Rapidly Progressive Erythroderma Caused by Pityriasis Rubra Pilars

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Abstract

We present the case of a 50-year-old male who developed rapidly progressive erythroderma as a complication of pityriasis rubra pilars (PRP), requiring hospital admission. The initial eruption developed following a sunburn. Following hospital discharge, the patient has experienced a protracted course of erythroderma, which was treated with cyclosporine and acitretin as well as topical corticosteroids. We briefly review the various classifications of PRP as well as potential treatment options and prognosis.

Introduction

Erythroderma is a generalized redness to the skin with or without scaling. It can be a manifestation of many common primary disorders of the skin including psoriasis, atopic dermatitis, or drug reactions; or less common disorders such as cutaneous lymphoma or pityriasis rubra pilars. Potential complications of erythroderma include peripheral edema, hypothermia, electrolyte imbalance, and high output heart failure. Prompt identification of the underlying disorder and treatment of erythroderma can prevent many complications and potentially be life saving.

Case Presentation

A 50-year-old Caucasian male presented with a three-day history of mildly pruritic erythematous papules and patches progressing from his head to his chest and upper arms after experiencing a sunburn during work. He also complained of redness to his hands and feet. The rash began two months earlier as a single, red, scaly patch on his scalp, which appeared after a mushroom hunting excursion. He had treated the patch with a mid potency topical corticosteroid without resolution. A presumptive diagnosis of psoriasis. The patient had a family history significant for psoriasis, but no other skin disorders. His past medical history was significant for hypercholesterolemia. A review of systems was negative for constitutional symptoms at his initial presentation.

Physical examination revealed a well appearing male with brightly erythematosus, hypertrophic, follicular-based papules and scaly patches coalescing on the scalp, face, chest, and upper extremities (Figure 1). Examination of his hands and feet revealed erythema and hyperkeratosis of the palms and soles (Figure 2).

Clinical differential diagnosis included erythrodermic psoriasis, pityriasis rubra pilars, and drug induced photosensitivity.

Initial laboratory evaluation was within normal limits and included complete blood count with differential, comprehensive metabolic panel, and urinalysis.

Two 4mm punch biopsies were obtained and revealed elongation of rete ridges, hyperkeratosis and confluent parakeratosis. There was a mild superficial perivascular lymphocytic and neutrophilic infiltrate as well as the presence of extravasated red blood cells. PAS stain was negative for fungi and colloidal iron stain was negative for mucinosis. A diagnosis of pityriasis rubra pilars was rendered.

At the initial visit, the patient was started on triamcinolone 0.1% cream and instructed to follow up in two days to review his biopsy results. Initial follow-up revealed progressing erythroderma in a cephalic to caudal direction with islands of spared skin as well as more extensive hyperkeratosis of the palms and soles with fissuring and marked edema. At this time, he was started on oral cyclosporine and acitretin. Despite these medications the erythroderma progressed and he developed 3+ pitting edema of the lower extremities. He was admitted to the hospital for fluid and electrolyte management. During the hospitalization, his laboratory abnormalities included a mild hypoalbuminemia and hyponatremia. Following hospital discharge, the patient’s dose of cyclosporine had been progressively tapered, and the dose of acitretin had been increased. The patient remained erythrodermic, but had experienced much less scaling, and the fissuring to his palms and soles had resolved. He continued to experience moderate pruritis and difficulty in body temperature regulation.

Discussion

Pityriasis rubra pilars (PRP) is an uncommon, chronic skin condition of unknown etiology. It is characterized by hyperkeratotic follicular papules and palmoplantar keratoderma. The coalescence of papules bordered by uninvolved skin creates the appearance of “islands of sparing” between salmon-colored, scaling plaques. Progression to erythroderma is a potential complication.

PRP affects approximately 2.5 per million of the population and does not differ based on race or gender. There is a bimodal distribution for age of onset, including childhood for familial cases and the fifth or sixth decade for acquired cases2-6.

The Griffiths’ classification scheme describes six different types of PRP, differing in clinical presentation, lesion distribution, course, and duration (Table 1). The majority of patients are Type I, “classic adults,” with generalized distribution and a cephalic to caudal progression. In addition to the cosmetic and functional implications of the tight scales of the scalp and face, the waxy, thickened skin of the soles and palms can crack resulting in painful fissures. The onset is acute and 80% resolve within a three-year period.

Table 1

<table>
<thead>
<tr>
<th>Clinical Type</th>
<th>Name</th>
<th>% of PRP Patients</th>
<th>Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Classic adult</td>
<td>55%</td>
<td>Generalized distribution, cephalic to caudal spread, red-orange plaques with “islands of sparing”, Perifollicular keratoderma, waxy palmoplantar keratoderma</td>
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<tr>
<td>II</td>
<td>Atypical adult</td>
<td>5%</td>
<td>Generalized distribution, areas of eczematous dermatitis with ichthyosiform scale on legs, keratoderma with coarse lamellated scale, occasional alopecia</td>
</tr>
<tr>
<td>III</td>
<td>Circumscribed juvenile</td>
<td>25%</td>
<td>Focal distribution, elbows and knees show erythema &amp; follicular papules, prepuberal onset</td>
</tr>
<tr>
<td>IV</td>
<td>Classic juvenile</td>
<td>10%</td>
<td>Generalized distribution with clinical findings similar to Type I, onset in first 2 years of life or in adolescence</td>
</tr>
<tr>
<td>V</td>
<td>Atypical juvenile</td>
<td>5%</td>
<td>Generalized distribution with follicular hyperkeratosis and erythema, scleroderma-like changes of hands and feet, onset in first five years of life</td>
</tr>
<tr>
<td>VI</td>
<td>HIV-associated follicular syndrome</td>
<td></td>
<td>Generalized distribution with findings similar to Type I, can occur in association with acne conglobata and hidradenitis suppurativa in HIV-infected individuals</td>
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Griffith’s classification scheme of pityriasis rubra pilars. Adapted from Bolognia, 3rd Ed. Fig. 9 p 164

While the pathogenesis of PRP remains uncertain, abnormal vitamin A metabolism, specifically a deficiency of retinol binding protein, and human immunodeficiency virus (HIV) have been studied as possible causes. Autoimmune diseases, sunburn, infections, and malignancies are linked as trigger factors; however, most cases occur without an inciting event.

The familial type of PRP, Type V, follows an autosomal dominant mode of inheritance, early age of onset, incomplete penetrance, and variable expression. In a recent study, Fuchs-Teelen et al. showed that mutations in CARD14, which regulates inflammatory processes through nuclear factor kappa B (NF-κB) and is strongly expressed in the skin, cause familial PRP.

Conclusion

Pityriasis rubra pilars is an uncommon chronic skin condition, which can potentially lead to erythroderma. The majority of PRP patients will present as adults with a generalized eruption beginning on the head and neck, which then generalizes in a caudal direction. Unique features include “islands of sparing” and a waxy palmoplantar keratoderma. Although the etiology is unknown most presenting with classic symptoms will experience resolution within three years. There are no universally successful treatments and the patient approach must be individualized.

References

9. Chapter 9, Other Papulosquamous Disorders: p. 157-69