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Case Report

Circumscribed juvenile pityriasis rubra pilaris

局限性幼年型毛髮紅糠疹

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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.

毛髮紅糠疹（PRP）是一種罕見及病因不明的丘疹鱗屑性炎性皮膚病。其最顯著的臨床表現是掌蹠角化過度、橙紅色的鱗片剝落狀斑塊和過度角化的毛囊性丘疹。它的影響範圍變化可以由局部亞型至泛發型伴隨著界限清晰而不受影響的正常皮膚部份（"倖免的島嶼"），甚至是全身的紅皮病。常用的格雷佛氏分類把毛髮紅糠疹分爲五個臨床亞型，診斷是建基於臨床表現和組織病理學的發現。局限性幼年型毛髮紅糠疹（第四型）患者的首要鑑別診斷是銀屑病，其他要排除的皮膚病包括毛髮角化病、小棘苔蘚和濾泡型魚鱗癬。毛髮紅糠疹的治療仍有爭議之處，源於其缺乏大規模的臨床隨機對照試驗及本身難以預計的自我痊癒可能。

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

關鍵詞：兒童、鑑別診斷、丘疹鱗屑性皮膚病、毛髮紅糠疹

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Introduction

Pityriasis rubra pilaris (PRP) is an uncommon papulosquamous inflammatory dermatosis of unclear aetiology.[1,2] 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[3] 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 initiatied

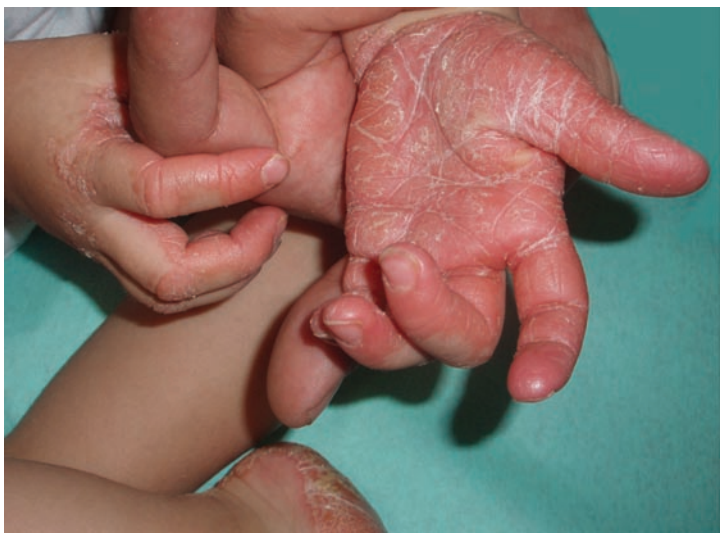

Figure 1. Waxy orange-red palmoplantar hyperkeratosis with fissures.

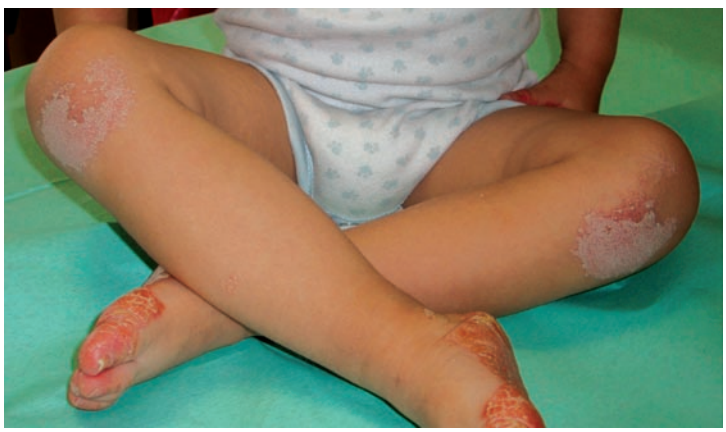

Figure 2. Erythematous scaly plaques on both knees.

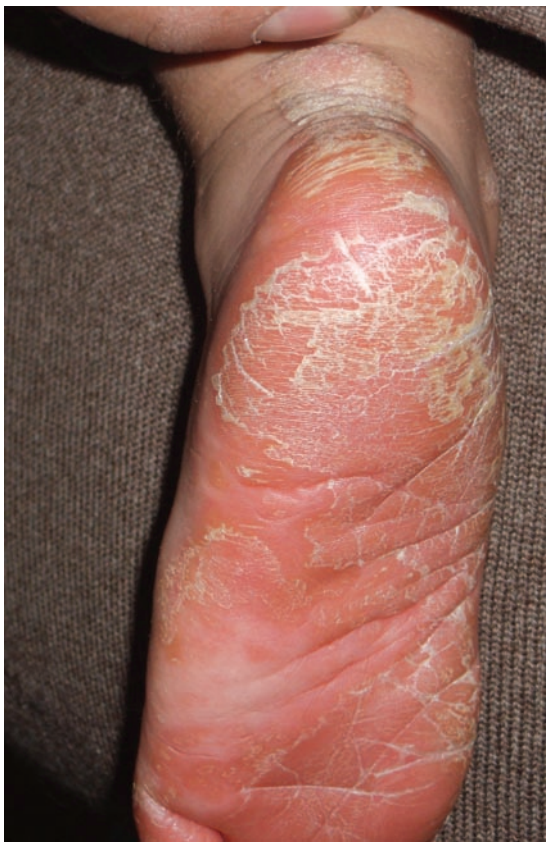

Figure 3. Orange-red waxy keratoderma on the soles.

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.[1] 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.[5]

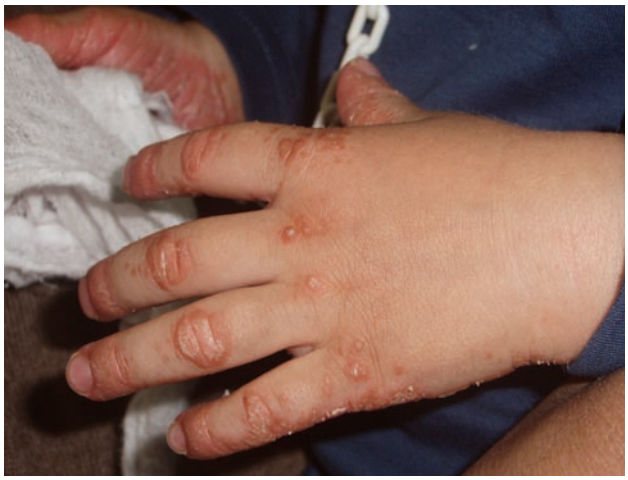

Figure 4. "Nutmeg grater" sign.

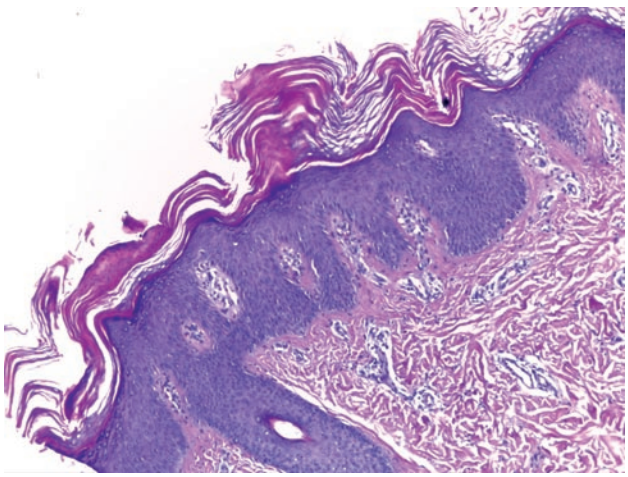

Figure 5. Papillomatosis with marked hyperkeratosis, alternating orthokeratotic and parakeratotic areas. Dermal capillaries are dilated and a mild mononuclear inflammatory cell infiltrate is present (H & E, x 20).

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.[6]

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.[7] 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.[8]

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.

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