Residência RP Pediátrica
Publicação Oficial da Sociedade Brasileira de Pediatria
ISSN-Online: 2236-6814

Submitted on: 16/04/2023
Approved on: 23/05/2023

INTERACTIVE CLINICAL CASE

Sclerodermiform alterations in the hands of an adolescent.

Gabriel Castro Tavares[1], Alexandra Rech Vieira[1], Tullia Cuzzi Teichner[2], Nurimar Conceição Fernandes[1]

This is an 18-year-old female adolescent, with generalized pruritic skin lesions since the age of 3 months, treated only with antihistamines. She reported no comorbidities, allergies, or family history of skin diseases. She has a history of unplanned pregnancy for a year. Upon examination, follicular keratosis was noted alternating with islands of healthy skin on the anterior and posterior torso (Figure 1); ichthyosiform desquamation on the lower limbs (Figure 2); sclerodermiform changes in the hands and feet (Figure 3) and palmoplantar keratosis.

1. What is the main diagnostic hypothesis?
a) Psoriasis
b) Scleroderma
c) Progressive symmetrical erythrokeratoderma
d) Pityriasis rubra pilaris

A biopsy of the lesion on the back was performed, which showed psoriasiform dermatitis with horizontally alternating orthokeratosis and parakeratosis (Figure 4), compatible with the clinical hypothesis of pityriasis rubra pilaris (PRP).

Pityriasis rubra pilaris is a rare inflammatory papulosquamous dermatosis. Genetic factors, changes in vitamin A metabolism and infections have been proposed as mechanisms that trigger the disease, but its etiology and pathogenesis are still unknown. It has



Figure 1. Follicular keratosis alternating with islands of healthy skin on the torso.

[1] Hospital Universitário Clementino Fraga Filho, Dermatologia - Rio de Janeiro - Rio de Janeiro - Brasil.
[2] Hospital Universitário Clementino Fraga Filho, Patologia - Rio de Janeiro - Rio de Janeiro - Brasil.

Correspondence to:
Gabriel Castro Tavares.
Hospital Universitário Clementino Fraga Filho. R. Prof. Rodolpho Paulo Rocco, 255 - Cidade Universitária da Universidade Federal do Rio de Janeiro, Rio de Janeiro - RJ, 21941-617. E-mail: gabrielcastrotavares@gmail.com

Resid Pediatr. 2024;14(1). DOI: 10.25060/residpediatr-2024.v14n1-1094

This work is licensed under a Creative Commons Attribution 4.0 International License.

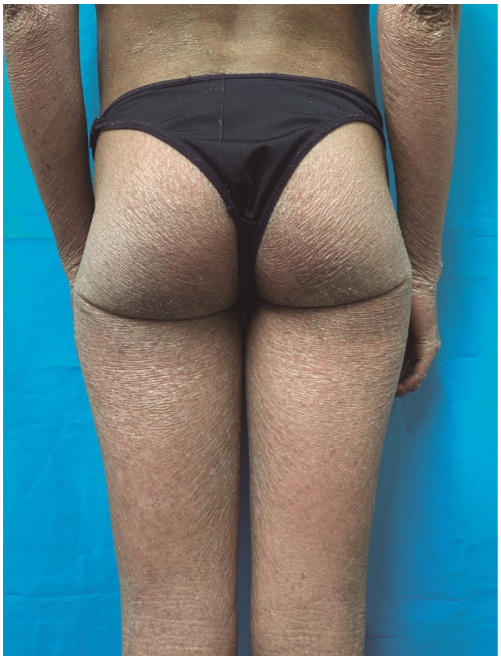


Figure 2. Ichthyosiform scaling on the lower limbs.



Figure 3. Sclerodermiform changes in the hands.

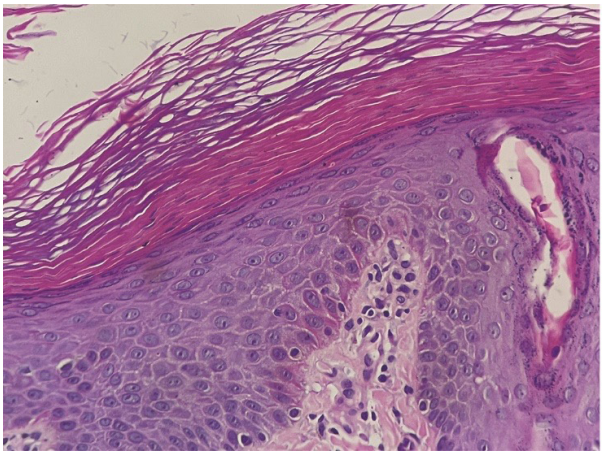


Figure 4. Horizontally alternating parakeratosis.

been described in association with HIV, drugs, autoimmune diseases and visceral neoplasms. It affects all races and sexes equally, with distribution peaks in the first and fifth decades of life.

2. What is the subtype of the disease in question?
a) Adult classic
b) Atypical of adults
c) Youth classic
d) Juvenile atypical

The juvenile atypical subtype is the least common (5% of cases) and accounts for the majority of familial cases. It appears in the first years of life and has a chronic course. It is characterized by follicular keratosis, erythema, ichthyosiform scaling on the lower limbs, gross palmoplantar keratosis and sclerodermiform changes on the hands and feet.[1,2,3]

3. What is the systemic treatment of choice?
a) Methotrexate
b) Acitretin
c) Prednisone
d) Azathioprine

Acitretin is the systemic medication of choice, but there is a risk of teratogenicity. Narrowband UVB phototherapy is a second-line therapeutic option, as well as other immunosuppressants. Considering the impossibility of guaranteeing effective contraception and the unavailability of phototherapy, treatment consisted of the topical use of emollients and keratolytics, with partial improvement of the condition.[4,5]

REFERENCES

1. Zaouak A, Chamli A, Koubaa W, Hammami H, Fenniche S. Pityriasis rubra pilaris. Presse Med. 2019 Jun;48(6):730-731. doi: 10.1016/j.lpm.2019.05.009.
2. Roenneberg S, Biedermann T. Pityriasis rubra pilaris: algorithms for diagnosis and treatment. J Eur Acad Dermatol Venereol. 2018 Jun;32(6):889-898. doi: 10.1111/jdv.14761.
3. Eastham AB. Pityriasis Rubra Pilaris. JAMA Dermatol. 2019 Mar 1;155(3):404. doi: 10.1001/jamadermatol.2018.5030.
4. Abrouk M, Nakamura M, Zhu TH, Farahnik B, Koo J, Bhutani T. Atypical Presentation of Pityriasis Rubra Pilaris: Challenges in Diagnosis and Management. Cutis. 2021 Sep;108(3):E33-E35. doi: 10.12788/cutis.0372.
5. Wang D, Chong VC, Chong WS, Oon HH. A Review on Pityriasis Rubra Pilaris. Am J Clin Dermatol. 2018 Jun;19(3):377-390. doi: 10.1007/s40257-017-0338-1.