristic findings of erythroderma with islands of sparing and waxy palmoplantar keratoderma that completely resolved in 4 years. It was unlikely that he had type II PRP, based on his self-limited course and absence of ichthyosiform dermatitis.

Our patient presented 3 years following PRP remission with a rash similar to his initial clinical presentation. Based on our clinical documentation and patient reported history of remission, the repeat diagnosis of PRP was consistent with recurrence. To our knowledge, recurrence has not been described for Type I PRP. On review of the literature, only 2 cases of PRP recurrence have been reported, both of which were in children with juvenile type III PRP.[5,6] In both cases, the recurrence was preceded by a viral infection.

In our patient, it is interesting to consider the association of malignancy. The recurrence of PRP occurred 3 months after a positron emission tomography scan suggested recurrence of his lung adenocarcinoma. The timing of the PRP recurrence together with probable active cancer fulfills Curth postulates of paraneoplastic disease.[7] Paraneoplastic PRP has been described but is relatively rare. It has been shown to occur with a variety of solid organ malignancies, including lung adenocarcinoma.[3] In these cases, the PRP diagnosis occurred 1 to 11 months before the diagnosis of cancer. While the timing in our patient differs from prior case reports, it is consistent with what has been described with other paraneoplastic syndromes, which can occur within 5 years of cancer diagnosis.[8]

The pathogenesis of paraneoplastic PRP is poorly understood. It is thought that paraneoplastic skin conditions may arise from immunologic responses triggered by the underlying cancer that cross-react with the skin.[9] Mutations in CARD14, a protein that regulates keratinocyte responses to cytokine signals,[10] have been identified in both familial and sporadic PRP.[4] It could be hypothesized that immune activation from a neoplasm could exaggerate the keratinocyte response when a CARD14 variant is present and provoke recurrence of a skin condition otherwise expected to remain in remission.

In summary, while Type I PRP is usually an idiopathic and self-limited process, our case provides support that Type I PRP may not only be recurrent but also paraneoplastic. It may be prudent to obtain age-appropriate cancer screening in patients with newly diagnosed Type I PRP. In the rare case of recurrent PRP, suspicion for an underlying malignancy should be even higher. In addition to age-appropriate cancer screening, these patients may require an additional malignancy work-up that includes urinalysis, fecal occult blood test, and chest radiograph. Depending on the initial work-up, there should be a low threshold for computed tomography scans of the chest, abdomen, and pelvis.[11]

Conflicts of interest

None disclosed.

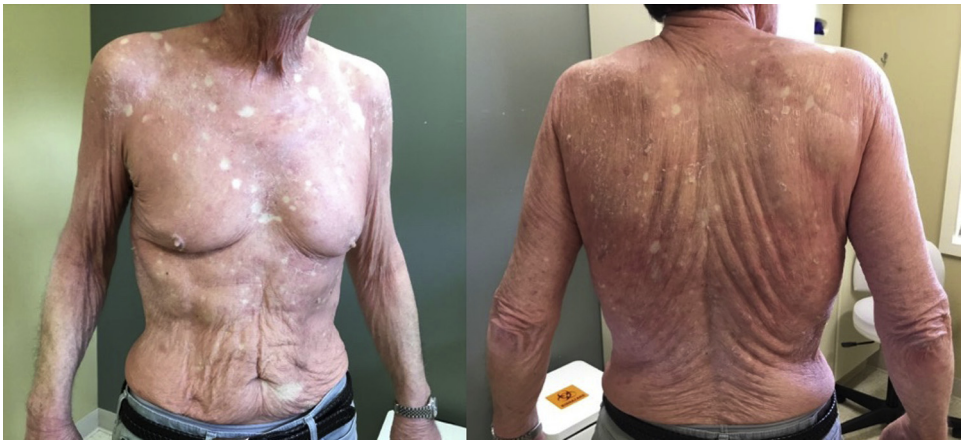

Fig 2. Clinical photos of recurrent PRP showing confluent erythematous scaly plaques and islands of sparing affecting the chest, abdomen, back, and upper extremities.

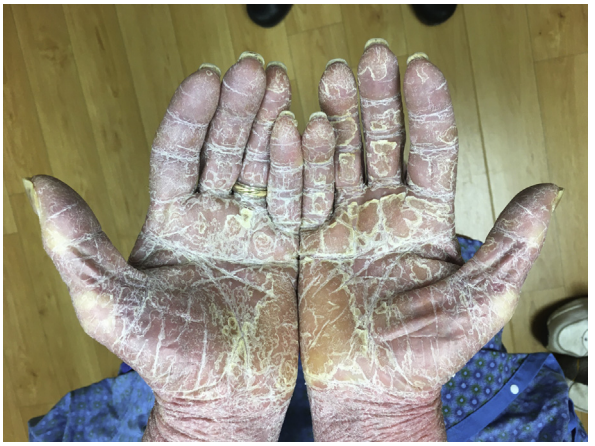

Fig 3. Clinical photo of recurrent PRP showing bilateral palmar hyperkeratosis.

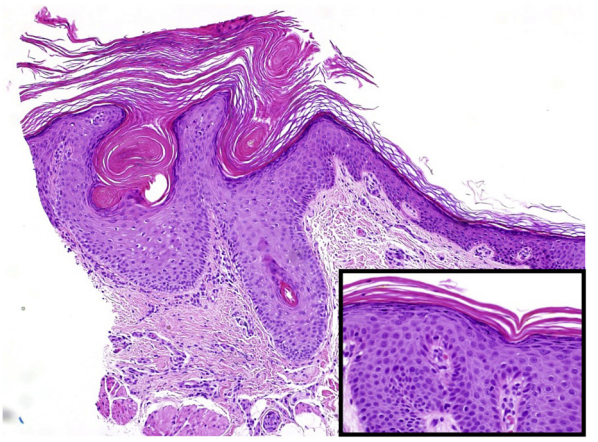

Fig 4. Skin biopsy of recurrent PRP in our patient demonstrating prominent follicular plugging (Hematoxylin and eosin; original magnification: ×100). Inset demonstrates compact hyperkeratosis, alternating parakeratosis, and acanthosis with a scant perivascular lymphocytic infiltrate. (Hematoxylin and eosin; original magnification: ×400).

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