Case Report

Circumscribed juvenile pityriasis rubra pilaris

局限性幼年型毛髪紅糠疹

OM Moreno-Arrones, B Perez-Garcia, A Pecharroman

Pityriasis rubra pilaris (PRP) is an uncommon papulosquamous inflammatory dermatosis of unclear aetiology. Its most remarkable clinical findings are palmoplantar hyperkeratosis, orange-red scaling plaques and hyperkeratotic follicular papules. Its extension may vary from localised subtypes to generalised skin involvement with well-demarcated areas of spared skin ("islands of sparing") to erythroderma. Griffiths' classification is used to distinguish PRP in five clinical subtypes. Diagnosis is based on clinical and histopathological findings. The main differential diagnosis in a patient with circumscribed juvenile PRP (type IV) is psoriasis although keratosis pilaris, lichen spinulosus and follicular ichthyosis must also be ruled out. Treatment of pityriasis rubra pilaris remains a controversial topic due to the lack of large-scale clinical randomised trials and its unpredictable spontaneous resolution.

Keywords: Children, differential diagnosis, papulosquamous dermatosis, pityriasis rubra pilaris

Introduction

Pityriasis rubra pilaris (PRP) is an uncommon papulosquamous inflammatory dermatosis of unclear aetiology. Its most remarkable clinical findings are palmoplantar hyperkeratosis, orange-red scaling plaques and hyperkeratotic follicular papules. Its extension may vary from localised subtypes to generalised skin involvement with well-demarcated areas of spared skin ("islands of sparing") to erythroderma. Griffiths' classification
is used to distinguish PRP in five clinical subtypes according to the age of onset, duration and clinical presentation of the disease. Recently; a sixth subtype, associated to HIV, was added.\(^4\) Whereas type I PRP is the most frequent presentation, type III and IV are much more common in children.\(^1\)

**Case report**

A previously healthy 3-year-old Maghrebian boy presented with a two-month history of non-pruritic cutaneous lesions on both hands, feet and knees. Physical examination revealed a waxy orange-red palmoplantar hyperkeratosis with fissures (Figure 1). On both knees and ankles erythematous scaly plaques were present (Figure 2). An orange-red waxy keratoderma affected the soles (Figure 3). "Nutmeg grater" sign was present on the back of the proximal phalanges of the fingers (Figure 4). The mucosa, nails and elbows were spared. Biopsy specimens were obtained from representative lesions of the left knee. Histopathological examination revealed papillomatosis with marked hyperkeratosis, alternating orthokeratotic and parakeratotic areas. Dermal capillaries were dilated and a mild mononuclear inflammatory cell infiltrate was present (Figure 5). A diagnosis of type IV PRP was made based on the clinical and histopathological findings. Therapy was initiated...
Type IV pityriasis rubra pilaris

with medium potency topical corticosteroid. The patient was symptom-free four months after the introduction of treatment. One year later, there was no relapse of the condition at the follow-up visit.

Discussion

Type IV PRP (circumscribed juvenile) represents approximately 25% of PRP. It has a relapsing and remitting unpredictable course and very rarely progresses to erythroderma. It has been published that a third of patients affected with this PRP subtype achieve disease remission within three years.

It typically affects the elbows and knees, appearing as follicular hyperkeratotic papules that tend to coalesce. In addition, palmoplantar hyperkeratosis is not an uncommon finding. Its age of onset is usually before the adolescence.

Psoriasiform dermatitis with alternating vertical and horizontal ortho- and parakeratosis in a checkerboard pattern is a characteristic histopathological finding of PRP. Acanthosis, spongiosis and a mild lymphohistiocytic inflammatory cell infiltrate in dermis might be present.

The main differential diagnosis in a patient with circumscribed juvenile PRP is psoriasis although keratosis pilaris, lichen spinulosus and follicular ichthyosis must also be ruled out.

Treatment of pityriasis rubra pilaris remains a controversial topic due to the lack of large-scale clinical randomised trials and its unpredictable spontaneous resolution. For localised disease, topical treatment with corticosteroids, vitamin D analogues and calcineurin inhibitors are generally the mainstays of treatment. Systemic drugs such as isotretinoin, methotrexate, ciclosporin, azathioprine, ustekinumab and TNF-alpha inhibitors are usually reserved for recalcitrant and severe disease. Although PRP may be exacerbated by ultraviolet exposure, it has been reported that some patients may respond to phototherapy.

Conclusion

Pityriasis rubra pilaris is a rare skin condition that may affect children. As psoriasis is its main differential diagnosis, a histopathological study is mandatory. Similar to psoriasis, its evolution is unpredictable and thus the patient might require chronic treatment.
References


