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CASE REPORT

Circumscribed Juvenile Pityriasis Rubra Pilaris in a 4-Year-Old Filipino Female Treated with Acitretin: A Case Report

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ABSTRACT: Pityriasis rubra pilaris (PRP) Type IV, the circumscribed juvenile form, is a rare, chronic papulosquamous disorder in children. It can be misdiagnosed as paediatric psoriasis because both share erythematous plaques and hyperkeratotic lesions. PRP Type IV typically presents with well-demarcated, scaly, reddish-orange plaques on the elbows, knees, hands, and feet. Orange-hued palmoplantar keratoderma may also accompany the condition. Diagnosis relies on clinicopathological correlation, with biopsy typically revealing hyperkeratosis and a checkerboard pattern of alternating parakeratosis and orthokeratosis. Management is challenging because no standardised treatment guidelines exist, although combined topical and systemic therapy may be effective. In this report, a 4-year-old girl presented with a 2-year history of persistent pruritic plaques. She was treated with low-dose oral acitretin, topical emollients, corticosteroids, and vitamin D analogues, resulting in marked clinical improvement and good treatment tolerance. This case highlights the importance of accurate diagnosis and demonstrates that early, appropriately tailored therapy can achieve favourable outcomes and improve quality of life in paediatric patients with PRP Type IV.

Keywords: Acitretin, Pityriasis rubra pilaris, PRP, Treatment, Type IV

INTRODUCTION

Pityriasis rubra pilaris (PRP) is a rare, chronic papulosquamous disease of keratinisation. Its aetiology remains unknown [1]. The incidence is estimated at 1 in 5000, with an equal male-to-female ratio [2]. PRP is classified into six types, each characterised by distinct clinical features, age of onset, and disease progression [1]. PRP Type IV, the circumscribed juvenile form, usually occurs in prepubertal children. Its primary characteristics include hyperkeratotic perifollicular papules that develop into sharply defined erythematous plaques, usually on the elbows, knees, and the dorsal aspects of the hands and feet. Orange-hued palmoplantar keratoderma often accompanies the condition [1]. Because this dermatosis is rare, with limited evidence from clinical studies and a lack of clinical practice guidelines, treatment remains challenging. This report describes the clinical, histopathological, and therapeutic features of PRP Type IV in a 4-year-old girl, as well as the clinical response to a combined topical and systemic treatment approach.

CASE PRESENTATION

We report the case of a 4-year-old Filipino girl with a 2-year history of persistent, very itchy, scaly, reddish-orange-colored plaques on the palms, soles, and both knees. The patient consulted a private physician with unrecalled diagnosis and was only given moisturising lotion, with no other treatment. Review of systems was unremarkable. No similar cases were reported in the family. The parents are non-consanguineous. Dermatological examination revealed multiple, well-defined, irregularly shaped, erythematous, scaly plaques and patches on the hands, feet, and knees, some with excoriations (**Figure 1**). Hair and nail examinations were normal. Systemic examination was unremarkable, and laboratory examinations including complete blood count, liver enzymes, serum chemistries, and chest radiograph were all within normal limits.

A 4-mm skin punch biopsy was taken from the erythematous plaque on the right sole. Histopathological examination with hematoxylin and eosin (H&E) staining

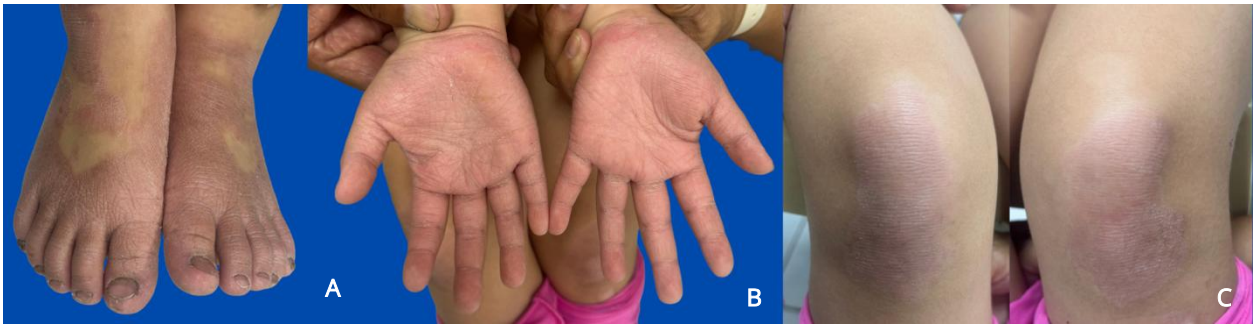

Figure 1. Erythematous, scaly plaques with follicular accentuation on the feet **(A),** hands **(B),** and knees **(C)** and diffuse symmetrical orange, waxy palmoplantar keratoderma with fissures.

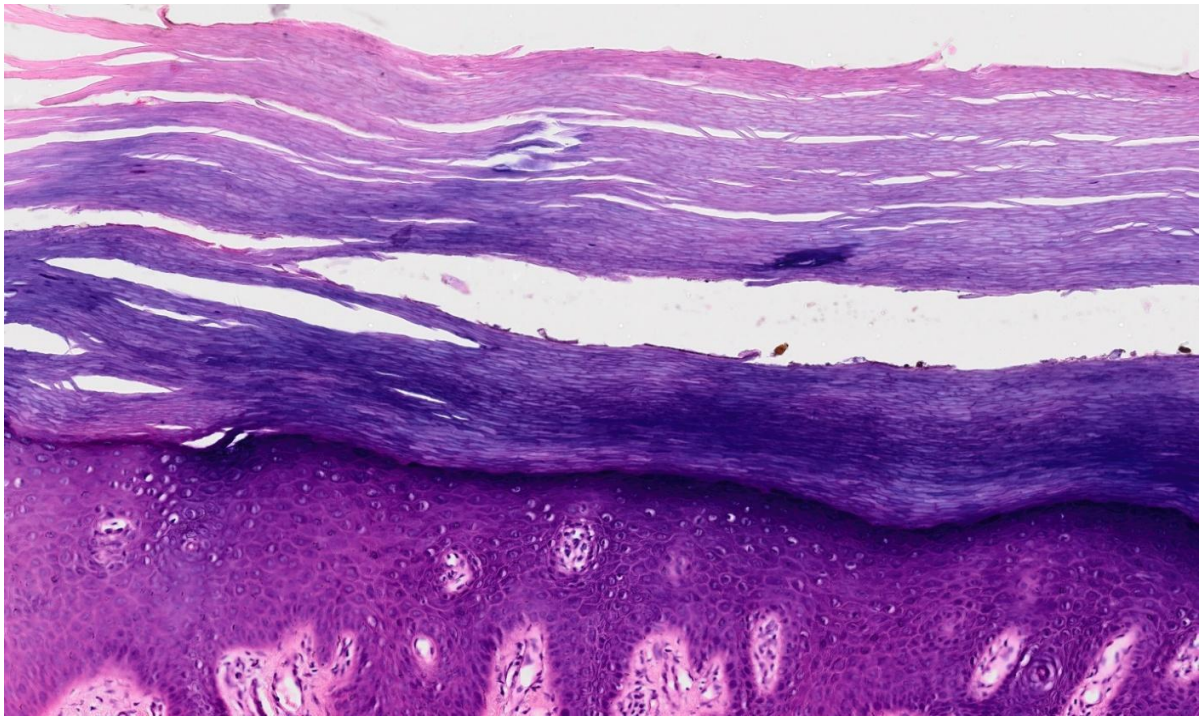

Figure 2. H&E staining of a skin punch biopsy specimen obtained from the right sole shows prominent acanthosis of the epidermis, hyperkeratosis, and alternating vertical and horizontal parakeratosis of the stratum corneum in a checkerboard pattern.

revealed hyperkeratosis and alternating vertical and horizontal parakeratosis in a checkerboard pattern in the stratum corneum. There was prominent acanthosis of the epidermis. The dermis showed a mild, superficial perivascular lymphocytic infiltrate (**Figure 2**). Based on the aforementioned clinicopathological findings, the diagnosis of PRP (Type IV) was established. Written informed consent for publication was obtained from the patient's legal guardian.

MANAGEMENT AND OUTCOME

The patient was started on acitretin 10 mg once daily (0.43 mg/kg/day), together with 30% urea lotion and clobetasol propionate 0.05% ointment applied twice daily for 2 weeks. After 2 weeks, clobetasol propionate was changed to calcipotriol, while the other medications were continued. The initial treatment plan was to administer acitretin for 3–6 months, depending on the clinical response. After 12 weeks of once-daily acitretin at 10 mg, complete resolution of the skin lesions on the bilateral knees, hands, and feet was observed (**Figure 3**). The acitretin dose was subsequently reduced to 10 mg every other day. However, the patient took the medication for only 2 weeks before being lost to follow-up. After discontinuing treatment for 2 months, the patient returned with recurrence of skin-coloured to yellowish, waxy plaques on the palms and soles.

Following the relapse, acitretin was reintroduced at 10 mg once daily, together with topical clobetasol propionate ointment. The lesions again responded favourably following treatment reintroduction. Appropriate laboratory monitoring was performed throughout acitretin therapy, and no treatment-related adverse effects were observed. The total duration of treatment was 16 weeks, comprising 12 weeks of once-daily acitretin, 2 weeks of alternate-day dosing and 2 weeks of once-daily treatment following reintroduction. Treatment was interrupted by a period of approximately 2 months, during which the patient was lost to follow-up. The patient attended six follow-up visits in total.

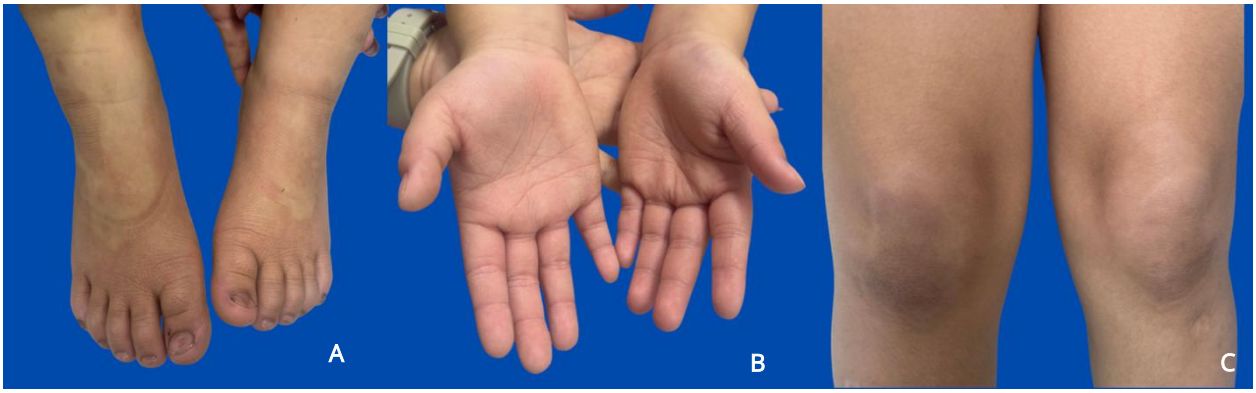

Figure 3. After 12 weeks of acitretin treatment showing complete flattening of the lesions on the feet **(A),** palms **(B),** and knees **(C).**

DISCUSSION

PRP is a rare inflammatory papulosquamous dermatosis. Griffiths originally classified it into five types. Later, a sixth category (HIV-associated) was added [1]. The exact aetiology and pathogenesis remain unclear. Several factors may be associated, such as heredity, endocrine dysfunction, vitamin A deficiency, autoimmune diseases, infections, trauma, and vaccination [3]. Most PRP types have generalised skin lesions, except Type IV, where the lesions are typically localised to the elbows, knees, palms, and soles [4]. This is the most common PRP type in children, comprising about 25% of paediatric cases. It is characterised by well-demarcated, hyperkeratotic erythematous plaques on the elbows and knees, resembling localised psoriasis. Palmoplantar keratoderma is characteristic of Type IV PRP but may also be absent [1].

PRP can be differentiated from psoriasis by the absence of characteristic silvery scales and the relative lack of scalp and nail involvement, which are common features of psoriasis. Dermoscopy further aids in distinguishing the two conditions. PRP typically demonstrates focal yellow-orange structureless areas with fine whitish scales, whereas psoriasis characteristically shows regularly distributed red dots and diffuse white scales on a pink to light red background. In the case presented, the presence of palmoplantar keratoderma along with erythematous plaques on the body initially led us to consider a diagnosis of paediatric psoriasis. A biopsy showing alternating parakeratosis and orthokeratosis in a checkerboard pattern supported the diagnosis of PRP. Other histopathologic features include hyperkeratosis and focal acantholytic dyskeratosis [1]. This underscores the diagnostic challenge of PRP, which can mimic other benign inflammatory dermatoses.

Due to the heterogeneity of the disease, there is no single standardised treatment approach, and various therapies may need to be considered to achieve adequate disease control. Currently, paediatric patients with PRP may be managed with topical treatment alone, particularly in less severe disease, while oral retinoids are widely accepted as the first-line option for systemic therapy [5]. Topical treatment includes medium- to high-potency corticosteroids, keratolytics, emollients, and vitamin D analogues (e.g., calcipotriol) [1,2]. Combining topical and systemic therapies can be effective in managing hyperkeratosis and palmoplantar keratoderma [2].

In our patient, a combination of systemic acitretin and topical therapies was selected to target both the underlying disorder of keratinisation and the localised hyperkeratotic inflammation. Marked clinical improvement was observed during treatment. A relatively low dose was used to achieve disease control while limiting potential adverse effects associated with long-term retinoid therapy in children. Topical clobetasol propionate was initially prescribed to rapidly suppress inflammation and improve symptomatic hyperkeratotic plaques. Because prolonged use of superpotent topical corticosteroids is associated with cutaneous adverse effects, clobetasol was limited to 2 weeks and subsequently replaced with calcipotriol for maintenance therapy. Emollients, mild soap, and 30% urea lotion were included to restore the skin barrier, reduce hyperkeratosis, and improve scale removal.

Clinical improvement in PRP is generally reported within 3–6 months of oral retinoid therapy [6]. A 2024 scoping review reported that, among 32 patients treated with acitretin, 59.4% achieved a significant response, 25.0% achieved a partial response, and 3.1% showed no response. These findings support the potential role of oral retinoids in the management of PRP [7]. The recommended oral acitretin dosage is 0.5 to 0.75 mg/kg/day [2]. In our patient, a relatively low dose of acitretin was used, which was associated with complete clinical remission when combined with adjunctive topical therapy, without treatment-related adverse effects. High doses of retinoids are often poorly tolerated because of frequent mucocutaneous adverse effects. The most common side effects of retinoids include skin dryness and chapped lips. Elevated lipid levels, elevated liver enzymes, and visual or skeletal changes can also occur. In children, retinoids may cause premature epiphyseal closure, especially in prepubertal patients receiving high doses [2]. These safety considerations may be particularly relevant when treating paediatric patients with PRP, and lower doses may potentially improve treatment tolerability and adherence.

A notable aspect of this case is the documented disease recurrence following treatment interruption due

to financial constraints rather than documented therapeutic failure. The recurrence of lesions approximately 1 week after treatment discontinuation, followed by a favourable response after reintroduction of acitretin at an increased dosing frequency alongside topical therapy, highlights the potential importance of treatment adherence and continuity in maintaining disease control. This case adds to the clinical experience with acitretin in PRP and illustrates the possibility of disease recurrence following treatment discontinuation.

Unlike previously reported cases of refractory or extensive PRP requiring escalation to immunosuppressive therapies or biologic agents, our patient achieved complete clinical remission with low-dose acitretin combined with topical therapies. Nevertheless, the long-term prognosis remains uncertain given the variable natural history of juvenile type IV PRP. Although some cases resolve within 1–3 years of disease onset, others achieve complete remission by late adolescence [8].

CONCLUSION

PRP can significantly affect patients' quality of life, particularly in its chronic and treatment-resistant forms. This case highlights the clinical response to low-dose acitretin combined with topical therapy in achieving complete clinical remission of circumscribed juvenile PRP (Type IV) without treatment-related adverse effects. The recurrence following treatment interruption highlights the importance of treatment adherence and continuity in maintaining disease control. In the absence of standardised treatment guidelines, this case adds to the limited clinical evidence regarding the use of low-dose acitretin in paediatric Type IV PRP.

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None.

CONFLICT OF INTEREST

The authors declare no conflicts of interest.

REFERENCES

1. Kang S, Amagai M, Bruckner AL, Enk AH, Paller AS, Watsky KL, et al. Fitzpatrick's Dermatology in General Medicine. 9th ed. New York: McGraw-Hill Education; 2019.
2. Moretta G, De Luca EV, Di Stefani A. Management of refractory pityriasis rubra pilaris: challenges and solutions. Clin Cosmet Investig Dermatol. 2017;10:451-7. DOI: 10.2147/CCID.S124351.
3. Zhang X, Wei K, Song H, Chen X, Yang J, Zhao J, et al. Efficacy and safety of small molecule drugs in the treatment of pityriasis rubra pilaris—a systematic review. Front Med (Lausanne). 2025;12:1544197. DOI: 10.3389/fmed.2025.1544197.
4. Albrakati BA, Alshareef IA, Alhawsawi WK, Al Hawsawi KA. Atypical juvenile pityriasis rubra pilaris: a case report of early onset with late diagnosis. Cureus. 2022;14(10):e30234. DOI: 10.7759/cureus.30234.
5. Guenther J, Novack D, Kamath S, Worswick S. Treatment options for juvenile pityriasis rubra pilaris. Paediatr Drugs. 2023;25(2):151-64. DOI: 10.1007/s40272-022-00549-4.
6. Menta N, Vidal SI, Stines A, Friedman A. Precision, research, progress: updates in the management of pityriasis rubra pilaris. J Drugs Dermatol. 2025;24(8):833-4. DOI: 10.36849/JDD.10825.
7. Zhou T, Al Muqrin A, Abu-Hilal M. Updates on pityriasis rubra pilaris: a scoping review. J Cutan Med Surg. 2024;28(2):158-66. DOI: 10.1177/12034754231223159.
8. Sehgal VN, Srivastava G. (Juvenile) Pityriasis rubra pilaris. Int J Dermatol. 2006;45(4):438-46. DOI: 10.1111/j.1365-4632.2006.02666.x.