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Pityriasis Rubra Pilar with Preceeding Herpes Infection in Childhood – Successful Treatment with Methotrexate

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Abstract

The Pityriasis rubra pilaris (PRP) is a rare, inflammatory, chronic disease of unknown etiology characterized by follicular papules, pinkish-yellow squamous plaques and palmoplantar keratoderma. The clinical diagnosis is very important and is confirmed by biopsy of the skin. Often, the clinical presentation can be confused with other diagnosis such as psoriasis and atopic dermatitis. Currently, the first-line therapy is the systemic retinoid and other treatments like methotrexate. The association with viral disease is uncommon. Our purpose is to report a case of PRP in a young boy associated with a prior primo-infection by herpes simplex showing a favorable response to oral treatment with methotrexate and without recurrence after 10 months of follow-up.

Keywords: *Herpes infection, Methotrexate, Pityriasis rubra pilaris.*

Introduction

Pityriasis rubra pilaris (PRP) is a rare, inflammatory, chronic disease of unknown etiology. The clinical appearance of PRP is highly variable, as is the individual prognosis. The most common presentation is characterized by follicular papules, pinkish-yellow squamous plaques and palmoplantar keratoderma. There are areas of unaffected skin our "islands of sparing" (1,2). The majority of patients who develop pityriasis rubra pilaris are over 50 years of age, but it can occur in individuals of any age, race and nationality (3). The estimated incidence is 1 in 400.000 and the precise prevalence is unknonwn (4). There are reports in the literature showing an association with immune system dysregulation, abnormal vitamin A metabolism, autoimmune disease, infection, and malignancy, however, it is not common found in association with primoinfection by herpes simplex (1).

Griffiths proposed its classification into five groups based on clinical appearance, behavior and prognosis (5). A sixth type was proposed, associated with HIV (6).

The diagnosis of PRP is clinic, but can be challenging to make, because it can be confused with psoriases and atopic dermatitis. The skin biopsy confirms the diagnosis (4).

Currently, the first-line therapy is the systemic retinoid and other treatments, like methotrexate can be possible (7).

Case Report

A 20 month old male infant from Rio de Janeiro/ Brazil, presented with intermittent fever and upper respiratory tract infections. After the 15 days, the boy had onset lesions of bilateral erythema and desquamation on the scalp, face, axilla, trunk and limbs, showing symmetrical and diffuse psoriform plaques of rosy-orange colored aspect associated with islets of normal skin. Also see was a presence of lesion the region palmoplantar region showing hyperkeratosis (Figures 1 a-c).

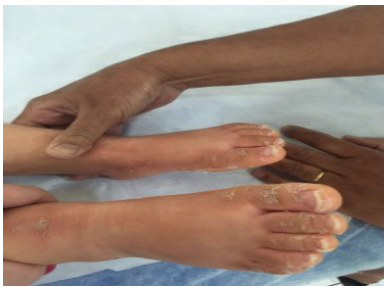
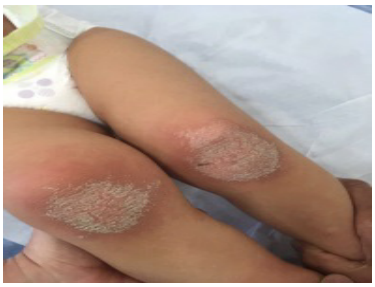


Fig1A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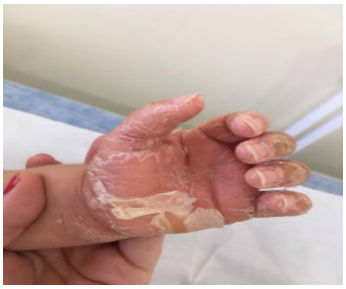
Fig1C

Fig1 (A-C). *Clinical presentation of the patient before the treatment with metotrexate.*

The emergency pediatrician made a diagnosis of atopic dermatitis and referred to allergist/ immunologist. Personal and family history for atopic diseases and psoriasis was negative. The diagnosis of atopic dermatitis was discarded and then referred to dermatology was performed resulting in a clinical diagnosis of PRP.

Complementary tests were performed revealing leukocytosis. Serology for herpes simplex virus I and II reactive IgM, and non-reactive IgG was also discovered. Serology for HIV I and II, HTLV I and II, Parvovirus B19, Ebstein Barr, Cytomegalovirus, Coxcksakie A and B, non-reactive IgM and IgG, antinuclerar antibodies non-reactive. The urinoculture and parasitological stool examination negative.

A skin biopsy was performed showing follicular infundibula filled with parakeratótic of the corneous laminae and the epidermis alternating areas of parakeratosis and orthokeratosis, with discrete perivascular inflammatory infiltrate of mononuclear cells in the superficial dermis (figure 2 a-b).

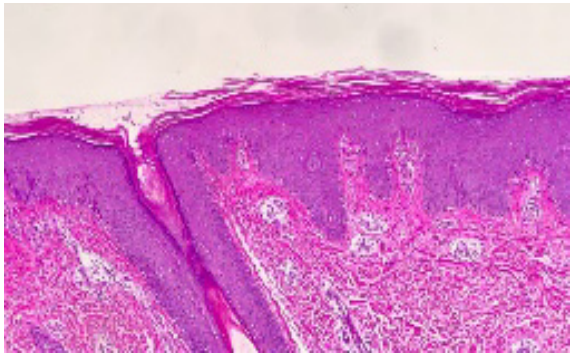
Fig2A

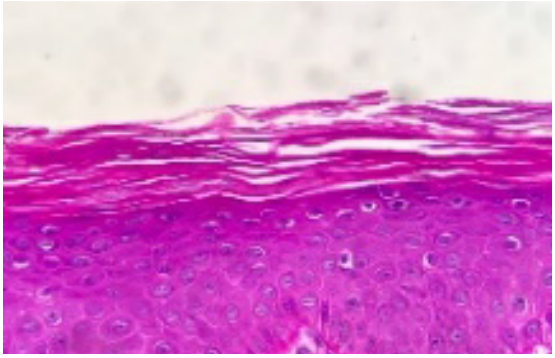
Fig2B

Fig2 (A-B). *Histopatology of a biopsy taken form a skin lesion of the right leg (hematoxylin and eosin, original magnification x 100)*

Treatment was initiated using oral with methotrexate 2.5 mg/week for 7 weeks. The patient presented with a clinical remission of symptoms. The medication was suspended by the child's mother of her own volition. Regardless of abrupt withdrawal of the the medication, the lesions did not return. After 10 months of discontinuation of treatment, follow-up did not show recurrence of the condition (figure 3 a-c).

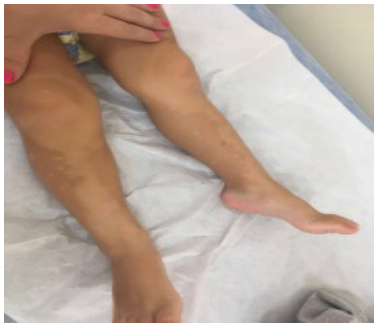
Fig3A

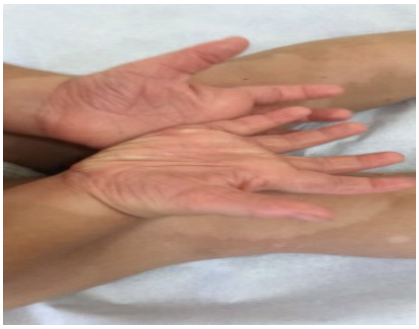
Fig3B

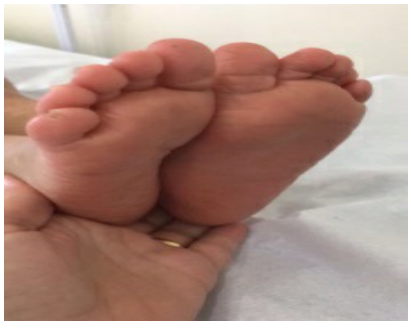
Fig3C

Fig3 (A-C). *Clinical presentation of the paciente after 4 moths of oral methotrexate.*

Discussion

PRP is rare pathology in childhood, especialy in the age under two years. There exit two periods of onset: one during the first decade of life and a second one in the sixth and seventh decade of life (4). The disease equally occurs in both sexes (8).

Diagnosis is essencially clinical and confirmed by biopsy. The histopatology reveled a follicular infundibulum filled with a parakeratótic corneous laminae and the epidermis with alternating areas of parakeratosis and orthokeratosis, referred to as the "checkerboard pattern", as in the literature (4).

Frequently, the lesions are confused with other dermatological diseases such as atopic dermatitis and psoriasis. In the case reported, initial diagnosis was atopic dermatitis and subsequently PRP. Histopathology confirmed the diagnosis. Acantholysis and focal acantholytic dyskeratosis within the epidermis have been described in the literature, features which have been suggested to be helpful in distinguishing PRP from psoriasis (4).

Most of the cases of PRP are acquired and only a few reports have been described as familial variants, including autosomal dominant as well as autosomal recessive forms (8).

The etiology is unknown, however there are reports of association with myasthenia gravis, sprue, hypothyroidism, trauma, neoplasias, infectious diseases (HIV, cytomegalovirus infection, chronic hepatitis C), but only rarely with herpes simplex primo-infection (1,8). The patient presented with upper respiratory symptoms and fever two weeks prior to the appearance of the lesions. Complementary exams revealed a reactive IgM and non-reactive IgG for *Herpes simplex*, confirming the infectioous etiology.

The patient in this case report, presented with clinical remission following only 7 weeks of treatment. Ross NA *et al*. reported that only 28% of adult and pediatric patients had entered remission before their study was completed, suggesting that PFP, can be a self-limiting condition in some cases and can persist for 2-3 years in others cases (4).

Most reports on therapy for PRP are based upon empirical data because of the lack of large-scale randomized trials (8). Retinoids are the most described treatments in both children and adult (9), but a few cases have been reported methotrexate as being successful therapy when used in children (10). In patients who fail to respond to systemic retinoids or in whom the use of retinoid therapy is contraindicated, methotrexate was proposed as a secondary-line systemic therapy (2). A cohort of 100 patients indicated that the most commonly used systemic therapies were retinoids (64%), methotrexate (42%) and light therapy (26%) (4). Griffiths reported a response rate to methotrexate of only 39%. Our option for the treatment was to prescribe the use of methotrexate for seven weeks.

Although, the literature does not agree with the abrupt cessation of methotrexate due to the possibility of rebound, our patient did not presented new lesions and after 14 weeks.

The patient had complete remission of the lesions, except showing a discreet residual hypochromia. There are no recurrence of the condition even after 10 months of follow-up.

Conclusion

This is a case rare showing the association of the PRP and primo-infection for herpes. The option of oral methotrexate therapy for pityriasis rubra pilaris in children showed a favorable response and was well tolerated by the patient and can be considered as a good option in this case, resulting in remission and sucessful treatment.

References

[1] Polcari IC, Boyod AH. Methotrexate treatment in a case of juvenile pityriasis rubra pilaris. *Pediatric Dermatology*.2017;1-2.

[2] Roenneberg S, Biedermann T. Pityriasis rubra pilaris: algorithms for diagnosis and treatment. *J Eur Acad Dermatol Venereol*. 2018;32(6):889–898.

[3] Allison DS, el-Azhary RA, Calobrisi SD, et al. Pityriasis rubra pilaris in children. *J Am Acad Dermatol* 2002;47:386-389.

[4] Ross NA, Chung HJ, Li Q, Andrews JP, Keller MS, Uitto J. Epidemiologic, Clinicopathologic, Diagnostic, and Management Challenges of Pityriasis Rubra Pilaris: A Case Series of 100 Patients. *JAMA Dermatol*. 2016;152(6):670–675.

[5] Griffiths WAD. Pityriasis rubra pilaris. *Clin Exp Dermatol*. 1980;5:105-12.

[6] Miralles ES, Núñez M, De Las Heras ME, Pérez B, Moreno R, Ledo A. Pityriasis rubra pilaris and human immunodeficiency virus infection. *Br J Dermatol.* 1995;133:990-3.

[7] Franzotti AM, Avelar JC, Cardoso TA, Pires MC, Vidigal Mdo R. Pitiríase Rubra Pilar e hipotireoidismo. *Um Bras Dermatol* . 2014 maio-junho; 89 (3): 497-500.

[8] Ivanova K, Itin P, Haeusermann P. Pityriasis rubra pilaris: treatment with biologics - a new promising therapy?. *Dermatology*. 2012;224(2):120–125.

[9] Alazemi A., Balakirski G., AlShehhi F., Lehmann S., Tenbrok K, Megahed M. Juvenile pityriasis rubra pilaris: successful treatment with methotrexate. *Clinical and Experimental Dermatology*. 2017; 43(1): 110-112.

[10] Genmmeke A. Schonlebe J, Koch A. Wollina U. Pityriasis rubra pilaris – a retrospective single center analysis over eight years. *J. Dtsch Dermatol Ges* 2010; 8: 439-44.

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