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Rare etiology of maculopapular rash

Etiologia rara de exantema maculopapular

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Keypoints

What is known

– Pityriasis rubra pilaris is a rare inflammatory dermatosis with unknown etiology.
– It can present in any age, being more common at 6 to 7 years of age and fifth to sixth decade of life. The incidence is unknown, appearing to be similar in both females and males.
– It usually presents has a cephalocaudal hyperkeratotic scaly-erythematous maculopapular rash that progresses to a generalized erythroderma with islands of sparing.

What is added

– Due to its rarity, a high index of suspicion is needed. It should be suspected in the presence of maculopapular hyperkeratotic rashes with erythroderma.
– Although spontaneous remission can occur, PPR usually persists for several years, having a significant impact on quality of life.

A sixteen-year-old boy, with history of a suspected cutaneous antibiotic adverse reaction, was admitted to the emergency department due to a 10 day evolution pruriginous scaly-erythematous maculopapular rash. The lesions appeared on face with caudal progression. He was treated on the third day with oral and topical corticosteroid and oral antihistamine. On the sixth day, he initiated cough and, on the ninth day, fever. Three weeks earlier, he had an upper airway infection medicated with a mucolytic agent (ambroxol), paracetamol and nasal phenylephrine. Ingestion of other drugs was denied.

Physical examination revealed exuberant erythematous and coalescing maculopapules on face, trunk, perineum and limbs, lip fissures and tonsillar hyperemia (Figs. 1-3). Laboratory workup revealed normal hemoleucogram, normal kidney and liver function, CRP 20.1mg/L and negative serological testing for CMV, HIV, EBV, parvovirus B19 and mycoplasma. Paul-Bunnell reaction, antistreptolysin O titer, group A Streptococcus rapid antigen detection test and throat culture were also negative. Herpes simplex virus I/II IgM antibody was negative and IgG was inconclusive. The previous treatment with ambroxol and paracetamol led to toxicodermia suspicion. The case was discussed with Dermatology and he was started on hydroxyzine and prednisolone with mild erythema improvement but with the appearance of new lesions. Because of the scarlatinous appearance of the lesions, a single dose of intramuscular penicillin was administered, without any improvement. A skin biopsy was then performed and the patient was discharged to Dermatology

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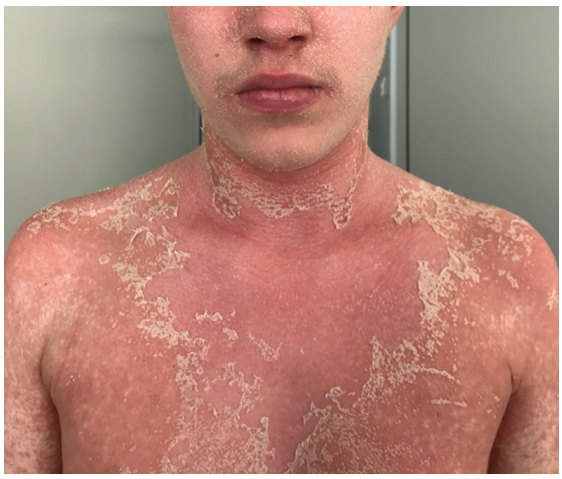

Figure 1. Clinical image showing exuberant scaly-erythematous maculopapular rash on face and trunk.

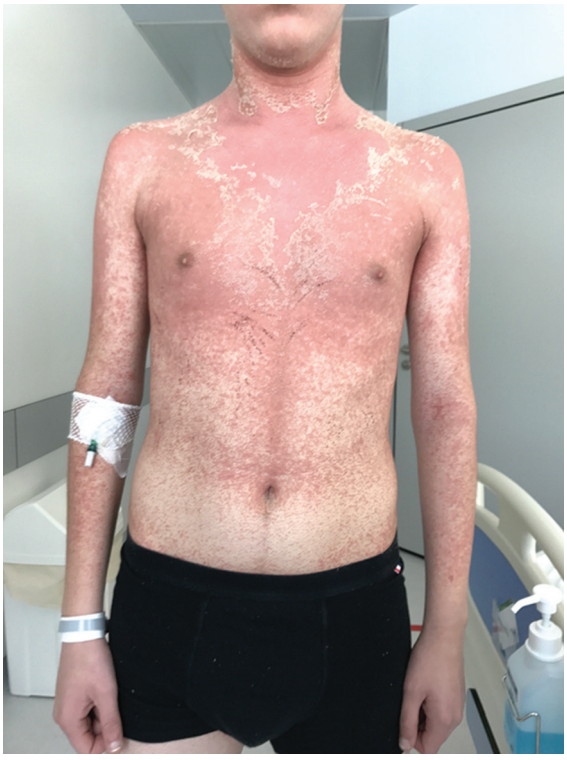

Figure 2. Clinical image showing scaly maculopapular rash on trunk, sparing the hands.

consultation on weaning prednisolone. The skin biopsy results revealed pityriasis rubra pilaris (PRP) and the patient initiated systemic isotretinoin for 16 months with

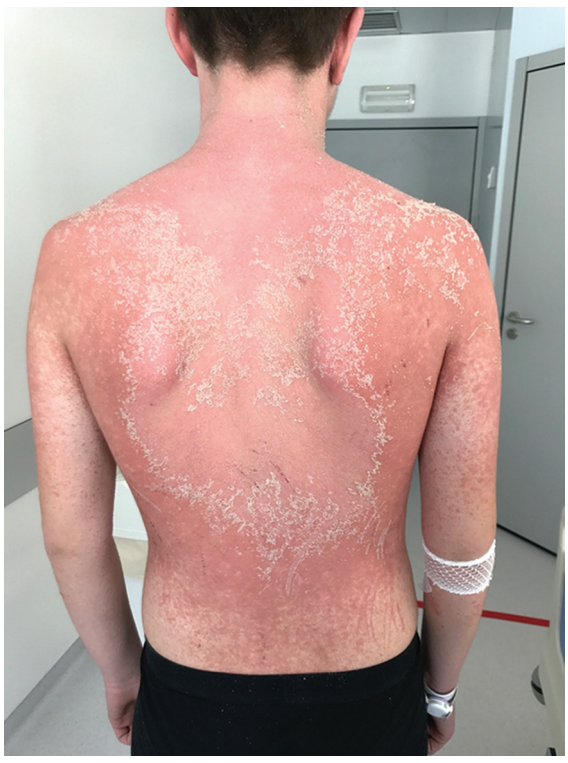

Figure 3. Clinical image showing marked scaly lesions.

improvement, but with persistence of desquamation and pruritus. Three years later, a tenuous erythema remains along with keratoderma and desquamation.

PRP is a rare idiopathic inflammatory dermatosis that typically presents as cephalocaudal follicular hyperkeratotic papules that progress to a generalized erythroderma with islands of sparing. Most cases are sporadic, but some (< 5%) have autosomal dominant transmission[1-3]. Familial cases have been linked to gain of function mutations in the caspase recruitment domain family member 14 (CARD14 gene), leading to nuclear factor-kB activation and cutaneous inflammation[1]. Drug induced PRP has also been described, being tyrosine kinase inhibitors and phosphoinositide 3-kinase inhibitors the most frequently reported. It was classified by Griffiths into 5 subtypes: type I-classical adult, type II-atypical adult, type III-classical juvenile, type IV-circumscribed, type V-atypical juvenile. Recently, type VI-HIV associated PRP was added[1,2].

Circumscribed PRP is the most common type in children. Due to its rarity, a high index of suspicion is needed. The diagnosis is based on clinical and histopathological findings[1-3]. There is no treatment consensus[1-4]. The first line therapies are topical emollient,

keratolytic agents and systemic retinoids. Methotrexate, phototherapy and biologic IL-17 or IL-23 inhibitor are also effective[1,2,4]. Although spontaneous remission can occur, PPR usually persists for several years, having a significant impact on quality of life.

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Conflicts of interest

None.

Ethical disclosures

Protection of human and animal subjects. The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

Use of artificial intelligence for generating text. The authors declare that they have not used any type of generative artificial intelligence for the writing of this manuscript, nor for the creation of images, graphics, tables, or their corresponding captions.

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