

CASE REPORTS

Acute Postinfectious *Pityriasis Rubra Pilaris*: A Superantigen-mediated Dermatitis

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Abstract. Acute postinfectious pityriasis rubra pilaris (PRP) is a variant of juvenile PRP (Griffiths type III) characterized by no family history, an acute course associated with a prior fever, and good prognosis. Clinical features may resemble other superantigen-mediated diseases, such as scarlatiniform rash or staphylococcal scalded skin syndrome, but its histology and treatment are different. We present 4 cases of acute postinfectious PRP that illustrate the clinical features of this uncommon disease and we review possible underlying pathogenic mechanisms.

Palabras clave: *pityriasis rubra pilaris*, erythroderma, infection, childhood, juvenile, superantigens.

PITIRIASIS RUBRA PILARIS AGUDA POSTINFECCIOSA: UNA DERMATOSIS MEDIADA POR SUPERANTÍGENOS

Resumen. La pitiriasis rubra pilaris (PRP) aguda postinfecciosa es una variante de la forma juvenil de PRP (tipo III de Griffiths) caracterizada por la ausencia de antecedentes familiares, curso agudo relacionado con un episodio febril previo y buen pronóstico. Clínicamente puede simular otras enfermedades mediadas por superantígenos, como los exantemas escarlatiniformes o el síndrome de la escaldadura estafilocócica; sin embargo, su histología y tratamiento son distintos. Presentamos 4 casos de PRP aguda postinfecciosa que ilustran las características clínicas de este proceso infrecuente y revisamos los posibles mecanismos fisiopatogénicos subyacentes.

Palabras clave: *pitiriasis rubra pilaris*, eritrodermia, infección, infantil, juvenil, superantígenos.

Introduction

Pityriasis rubra pilaris (PRP) is a chronic skin disease rarely found in children that is characterized by the appearance of follicular hyperkeratotic papules, erythematous plaques, and palmar-plantar keratoderma.^{1,2} The condition involves keratinization caused by accelerated epidermopoiesis, in which underlying defects in vitamin A metabolism, hypogammaglobulinemia, or isolated immunoglobulin (Ig) A deficiency are believed to be present.³ In many cases, patients have a history of infection, fever, sun exposure, or traumatic injury. In 1980 Griffiths⁴ proposed a classification of 5 groups. However, some cases do not fit any of these groups. In 1983 Larrègue et al⁵

proposed a new variant as a subtype of form III of this classification: acute or postinfectious juvenile PRP, a condition rarely reported in the literature.

Case Descriptions

We present 4 patients who attended the pediatric dermatology outpatient clinic for diagnosis and treatment of a generalized erythematous-desquamative rash that appeared 5 to 10 days after a fever. The most relevant clinical information is summarized in Table 1.

Patient 1

An 18-month-old girl came to the emergency room for a generalized rash with onset 3 days earlier. She was initially diagnosed with staphylococcal scalded skin syndrome (SSSS) and was admitted and treated with oral cloxacillin 250 mg/6 hours and neomycin ointment in the natural

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Table. Most Relevant Clinical Data of the 4 Patients Diagnosed with Acute Postinfectious Pityriasis Rubra Pilaris

	Patient 1	Patient 2	Patient 3	Patient 4
Age, y	1.5	1.5	3	2.5
Presenting complaint	Generalized rash	Erythroderma, fever, cold symptoms, diarrhea	Erythematous-desquamative rash, lack of response to oral corticosteroids or antihistamines	Generalized rash
Disease history	Recurrent bronchitis and otitis, seborrheic dermatitis	None of interest	Erythema multiforme	None of interest
History of infection before cutaneous symptoms	Yes (fever)	Yes, simultaneous onset	Yes (upper respiratory infections)	Yes (fever), ibuprofen
Physical examination	Good general health, erythema, very fine flaking, erythematous plaques. Palmar-plantar erythema and flaking in finger pads, vulvar, perianal, and intergluteal areas	Erythematous-desquamative plaques. Circumoral and periocular fissures, conjunctival purulent secretion, swollen lips, and thrush	Desquamative erythroderma, follicular papules, islands of healthy skin, orange-colored palmar-plantar keratoderma	Desquamative plaques, islands of healthy skin. Furfuraceous scaling on the scalp
Additional examinations	Laboratory workup: leukocytes 17 000 L/mm ³ (52% N, 40% L), all other counts normal. Blood, skin, and mucosa cultures negative. Lymphocytic population study normal. Parvovirus B19 serology negative. Skin biopsy: suggestive of PRP	Laboratory workup: leukocytes 14 900 L/mm ³ and AST 66 IU/mm ³ . Cultures: <i>S aureus</i> (exfoliative toxin B) in conjunctival exudate and enterovirus in stool culture	Skin biopsy: suggestive of PRP	No
Initial diagnosis	SSSS in acute postinfectious PRP	SSSS	Acute postinfectious PRP	Acute postinfectious PRP
Initial therapy	Cloxacillin po 250 mg/6 h, neomycin ointment	Cloxacillin iv, antipyretics, fusidic acid ointment	Topical emollients and salicylic acid	Hydroxyzine, methylprednisolone aceponate emulsion, oatmeal baths, coal tar shampoo
Outcome	See text (clinical cases)	See text (clinical cases)	Cure (3 weeks), residual hypochromic macules. No new flare-ups (1 year of follow-up)	Cure (5 weeks) residual hypochromic macules. No new flare-ups (1 year of follow-up)

Abbreviations: AST, aspartate aminotransferase; iv, intravenous route; L, lymphocytes; N, neutrophils; po, oral route; PRP, pityriasis rubra pilaris; SSSS, staphylococcal scalded skin syndrome.

orifices (Table). A reassessment 24 hours later showed a generalized rash, predominantly in the perianal and facial area, with islands of normal skin on the trunk, follicular hyperkeratosis, and orange palmar-plantar keratoderma (Figures 1 and 2). Based on the characteristic clinical symptoms and the skin biopsy, acute postinfectious PRP was diagnosed. The patient was discharged 8 days later with topical treatment (0.1% triamcinolone acetonide in Lanette cream) and hydroxyzine. New lesions appeared

after 1 week and she started oral therapy with acitretin capsules (0.6 mg/kg/d); the parents were advised to administer the medication in darkness, opening the capsule and sprinkling the content over a piece of toast with butter or creamy chocolate spread. After 2 months of treatment, only residual hypopigmented areas remained. The cumulative dose was 606 mg of acitretin over 5 months. After 1 year of follow-up, no new flare-ups had occurred and she was discharged from the clinic.



Figure 1. Patient 1. Peeling on the soles, with cracking in the fleshy part.



Figure 2. Patient 1. Lumbar erythematous plaque, fine flaking in intergluteal folds, and palmar keratoderma.

Patient 2

An 18-month-old boy born in Russia and adopted 6 months previously was admitted for generalized rash, fever of 39°C, cold symptoms, and diarrhea. The lesions were first seen in the circumoral and periocular area, but advanced toward erythroderma of cephalocaudal progression. Because SSSS was initially suspected, intravenous cloxacillin, antipyretics, and topical fusidic acid in the periorificial areas were started.

One week later, the patient exhibited large dry, erythematous-desquamative plaques composed of micropapules located in the extensor area of the limbs, midfacial area, scalp, and back of the neck (Figure 3). All nails showed distal onycholysis, yellowish discoloration, and hyperkeratosis. The symptoms suggested acute postinfectious PRP. Prednicarbate cream, hydroxyzine, and coal tar baths were prescribed, and the patient was discharged 3 days later. Because the lesions persisted, 1 month later oral acitretin (0.5 mg/kg) was prescribed for 2 months and the symptoms improved. However, over the next 6 months, the patient experienced mild flare-ups of rash that coincided with diarrhea or cold symptoms, but always improved with oral amoxicillin-clavulanic acid. Eleven months later, he was asymptomatic, except for mild yellowish discoloration of the second left toenail.



Figure 3. Patient 2. Dry erythematous-desquamative plaques, composed of follicular micropapules and islands of healthy skin.



Figure 4. Patient 3. Orange discoloration of palms.

The most relevant data for patients 3 (Figure 4) and 4 are summarized in Table 1.

Discussion

According to the Griffiths classification published in 1980, 5 forms of PRP are distinguishable, based on age of onset, course, clinical characteristics, and prognosis.⁴ The classic adult (type I) and classic juvenile (type III) forms differ only in the age of onset, and the course is more favorable in children compared with adults. Type III is the most common form in children and tends to recur.⁶ The circumscribed juvenile form (type IV) is a common form in children that is more limited in its extension and is characterized by the appearance of hyperkeratotic plaques on the elbows and knees. Type II is an atypical form in adults, and type V is the atypical juvenile form.⁴

In 1983 Larrègue proposed a new subgroup, acute postinfectious pityriasis rubra pilaris. This form is morphologically indistinguishable from type III. Its distinctive characteristics are: a) no family history; b) onset during childhood, after the first year of life; c) presence of a previous infectious episode; d) scarlatiniform erythema, followed by the appearance of follicular papules weeks later; e) no laboratory abnormalities, except for those derived from the infectious process; f) clinical appearance similar to classic juvenile PRP, and g) acute course with good outcomes, although resolution may be slow, and no tendency toward recurrence.^{5,7,8}

PRP is a keratinization disorder caused by accelerated epidermopoiesis. Nevertheless, the acute variant is remarkably similar at first to a superantigen-mediated disease, such as SSSS, scarlet fever, toxic shock syndrome, and Kawasaki disease.⁹ Common traits in all of these entities are raspberry tongue, shiny, chapped lips, flexural (particularly perineal) erythema followed by peeling, palmar-plantar erythema, and generalized rash.¹⁰ In PRP cases, the usual lesions (follicular papules, islands of normal skin on the trunk, orange-colored palmar-plantar keratoderma) appear days or weeks later. The epidermal hyperproliferative condition can be caused by a superantigen-mediated reaction triggered by a previous infection.

There is some evidence of a link between this form of PRP and superantigen-mediated diseases:

1. Similarity (particularly in the initial stages) with scarlet fever or staphylococcal scalding, already described by other authors.^{7,9} This is attributable to the release of a large quantity of cytokines often seen in these conditions. The subsequent development of typical PRP lesions may result from the inflammatory response acting on keratinocytes.
2. Existence of variants of diseases that typically exhibit an inflammation-mediated keratinization disorder, such as psoriasis (in particular, guttate psoriasis) known to appear after bacterial infections. It has also been reported after Kawasaki disease.¹¹
3. Existence of variants of diseases, such as Wong-type dermatomyositis, that initially present as a rash similar to PRP^{12,13} and has been related to an immunologic response to a parvovirus B19 infection.^{14,15}

In conclusion, this form of PRP should be included in the differential diagnosis of diseases that are superantigen-mediated in their initial phase, from which it may be indistinguishable. We believe that this is due to massive cytokine release in the skin. In the second phase, it seems a

more specific inflammatory response—arising from cross-reactivity with antigens of the trigger microorganism or idiosyncrasy—alters the differentiation of follicular and cutaneous epithelium (as occurs in hereditary PRPs) and causes the typical lesions.

Conflicts of Interest

The authors declare no conflicts of interest.

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