O Post-infectious pityriasis *rubra pilaris* in pediatric patients

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ABSTRACT

Pityriasis *rubra pilaris* (PRP) is a rare skin condition. The etiology of PRP is unknown; however, it has been associated with infections, autoimmune diseases, and neoplasms.

Here we describe the cases of 2 pediatric patients with PRP triggered by a respiratory syncytial virus infection concurrently with obstructive bronchial syndrome. PRP resolved after treatment with topical emollients, topical corticosteroids, and calcineurin inhibitors.

Keywords: pityriasis rubra pilaris; respiratory syncytial virus; corticosteroids, calcineurin inhibitors; emollients.

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INTRODUCTION

Pityriasis *rubra pilaris* (PRP) is a rare skin condition¹ characterized by the presence of hyperkeratotic follicular papules coalescing into reddish-orange scaly plaques leaving islands of sparing and palmoplantar keratoderma.² It is an idiopathic disease; however, it has been suggested that it could be triggered by viral or bacterial infections or autoimmune or neoplastic diseases.³

PRP affects both men and women. It may occur at any age, but it has been described mostly in the first and fifth decades of life.¹ Griffiths¹ classified PRP into 5 groups, according to its clinical appearance, behavior, and prognosis. In 1995, Miralles et al. proposed the classification of human immunodeficiency virus (HIV)-associated PRP as type VI.⁴ Table 1 shows the classification of PRP and its characteristics as per Griffiths and Miralles et al.

Here we describe 2 cases of PRP in pediatric patients triggered by respiratory syncytial virus (RSV) infection concurrently with obstructive bronchial syndrome.

CASE REPORT 1

A 10-month-old male patient developed respiratory distress with concomitant fever, consistent with obstructive bronchial syndrome associated with RSV infection. Simultaneously, he developed a skin condition consisting in erythematous scaly plaques with follicular accentuation, preferentially in his elbows and knees. During the course of his condition, he also experienced palmoplantar involvement, especially in part of his trunk, face, and neck (*Figure 1*). A biopsy showed psoriasiform dermatitis with alternating parakeratosis consistent with pityriasis *rubra pilaris*; given the clinical presentation, it was diagnosed as type IV: circumscribed juvenile pityriasis. The patient's symptoms were treated with emollients and clobetasol 0.05% topical ointment every 12 hours for 2 weeks. After 4 weeks, his condition resolved completely.

CASE REPORT 2

This was a 2-year-old female patient who, 1 week after experiencing obstructive bronchial symptoms secondary to RSV infection, developed multiple coalescent pinkish-orange erythematous plaques associated with palmoplantar keratoderma, with areas of unaffected skin, especially in the legs (Figure 2). The biopsy results were compatible with PRP. It was clinically classified as type III: classical juvenile PRP. The patient was treated with optimized topical therapy, including "wet wipes" in association with topical emollients, topical corticosteroids, and calcineurin inhibitors. Consecutively, clobetasol 0.05% cream was used on palmoplantar areas every 12 hours and tacrolimus 0.1% cream was used every 12 hours for 3 weeks on body areas that showed more symptoms and special areas, such as the face. Her condition's severity decreased and, in the course of 5 weeks, it resolved completely.

Туре	Clinical presentation	Cases (%)
I. Classical adult	It affects adults. Erythematous follicular papules coalescing into plaques with islands of sparing. It appears as palmoplantar keratoderma in most patients. ¹	55
II. Atypical adult	Ichthyosiform scales, mainly in the legs. Thick, laminated palmoplantar keratoderma. There may be a thinning of hair on the scalp. ¹	5
III. Classical juvenile	Similar to type I PRP. It affects infants in their first or second year of life.	¹ 10
IV. Circumscribed juvenile	It occurs in prepubertal children. It is characterized by sharply demarcated areas of follicular hyperkeratosis and erythema of knees and elbows. ¹	25
V. Atypical juvenile	It occurs in the first years of life. It is characterized by follicular hyperkeratosis. It may be associated with sclerodermoid changes of the hands and feet. ¹	5
VI. HIV-associated	It is observed in patients with HIV. It presents as severe nodulocystic form or pustular follicular eczema resembling acne or boils. ⁴	< 5

TABLE 1. Classification and characteristics of pityriasis rubra pilaris as per Griffiths and Miralles et al.



FIGURE 1. Skin lesions of patient described in case report 1

Erythematous scaly plaques with follicular accentuation in knees (A), elbows (B), and neck (C). D: palmoplantar keratoderma.

DISCUSSION

The incidence of PRP has been estimated at a new case for every 3500 to 5000 new patients seen at the dermatology clinic.⁵

The cases described here correspond, according to Griffiths' criteria, to types III and IV of juvenile PRP.

The most common form of presentation of PRP in the pediatric age group is the juvenile form, type III,⁶ which is characterized as sporadic, presenting on the upper body as a rash that spreads progressively in a caudal manner over weeks to months.⁷ It usually starts with a single erythematous plaque on the face, trunk or chest, and scalp desquamation. Then, follicular papules on an erythematous base and a keratinous plug appear, which tend to coalesce into extensive orange erythematous areas, leaving islands of sparing.⁷ Type IV is usually observed in children younger than 10 years, with well-circumscribed, erythematous plaques with follicular hyperkeratosis located in the elbows,

knees, and pretibial areas, usually accompanied by palmoplantar keratoderma.⁷

In 1983, Larrègue described the development of PRP following infections, a new etiology of PRP, characterized by a rash occurring in children older than 1 year after an infectious process, with no family history, that resolves spontaneously.⁷ Patients usually have a history of upper respiratory tract infection, diarrhea, or other infectious symptoms that, after a few days, are associated with the development of these follicular papules that extend progressively.⁷ The mechanism that triggers PRP is unknown; Betloch et al.⁸ suggested superantigen mediation as a possible mechanism.

The diagnosis of PRP is based on the recognition of clinical characteristics and histopathological findings,³ especially in some patients in whom it is difficult to distinguish it from psoriasis.⁹ In pediatric PRP, a differential diagnosis should be made with psoriasis, seborrheic dermatitis, erythrokeratoderma,



FIGURE 2. Skin lesions of patient described in case report 2

Coalescent pinkish-orange erythrodermic scaly plaques in the whole trunk, abdomen (A), and legs, which showed islands of sparing (B). Palmoplantar keratoderma is also observed (C and D).

follicular eczema, ichthyosis, and atypical keratosis pilaris.⁷ Histopathological findings include alternating vertical and horizontal orthokeratosis and parakeratosis, epidermal acanthosis, normal granular layer, and mild perivascular lymphohistiocytic infiltrate in the dermis.³

In 2018, Roenneberg and Biedermann developed a treatment algorithm based on a systematic review of the related bibliography. They proposed that the baseline treatment for all patients with PRP should be, first of all, basic skin care with emollients with or without keratolytic agents, such as urea or salicylic acid.³ Secondly, topical corticosteroids, topical calcineurin inhibitors, and topical retinoids should be indicated before escalating to systemic therapy with retinoids or phototherapy because topical treatment may be sufficient for localized forms of PRP. Phototherapy is not recommended consistently in all patients, as the response to ultraviolet light is variable for this condition, and may even worsen it. Notwithstanding this, cases of successful treatments have been reported, so a phototherapy test prior to use may be considered.^{2,3}

If after 6 weeks of treatment PRP is not controlled, systemic retinoids are recommended. Methotrexate should be considered a second-line treatment in patients who do not respond to systemic retinoids or in whom retinoids are contraindicated.³ In the absence of a marked clinical response after 12 weeks, a biological therapy may be considered. Biological drugs used for the treatment of PRP include tumor necrosis factor-alpha inhibitors (infliximab, etanercept), interleukin (IL)-17 inhibitors (secukinumab or ixekizumab), and ustekinumab (IL-12/IL-23 inhibitor).¹⁰ The association of biological therapy with retinoids or immunosuppressants, such as methotrexate or cyclosporine, has been beneficial

in some cases.³ Although retinoids have been the most common treatment described in children and adults, a possible side effect of this treatment is premature closure of the growth plate; this risk increases with prolonged treatment at high-dose retinoids.¹¹ Further studies are required to assess the efficacy and safety of the therapeutic options for PRP.

The treatment algorithm for PRP is based on clinical experience and case reports.³ There are ongoing clinical studies in different stages of development with new molecules that could expand future therapeutic alternatives.

Janus kinase inhibitors are a relatively new class of drugs in the field of dermatology. The use of upadacitinib, a selective Janus kinase-1 inhibitor, has been reported in cases of treatment-resistant PRP, resulting in almost complete disease control after its administration.¹⁰

In relation to the 2 cases described here, both patients developed typical PRP. Both cases resolved with topical treatments that included emollients, topical corticosteroids, and topical calcineurin inhibitors, with no need to escalate to systemic therapies and thus avoiding their side effects.

To conclude, the prognosis of post-infectious PRP is generally good.¹² This report describes the case of 2 pediatric patients with a history of RSV infection that resolved after weeks of treatment with topical emollients, topical corticosteroids, and calcineurin inhibitors. We could not find any report of cases of post-infectious PRP in association with RSV; however, PRP has been associated with infections caused by varicella-zoster,13 human immunodeficiency virus,⁴ cytomegalovirus,¹² and SARS-CoV-2;14 a case of PRP following the administration of the diphtheria, tetanus, pertussis vaccine and the oral poliovirus vaccine has been reported as well.¹⁵ Therefore, this case report contributes to the bibliography on infectionrelated PRP and on the corresponding treatment of choice.

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