Acad Med J 2025;5(Suppl 1):S152-S158.
UDC: 616.58-003.875-085-053.2
https://www.doi.org/10.53582/AMJ2551152p
Case Report

SPONTANEOUS IMPROVEMENT IN A PATIENT WITH JUVENILE PITYRIASIS RUBRA PILARIS

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Abstract

Pityriasis rubra pilaris (PRP) is a rare and complex dermatosis characterized by erythematous plaques, follicular papules, hyperkeratosis, and scaling. While its precise etiology remains largely unidentified, genetic predisposition, immune-mediated mechanisms, and environmental triggers are implicated in its pathogenesis.

We present a case of a 3-year-old child with a 1-month history of erythematous macules, initially affecting the hands, feet, perioral, and periocular regions. The lesions were initially recognized by a dermatologist and pediatrician as hand, foot, and mouth disease (HFMD). Two weeks later, the patient developed widespread involvement of 80% of the skin surface, including the face. Considering the evolutionary progression of the clinical changes, a diagnosis of pityriasis rubra pilaris (PRP) was established.

After 6 weeks of local treatment with corticosteroids and emollients, significant reduction in body surface area (BSA) was observed. The idea of initiating systemic therapy with acitretin was dismissed, given the favorable course of the disease. After 5 months of clinical observation, the patient achieved complete resolution of the skin changes, highlighting the self-limiting nature of the juvenile form. The patient underwent frequent check-ups for 1 year, and no changes or relapses were observed. This case highlights the value of conservative management and vigilant monitoring in achieving long-term resolution and minimizing the need for systemic interventions.

Keywords: pityriasis rubra pilaris; juvenile form; local treatment; children

Introduction

Pityriasis rubra pilaris (PRP) is a rare, chronic skin disorder characterized by erythematous plaques, follicular papules, and hyperkeratosis, primarily affecting the palms and soles[1]. The exact etiology remains unclear, though genetic predispositions, immune dysregulation, and environmental factors such as infections, trauma, and certain medications are thought to contribute to disease onset and exacerbation [2,5].

Griffiths' classification of PRP includes 5 distinct types: classic adult PRP, atypical adult PRP, classic juvenile PRP, circumscribed juvenile PRP, and atypical juvenile PRP, while an additional sixth type was introduced by Miralles *et al*., specifically related to HIV patients, differing in presentation and being significant for diagnosis and treatment [3,4]. With an estimated incidence of approximately 1 case per 5,000 dermatology patients, this rare dermatological disorder remains uncommon within the field. Both males and females can be affected by PRP, with male predominance in both juvenile and adult forms [5]. The adult form of PRP, the most common type, typically appears between 40-60 years of age, starting with erythematous, scaling plaques on the scalp, trunk, and upper limbs, often progressing to larger,

confluent plaques. Unlike the juvenile form, it usually spares the face and flexural areas, progresses more slowly, and is more likely to become chronic. In contrast, the juvenile form is less common but tends to present more aggressively, often in children or adolescents, typically between the ages of 3-18. Griffiths classifies the juvenile form of PRP into 3 subtypes [3]. The classical juvenile form (type III) typically manifests with a rapid onset of extensive, confluent, scaling, pink-orange plaques, often accompanied by keratoderma of the hands and feet, with about 50% of cases resolving within 6 months. The circumscribed juvenile form (type IV), the most prevalent pediatric subtype, primarily affects the extensor surfaces of the knees and elbows, along with palmoplantar keratoderma, and generally begins between 3 and 9 years of age, showing a similar rate of spontaneous resolution. The atypical juvenile form (type V), which can appear from birth or early childhood, is associated with mutations in the CARD14 gene and often presents with an unusual distribution, commonly involving the face, neck, and upper arms, with less pronounced scaling, making diagnosis more complex [3,5].

The diagnosis of juvenile (PRP) is primarily clinical, and management begins with emollients, topical corticosteroids, and keratolytics [6]. In sensitive areas, calcineurin inhibitors like tacrolimus or pimecrolimus can be used as alternatives to corticosteroids, while calcipotriol, a vitamin D3 analog, is also considered as a therapeutic option. If there is insufficient improvement after a few weeks, systemic therapy with oral acitretin is introduced. If acitretin proves ineffective, second-line treatments such as methotrexate or cyclosporine are considered, and biologic therapies (e.g., TNF-alpha inhibitors, IL-17/IL-23 inhibitors) may be explored for refractory cases [7,8]. JAK inhibitors can also be a therapeutic option for treating pityriasis rubra pilaris (e.g. Tofacitinib, Upadacitinib, Baricitinib). Phototherapy, particularly narrow-band UVB, is also an option for widespread lesions or when systemic treatments are contraindicated [6,8].

We present a case of classical juvenile pityriasis rubra pilaris, with gradual symptom improvement regardless of the initially rapid and aggressive clinical presentation.

Case report

A 3-year-old Caucasian boy was admitted to our department with a one-month history of erythematous macules, initially localized to the palms and later extending to the soles, lower extremities, and periocular and perioral regions. The patient was examined by a dermatologist and a pediatrician, and a diagnosis of hand, foot, and mouth disease was established. The patient was treated with local emollients and antiseptic therapy (potassium permanganate), which was improperly applied in an undiluted form. Two weeks later, the dermatological condition worsened, with the appearance of diffuse erythema and desquamation on the face, erythematous papules and plaques on the torso and extremities, and hyperkeratotic plaques on the palms and soles. Consequently, the patient was referred to the University Clinic for Dermatology for comprehensive evaluation and management

During physical examination, the child was found to be in stable overall condition, exhibiting appropriate growth and development for his age. No familial history of similar cases was documented, and there was no consanguinity among family members. Additionally, there was no history of allergic reactions.

The patient's dermatological condition was characterized by erythematous macules and papules coalescing into larger patches, affecting both the trunk and extremities. Diffuse erythema and pityriasis-like desquamation were observed across the entire body, most prominently in the subaxillary regions, arms, back, sacral area, and thighs. The face exhibited diffuse erythema accompanied by pronounced pityriasis-like desquamation, consistent with the findings on the rest of the body. The skin also showed areas of unaffected skin (*nappes claires*), interspersed with numerous excoriations, creating a distinct contrast (Figure 1). The palms and soles exhibited a yellowish, waxy keratoderma with visible fissures and diffuse edema (Figure

2). Marked edema was also noted on the left knee (Figure 3). Additionally, discrete nail changes were observed, including nail plate thickening and subungual hyperkeratosis. There was no involvement of the mucous membranes, and no lymphadenopathy or organomegaly was present.

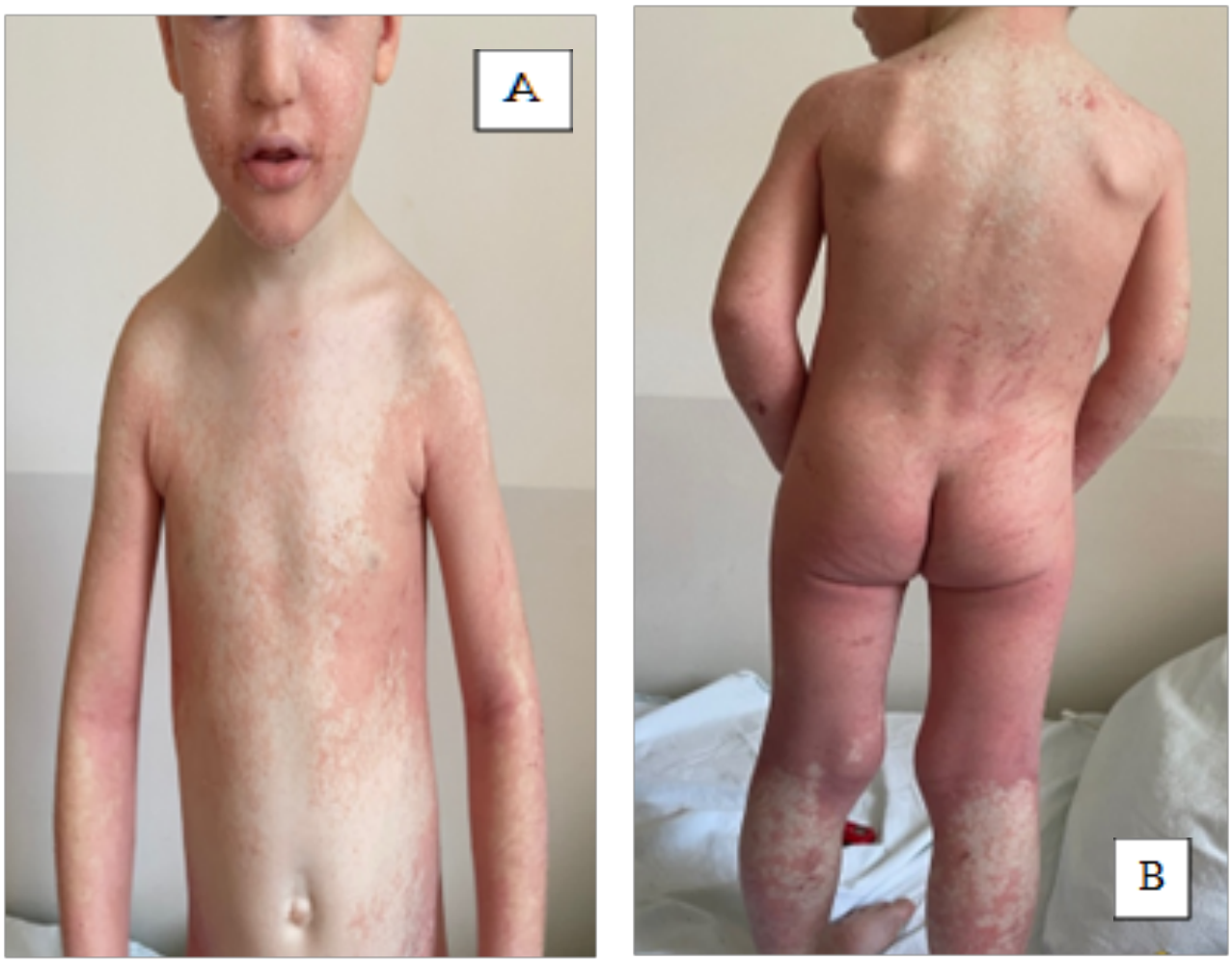

Fig. 1. The face showed diffuse erythema and pityriasis-like desquamation (A). Erythematous macules and papules coalesced into larger patches on the trunk and extremities, most prominently in the subaxillary regions, arms, back, sacral area, and thighs, with diffuse erythema, pityriasis-like desquamation, and areas of unaffected skin (nappes claires). Excoriations were also present (A,B).

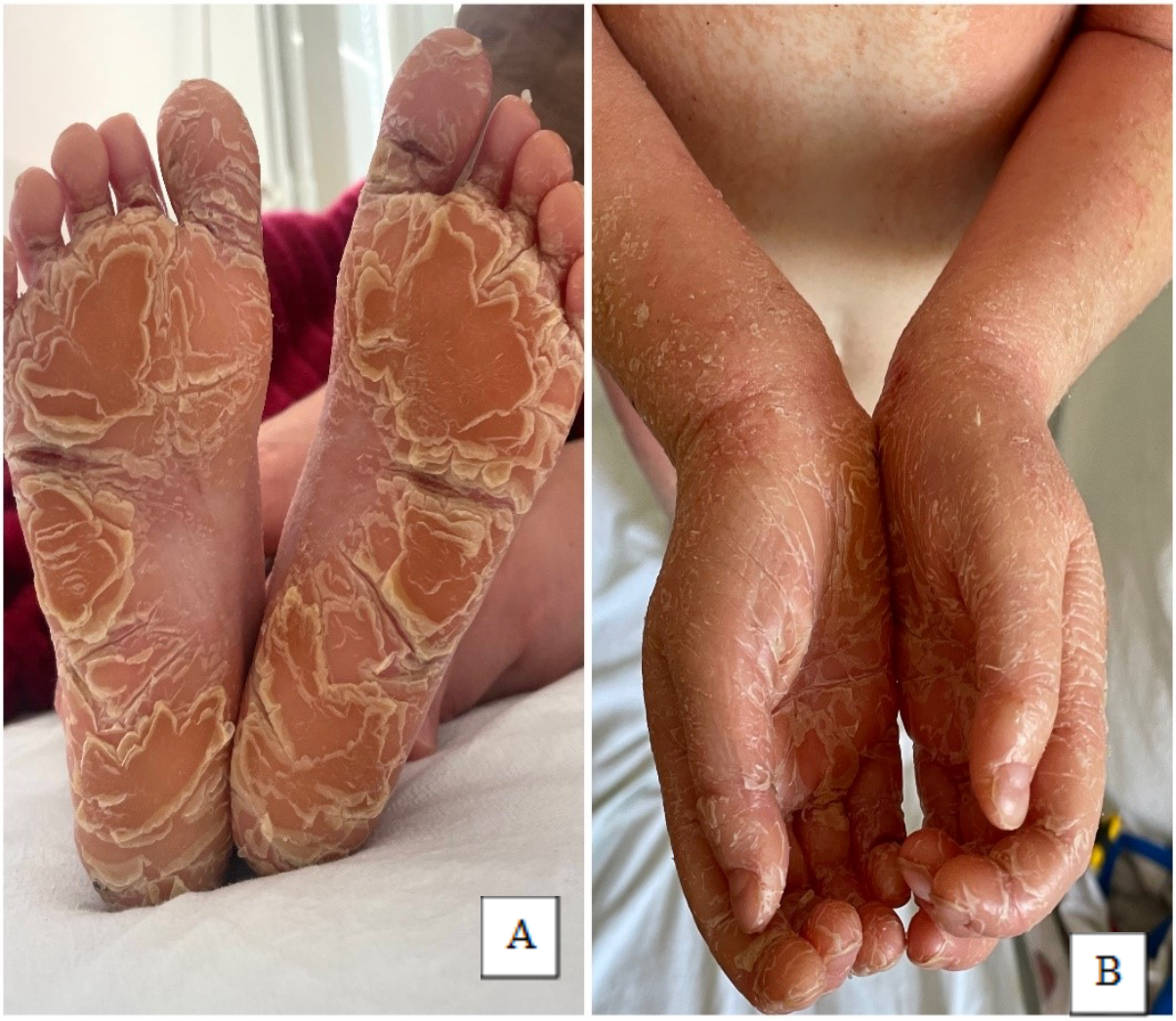

Fig. 2. The soles and palms exhibited yellowish, waxy keratoderma with visible fissures, accompanied by diffuse edema, and thickened, scaly patches (A,B).

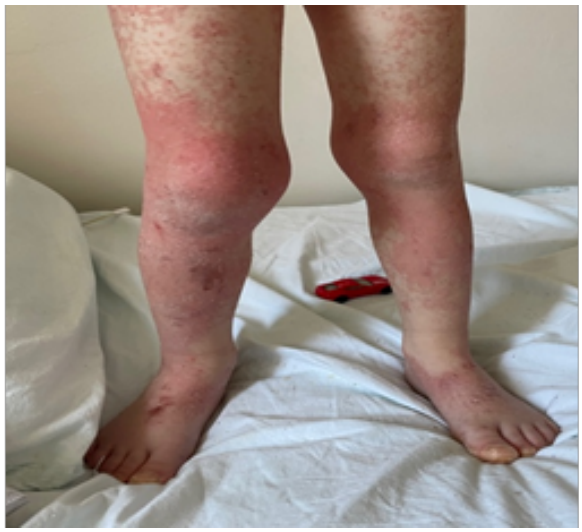

Fig. 3. Marked edema on the left knee

Given the child's age, ethical considerations, lack of cooperation, and the clear clinical presentation of the disease, it was decided to omit histopathological confirmation of the diagnosis.

Laboratory investigations revealed the following abnormalities (Table 1):

Table 1. Abnormal laboratory investigations

Laboratory tests	Results	Reference range
WBC	17.58 x 10^9 cells/L	5.0 – 15.5 x 10^9 cells/L
ASO titer	398IU/mL…562IU/mL	<200 IU/mL

Due to the elevated ASO titer, microbiological investigations were subsequently performed on throat, nasal, and skin swabs, which revealed the following pathological findings (Table 2):

Table 2. Microbiological swabs

Throat swab	*Streptococcus pyogenes, Staphylococcus aureus*
Nasal swab	Negative
Skin swab	*Staphylococcus aureus*

Based on the antibiogram, oral antibiotic therapy was initiated to target the identified infection. Additionally, oral antihistamines were prescribed to alleviate pruritus and reduce discomfort. Local treatment included an emollient, corticosteroid, and antibiotic, which were used to reduce inflammation and treat the infection. After significant improvement in cutaneous symptoms, the patient was discharged following a 13-day hospitalization, with instructions to continue using emollients and corticosteroids to maintain skin condition and control inflammation. Follow-up care was scheduled to monitor the patient's progress, ensure proper healing, and assess the response to ongoing treatment. The patient was also advised to remain vigilant for any recurrence of symptoms, such as increased redness, swelling, or pruritus, and to seek prompt medical attention if any new or worsening symptoms occurred.

Discussion

Pityriasis rubra pilaris (PRP) comprises 6 types [3]. According to Griffiths' criteria, our case corresponds to type III of juvenile PRP, which is typically associated with a more aggressive course and clinical presentation, accounting for approximately 10% of all juvenile PRP cases. The diagnosis of pityriasis rubra pilaris is primarily based on the clinical presentation, as the characteristic features are crucial for distinguishing it from other inflammatory skin conditions [6,8]. Histopathological examination may aid in confirming the diagnosis but is secondary to clinical evaluation [9]. The differential diagnosis includes psoriasis, seborrheic dermatitis, atopic dermatitis, nummular dermatitis, tinea corporis, lichen planus, ichthyosis vulgaris, pityriasis rosea, drug eruptions, systemic lupus erythematosus, etc[7].

The prognosis for the classic juvenile form of pityriasis rubra pilaris (PRP) is generally favorable, with most cases improving within the first year and typically resolving within 6 months to 2 years [10]. In some cases, however, the condition may persist for up to 2 years before fully resolving. It is believed to be triggered by viral infections such as rhinoviruses, enteroviruses, influenza virus, coxsackievirus, cytomegalovirus (CMV), Epstein-Barr virus (EBV), and human herpesvirus 6 (HHV-6), as well as bacterial infections like *Staphylococcus aureus* and *Streptococcus* species [1,3]. These infections suggest an immune-mediated response

that typically resolves over time. While the overall prognosis is generally positive, the disease course can vary, and the response to treatment may differ depending on factors such as age, the extent of involvement, and the presence of systemic symptoms. The temporal association between a preceding coxsackievirus infection and the onset of cutaneous manifestations suggests that the viral infection, as observed in our clinical case, may have acted as a potential trigger for the development of PRP.

In our case, significant improvement in the cutaneous lesions and a noticeable reduction in BSA involvement were observed after just 6 weeks of topical treatment. Based on this positive response, the option of initiating systemic therapy with acitretin was reconsidered and ultimately excluded, despite its initial consideration as a potential treatment approach. Three months later, the skin was nearly entirely normalized, with only a few minor scales and residual erythema on the torso and knees. After 5 months, complete resolution of the skin lesions was achieved, confirming the efficacy of the initial treatment and the self-limiting nature of the disease. This was further supported by the absence of cutaneous changes after one year, consistent with the typical course of juvenile pityriasis rubra pilaris. The method of regular follow-up and a 'wait and see approach proved to be crucial in monitoring the patient's progress and ensuring the resolution of the condition. This case highlights the importance of close monitoring and individualized management in achieving successful outcomes in classic juvenile PRP. It also emphasizes that, in many cases, conservative management with appropriate topical treatments can lead to substantial improvement, thereby potentially avoiding the need for systemic therapy, especially when the disease follows a typical self-limiting course.

Conclusion

Classic juvenile pityriasis rubra pilaris is a rare condition with a good prognosis, typically following a self-limiting course. Early recognition is key for accurate diagnosis. Given its self-limiting nature, a 'wait and see' approach is often appropriate, with vigilant monitoring to detect potential complications or relapses.

Conflict of interest statement. None declared.

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