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Case Report

Pityriasis rubra pilaris: an adult presenting as erythroderma with follicular papules

毛髮紅糠疹：首發病徵為紅皮症及毛囊丘疹的成人病例一宗

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This is a report of a 60-year-old Chinese man who presented with a two weeks' history of erythroderma and keratoderma. The clinical features and skin biopsy confirmed the diagnosis of adult onset classical pityriasis rubra pilaris. Oral acitretin was used with satisfactory response.

患者男性，60 歲，華人，首發病徵為紅皮症及毛囊丘疹，歷時兩週。其臨床表現及皮膚活檢確診為成人典型毛髮紅糠疹。以口服 acitretin 治療獲得滿意效果。

Keywords: Adult, classical, pityriasis rubra pilaris

關鍵詞：成人，典型，毛髮紅糠疹

Introduction

Pityriasis rubra pilaris (PRP) is an uncommon cause of erythroderma. The aetiology is unknown. The condition is classified according to the age of onset and clinical presentation. Both familial and acquired forms are present with different prognosis. The following is a case report of a 60-year-old man with the disease.

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Case Report

The patient is a 60-year-old Chinese man with good past health. He presented with progressive scaly generalised skin eruption starting from the face and upper body. The skin rash was not itchy and he had no fever. He did not take any medication or herbal medicine before the onset of rash. There was no associated joint pain or swelling. There were no precipitating factors and no family history of skin diseases.

On physical examination, there was exfoliative dermatitis with orange-red discoloration and islands of sparing of normal skin over abdomen, elbows and knees (Figure 1). Palmoplantar keratoderma and follicular papules on legs were detected (Figure 2). Nail involvement was not

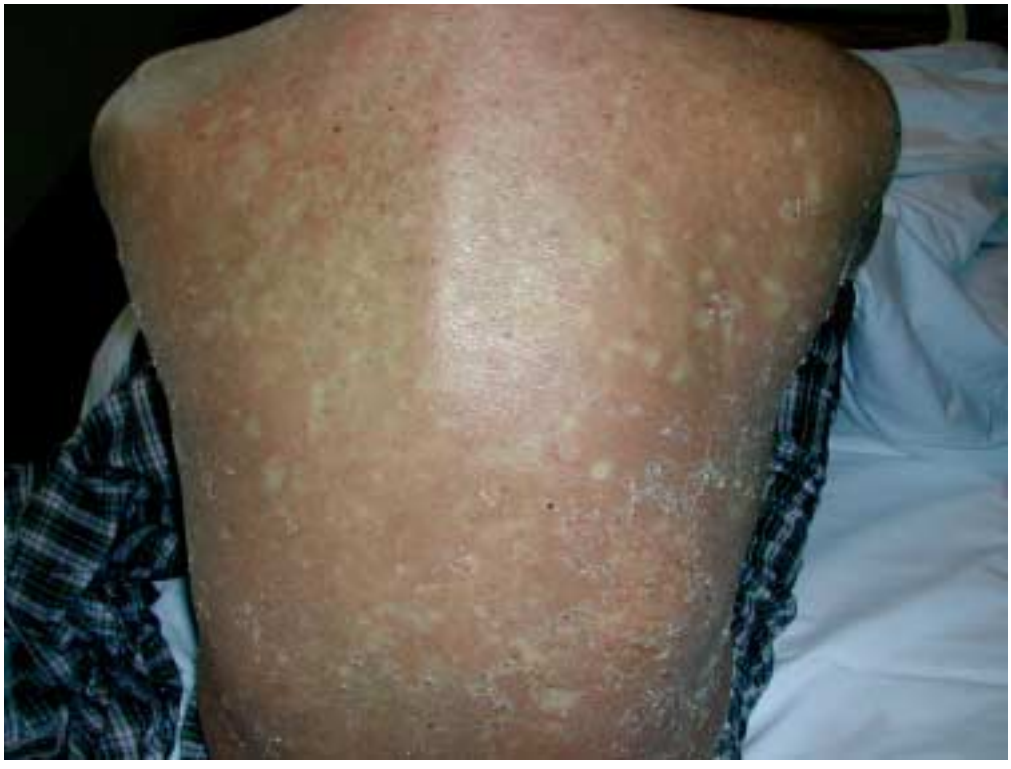

Figure 1. Erythroderma with an orange-red tinge and islands of sparing were noted on the trunk.

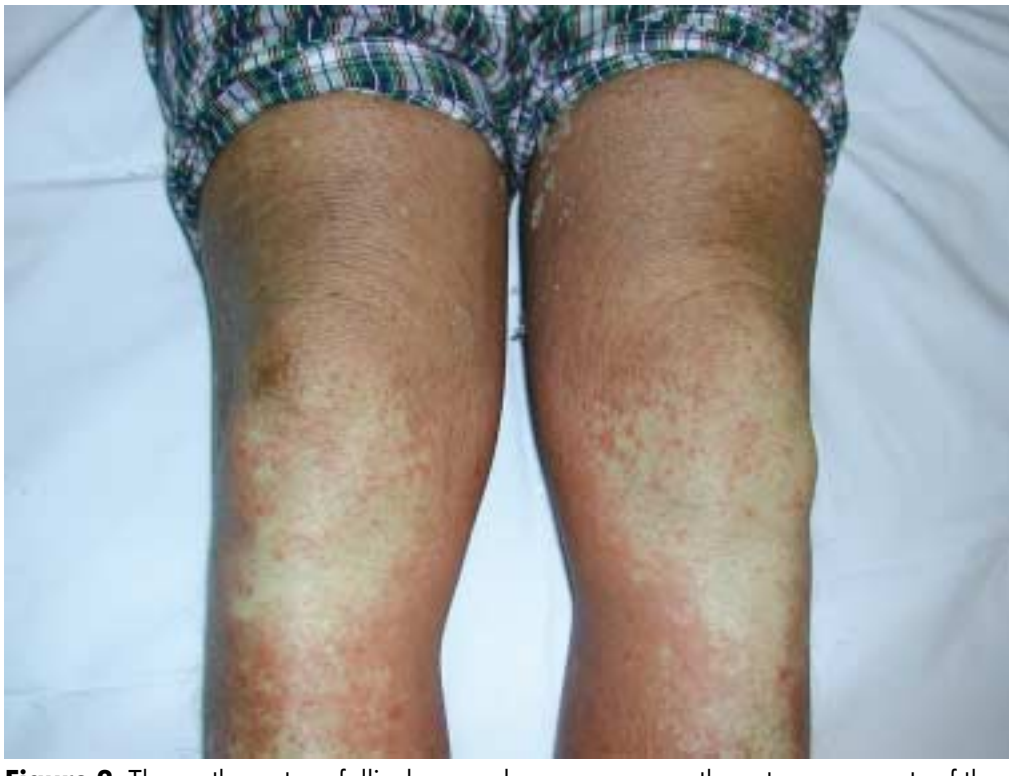

Figure 2. The erythematous follicular papules were seen on the extensor aspects of the lower limbs.

present. There was no lymph node enlargement or hepatosplenomegaly. The differential diagnoses included psoriasis, eczema, cutaneous T-cell lymphoma and drug eruption.

Incisional skin biopsy showed alternating orthokeratosis and parakeratosis in both vertical and horizontal directions, follicular parostial parakeratosis and follicular plugging. There was upper dermal perivascular infiltrate (Figure 3). The diagnosis was pityriasis rubra pilaris. The age of onset and the clinical pattern of the patient belonged to the type I (classic adult) according to the classification proposed by Griffiths.

The management included bed rest, emollient use and acitretin 0.5 mg/kg daily orally. The skin condition improved with decrease in exfoliation and erythema after eight weeks of treatment. He remained in remission two years after the initial presentation.

Discussion

Pityriasis rubra pilaris (PRP) is a rare chronic erythematous papulosquamous disorders. The condition has bimodal age distribution in the first and fifth decades.[1] Griffiths proposed a classification according to the age of onset, duration of disease and clinical pattern.[2] The most common type is the classical type of adult onset (type I, >50%), compared with the classical juvenile onset (type III, 10%). The atypical adult (type II), circumscribed juvenile (type IV) and atypical juvenile (type V) comprise 5%, 25% and 5% of all cases of PRP respectively.[3] The familial type has an autosomal dominant mode of inheritance in the types II, IV

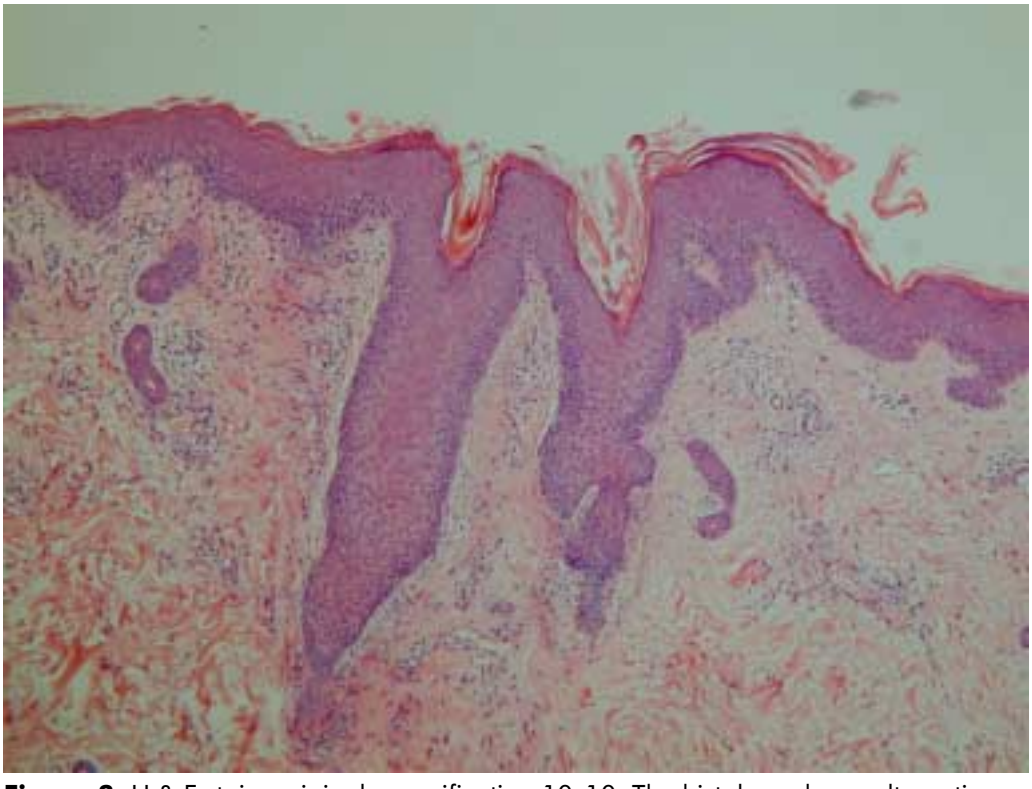

Figure 3. H & E stain, original magnification 10x10. The histology shows alternating parakeratosis and orthokeratosis in both vertical and horizontal directions. Parakeratosis was seen around the follicular openings, creating a shoulder effect. Irregular acanthosis accompanied a mild inflammatory infiltrate in the upper dermis was noted.

and V. The aetiology remains obscured. Vitamin A metabolism, genetic factors and infections have been proposed to play a role in the pathogenesis.[4]

Our patient presented with a classical picture of type I PRP, characterised by orange-red erythroderma, palmoplantar keratoderma, keratotic follicular papules and islands of normal skin. No definite precipitating factors were identified in our case. The eruption started on the face and upper trunk, and then spread caudally. The subsequent resolution of the erythroderma after systemic retinoid therapy was consistent with the favourable clinical course of type I PRP.

The atypical types are of longer duration and associated with ichthyosiform scales, sclerodactyly and alopecia.[5] In children with PRP, most cases present in late teens but PRP may occur before two years of age.[6] It is most common at elbows, knees and back in children. The diagnosis of PRP primarily depends on clinical features. Skin biopsy may further support the diagnosis when histology showed the characteristic alternating ortho-keratosis and parakeraotsis in both vertical and horizontal directions and follicular plugging.

Types I and III often follows a natural remitting course.[7] Topical steroid ointment can be used in limited diseases. Systemic treatment is necessary in extensive diseases.[8] Systemic retinoids including vitamin A, isotretinoin, etretinate and acitretin have been used. Seventeen patients (71%) showed 25% to 75% response after 16 weeks of oral retinoids in a retrospective review on 24 acquired PRP adults patients over 10 years' period.[3] Other systemic therapy includes methotrexate, psoralen plus ultraviolet A, azathioprine and stanozolol.[5]

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