MedMax Publishers

ISSN: 2768-5004

Global Journal of Clinical Case Reports Case Report

# Pityriasis Rubra in a Pediatric Patient: About A Case

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**Received**: 23 September 2021; **Published**: 05 October 2021

## Abstract

Pityriasis rubra is a rare erythematous squamous dermatosis of unknown etiology produced by an alteration in the keratinization of the epidermis. It presents a bimodal distribution with a higher incidence in the first and sixth decades of life and a heterogeneous clinical presentation classified into 6 subtypes according to Griffiths, depending on the prognosis. His main findings are follicular hyperkeratotic papules, palmoplantar keratoderma, and red-orange erythematous plaques that can progress to erythroderma, with islands of healthy skin. The histology is not specific but supports the diagnosis. The importance of identifying which subtype our patient presents is due to the fact that there are multiple therapeutic options depending on the extent and severity of the condition.

**Keywords:** Pityriasis rubra, Pediatric pityriasis Rubra

## Introduction

Pityriasis rubra (pr), also known as lichen ruber acuminatus or Devergie's disease, is a rare inflammatory papulosquamous disease with clinically heterogeneous manifestations. It was described for the first time by Claudius Tarral in 1828; Almost thirty years later, in 1856, Alphonse Devergie called it "pityriasis pilar" and, in 1889, Besnier named it pityriasis rubra pilar after having described several cases. The clinical characteristics that appear in prp are highly variable, as well as the prognosis in each patient [1].

Based on the age of onset of the disease and the clinical characteristics, Griffiths classified it into five types (Figure 1): i classic adult, ii atypical adult, iii classic juvenile, iv circumscribed juvenile, and v atypical juvenile [2- 6]. Recently, type vi was added, which is associated with human immunodeficiency virus (hiv) infection. Clinicopathologic findings are the gold standard for diagnosing the disease [7,8].

**Figure 1:** Classification of pityriasis rubra proposed Griffiths

| Type | Age | Clinical Description | Treatment |
|---|---|---|---|
| I | Adult onset | Large zones of follicular hyperkeratosis with an erythematous halo. Islands of unaffected skin covering the erythematous sheets. | Retinoids, isotretinoin, etretinate, acitretin |
| II | Adult onset | Atypical, long duration, increased scaling. Sparseness of scalp hair. Increased palmoplantar hyperkeratosis. | Treated on case by case basis |
| III | 1 - 2 years | Resembles Type I PRP. 90% self-limited in 3 years. | Keratolytics, topical steroids, emollients |
| IV | Pre-pubertal children | Sharply demarcated areas of follicular hyperkeratosis and erythema on knees and elbows. | Systemic retinoic acids, topical steroids |
| V | First years of life | Chronic. Follicular hyperkeratosis. Erythema not prominent. Most cases of familial PRP. | Retinoids, methotrexate, cyclosporins, azathioprine, gamma-interferon |

Although the etiopathogenesis of RP has not yet been elucidated, 1 a variant of RP type III has been described, called acute post-infectious RP, which would have the particularity of being triggered by a previous infectious process. It was described for the first time in 1983, when 3 cases of type III triggered RP were reported after an infectious fever [9].

RP, in its acute post-infectious variant, is morphologically indistinguishable from RP type III, characterized by not having a family history; onset during childhood (after the first year of life); presence of a previous infectious episode; beginning with a scarlatiniform erythema, followed by the appearance of follicular papules (image 2); have a good prognosis, self-limited and without recurrences [10].

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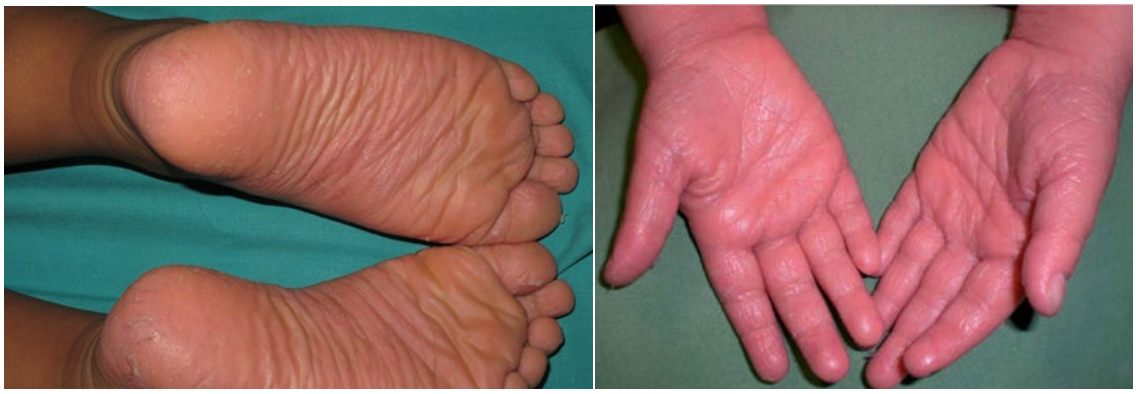

**Image 1:** Characteristic lesions of pityriasis Rubra in pediatric population

## Presentation of the clinical case

20-month-old female patient in good general condition, who attends the consultation in the company of her mother, due to a clinical picture of 72 hours of evolution, characterized by erythema in the hands and feet, accompanied by itching, without other symptoms. Self-medicated with loratadine without showing improvement.

On physical examination, patient with vital signs in normal parameters, hemodynamically stable, without alterations in the nervous system, isochoric pupils normo reactive to light, anicteric stairs, eyes with the presence of erythema, tear ducts with evident abnormal redness, moist oral mucosa without lesions, bilateral otoscope without alterations, mobile neck, without masses or palpable lymphadenopathy, symmetric expandable thorax, without pulling its sacks, on pulmonary auscultation, universal vesicular murmur, well-ventilated lungs, without added sounds, rhythmic heart sounds, well timbred, or murmurs, or aggregates. Abdomen without scars, water noises present, soft, not painful on superficial or deep palpation, without signs of peritoneal irritation, without masses or palpable megaly, unexplored external genitalia, symmetrical extremities, not tender on palpation, hydrated anicteric skin, without edema, with erythematous squamous lesions on the palms of both hands, soles of the feet and on the knees of both extremities (image 2). Conscious, alert patient, without apparent sensory or motor deficit, without neurological focus, Glasgow 15/15., with initial diagnostic impression of idiopathic thrombocytopenic purpura to be ruled out, hypersensitivity reaction and dermatitis. Hemogram and peripheral blood smear are requested to determine the status of the red and white lines.

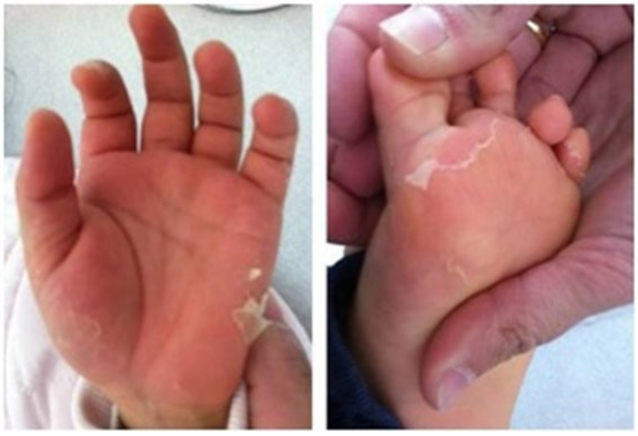

**Image 2:** Erythema on the patient's hands and feet

Hemogram with a report of lymphocytocis and the rest of the formed elements with percentages of normal ranges. With pityriasis rubra pilaris, treatment was started with emolliency, prednisone 0.5 mg / kg / day, and a biopsy was taken for hematoxylin / eosin staining, which reported acanthosis with a network of wide ridges and thick suprapapillary plaques. Hyperortokeratosis with foci of parakeratosis. Dilated follicular ostium with keratin plug and peripollicular parakeratosis. Mild perivascular lymphocytic inflammatory infiltrate is observed in the dermis. This histopathological image is compatible with the diagnosis of RP. At the 10-day follow-up, the patient presented clear improvement with clearing of the erythema and improvement of the erythematous squamous plaques, adding a brown ichthyosiform-looking scale and plantar keratoderma on the front of the legs. Due to good evolution, a descending dose of corticosteroids and tazarotene 0.1% 2 v / day in plates is indicated. Patient shows a good response to established treatment.

## Discussion

Due to the infrequency of the disease, especially in children, large-scale studies of juvenile RP are scarce, with disparate conclusions about its manifestations, classification, and treatment. This is why in the literature there are multiple publications of cases with atypical clinical presentation that fail to be pigeonholed in the Griffith classification. Acute juvenile presentations have been described, with scarlatiniform rash followed by PRP type III lesions, with early resolution. In addition to mixed presentations with type III and IV clinical manifestations [11]. Our patient presents an atypical type V PRP, characterized by palmoplantar keratoderma, associating an ichthyosiform scale on the anterior aspect of the legs. However, it presents with erythematous squamous plaques on the elbows and knees, not considered in this RP subtype [12]. The diagnosis is clinical supported by histology, which shows acanthotic epidermis with alternation of orthokeratosis and parakeratosis in horizontal and vertical direction associated with focal or confluent hypergranulosis. There is dilation of hair follicles with horn plugs. Occasionally there is the sign of "shoulder parakeratosis" given by parakeratosis on both sides of the follicle covered with keratin. Perivascular and peripollicular inflammatory infiltrate of lymphocytes and macrophages is often observed [13]. These findings were found in our patient. There are no standardized treatments in pediatric RP [14]. The association of topical and systemic treatments is frequently used. Topical treatments include emolliency, keratolytics, corticosteroids, calcipotriene, and retinoids [15]. Tazarotene is considered safe and well tolerated in patients with localized disease [16].

Tazarotene 0.1% 2 times a day was satisfactory in our patient as maintenance. The systemic treatment used in PRP includes corticosteroids, retinoids, vitamin A, methotrexate, cyclosporine, and biologics [17]. The use of prednisone in cases of erythroderma has been described [18].

The aforementioned supports the rapid resolution of the extensive erythema in our patient. In the literature there are case reports with disparate results with the use of vitamin A in both children and adults [19,20]. Systemic retinoids are considered the most effective treatments. Among these, the most widely used are isotretinoin and etretinate, which are synthetic derivatives of vitamin A that modulate the growth and differentiation of epithelial tissues. So far, isotretinoin has been approved by the FDA for use in children 12 years of age and older for the treatment of acne. Acitretin and etretinate have not yet been approved in children. The benefit of acitretin in the treatment of PRP is controversial, with different results in published cases [21,22].

Tretinoin is a panagonist of retinoid receptors, with an immunomodulatory and anti-inflammatory effect; and it could be considered as a safe treatment in recalcitrant juvenile localized RP [23]. When retinoids are not effective, methotrexate should be considered as a second-line treatment [24]. Phototherapy has proven to be an effective option. There are case reports of successful treatment with PUVA, RePUVA, UVB-BE, including the combination with acitretin (ReUVB) [25]. Biologics can be used as an option in recalcitrant RP in adults, with no evidence in children to date [26].

## Conclusion

In conclusion, RP is a rare pathology in children in which clinical forms III and IV are described. Due to its torpid evolution, it presents a therapeutic challenge. There are no serological markers for prp, however, laboratory tests can be obtained to evaluate possible coexisting conditions, such as neoplasms or hypothyroidism. It is also helpful in establishing baseline parameters for monitoring these laboratory tests during treatment. These studies should include a differential white blood cell count, a complete metabolic profile, liver tests, and a lipid profile if retinoids such as isotretinoin are used.

In addition, due to the association of type VI with hiv infection, it is advisable to perform a rapid test for the detection of the virus. The diagnosis of Pr is made clinically and confirmed with histopathology, which reveals irregular hyperkeratosis with alternation of orthokeratosis and parakeratosis in vertical and horizontal directions, forming the typical checkerboard pattern other frequent findings are hypergranulosis, follicular tamponade, broad ridges, narrow dermal papillae, mild superficial perivascular lymphocytic infiltrate and vasodilation.

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**Citation:** Estefanía Cordero Lugo, Alba Lucía Vellojín Olivares, Angie Katerine Rodríguez Paredes, Olga Patricia Hoyos Hernández, Ricardo Antonio Rendón Muñoz, et al.. PPityriasis Rubra in a Pediatric Patient: About A Case. G J Clin Case Rep 2021, 2: 1-4.