Hong Kong J. Dermatol. Venereol. (2016) **24**, 129-134

Views and Practice

Pityriasis rubra pilaris: Not just skin deep a case series

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Case series

Pityriasis rubra pilaris (PRP) is an idiopathic uncommon dermatosis characterised by follicular and palmoplantar erythroderma, scaly plaques and "islands of sparing". PRP occurs in all races and affects both genders equally. Diagnosis can usually be made clinically. Anxiety and depression are common amongst chronic sufferers of this dermatosis. It is of utmost importance to explore the impact of PRP on mood and quality of life during clinical reviews. Further research may be required to determine the prevalence of depression in PRP sufferers and to assess the impacts on quality of life.

PRP may be hereditary or acquired and affects both genders equally. There are six subtypes and diagnosis is based essentially on clinical grounds. We would like to present five cases of PRP and review its psychological impact.[1-3]

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Case 1

A 62-year-old Caucasian man was seen by a private dermatologist in December 2009 for erythroderma (Figure 1). His background consisted of ischaemic heart disease, hypertension, hyperlipidaemia, type 2 diabetes and osteoarthritis. A diagnosis of exfoliative PRP was made, confirmed on histopathology. He was commenced on acitretin 50 mg daily with topical betamethasone dipropionate cream twice daily but flared necessitating admission to hospital under our team in April 2010 for wet dressings. Acitretin was weaned to 10 mg daily and islands of spared skin appeared. By the end of 2011 only a few residual patches of PRP could be seen, mainly on the face, and acitretin was stopped.

However, in 2012 and 2013 the patient had two further flares requiring hospital admission, despite courses of acitretin and oral methotrexate. He became very depressed due to the appearance and odour of his skin, continual itch, the chronic course of PRP, and inability to work. A short course of anti-depressives was required. Furthermore in 2013 his PRP course was complicated by the development of DISH, requiring cessation of acitretin. He then developed polymyalgia rheumatica for which a fourth hospital admission and oral prednisone were required.

Another flare in PRP occurred in 2014, accompanied by depression. The patient was admitted to hospital and his skin improved on oral methotrexate 10-20 mg weekly. As of 2015,

his PRP has been gradually improving; he no longer has arthralgia and is being weaned off methotrexate, prednisone and antidepressants. Moreover, he is thrilled about the remarkable improvement of his stubborn and irritating ectropion (Figure 2).

Case 2

A 67-year-old Caucasian man presented to the dermatology clinic in May 2011 with erythrodermic PRP. Prior to this he had a two month history of a generalised erythematous rash which was thought to be a drug reaction and treated with oral prednisone. His medical history included atrial fibrillation, hyperlipidaemia, insomnia and anxiety secondary to family stressors.

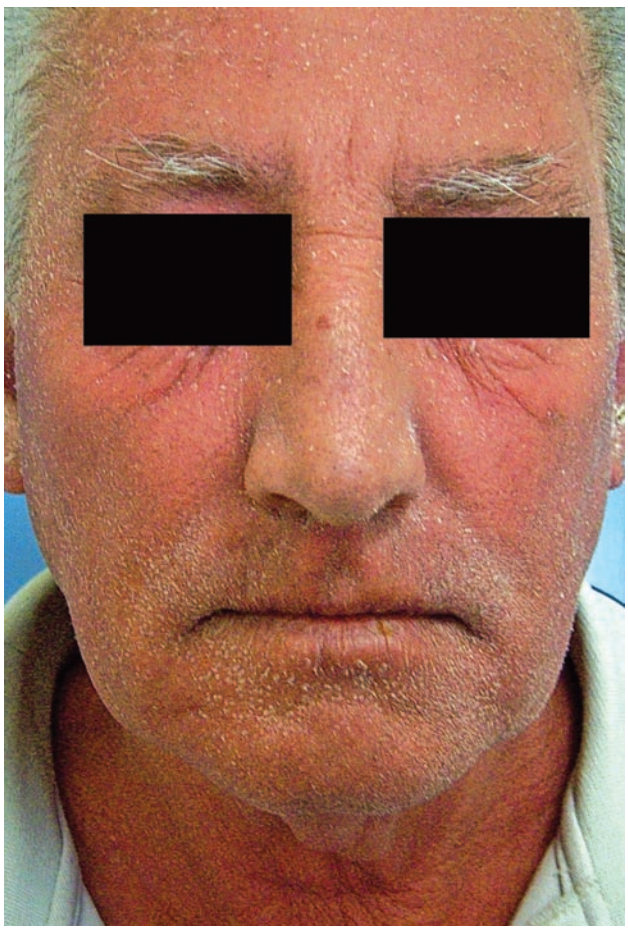

Figure 1. Erythrodermic in 2010.

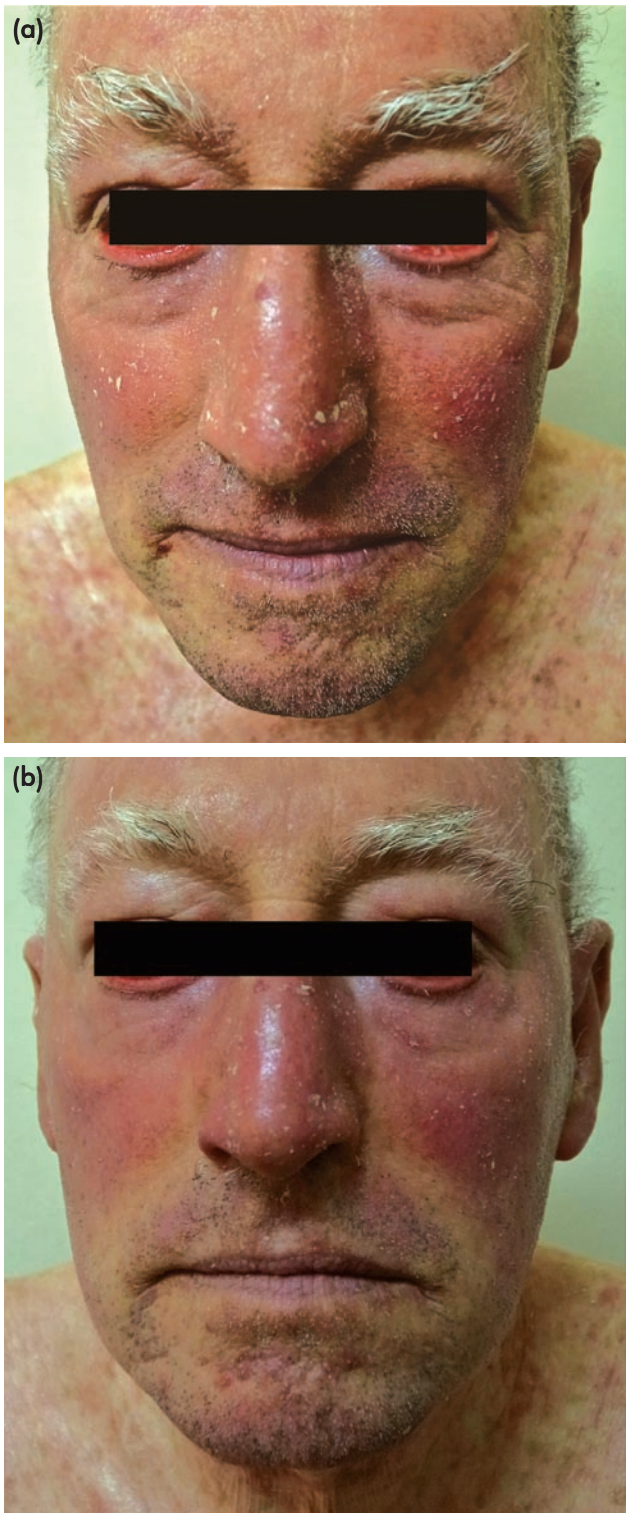

Figure 2. (a) Bilateral ectropion. (b) Bilateral ectropion improving. Also note perioral clearing.

On examination he was erythrodermic with follicular hyperkeratosis and keratoderma. There were small islands of sparing and a diagnosis of PRP was made. There was also mild ectropion. He was admitted to hospital for daily wet dressings, commencement of acitretin and cessation of prednisone. His PRP improved and over the next 12 months and acitretin was weaned down to 10 mg thrice weekly and then withdrawn altogether. At present, his PRP continues to improve with the use of emulsifying ointment daily. He describes his disease progress as 'drawn-out' and 'tries to keep busy' in order to lift his mood. There remain mildly erythematous plaques over the torso, arms and legs, involving less than 25% of body surface area.

Case 3

A 78-year-old Caucasian female presented to the Emergency Department in December 2011 with generalised PRP. This was preceded by six weeks of an erythematous desquamating rash. The rash commenced on the central chest then spread outwards to involve the entire body. She had seen another dermatologist who diagnosed PRP and commenced methotrexate with no effect. Her medical background included hypertension, hyperlipidaemia, gastro-esophageal reflux and mitral valve prolapse. Of relevance is a family history of depression and her son had committed suicide from depression.

She was admitted to hospital for wet dressings and commencement of acitretin at 10 mg daily. Methotrexate was ceased. Itch was particularly problematic, requiring the addition of antihistamines and doxepin. Malignancy screening was negative. Over the next six months the PRP slowly improved, allowing for a gradual reduction of acitretin down to 10 mg second daily. By nine months, as the rash had completely settled, acitretin was stopped. The patient remains well to date with no recurrence of PRP.

Case 4

A 41-year-old Argentinian man developed an erythematous rash on the scalp in August 2013, during a stressful period at work. The rash had spread caudally and the initial diagnosis was guttate psoriasis. Local doctors tried topical and oral steroids with little relief.

By January 2014, this patient became erythrodermic and was admitted to hospital for dermatological management. On examination there was widespread erythroderma with desquamation, distinct islands of sparing over the trunk (Figure 3), and orange/red palms (Figure 4) and soles: features that were strongly indicative of PRP. Infection and malignancy screens were negative. The patient was frustrated at having to suffer the skin discomfort and disfigurement for months without an earlier diagnosis of PRP. Moreover, he was anxious about the possibility of an underlying malignancy, and depressed about the uncertain course of his skin disease.

Initially he was treated with intensive topical steroids and wet wraps. These were changed to oral acitretin 25 mg daily fairly quickly with good effect. Acitretin was gradually weaned then ceased in October 2015. This patient continues to improve with less than 20% body surface area affected. He is no longer clinically depressed although he remains guarded regarding the long-term prognosis.

Case 5

A 74-year-old Chinese man presented with a pruritic rash in May 2014, on a background of hypercholesterolaemia, hypertension, ischaemic heart disease and osteoporosis. The rash first appeared on the abdomen then spread centripedally with sparing of the face and limbs. Biopsy showed changes consistent with PRP, and the overall clinical picture was consistent with PRP despite the unusual distribution.

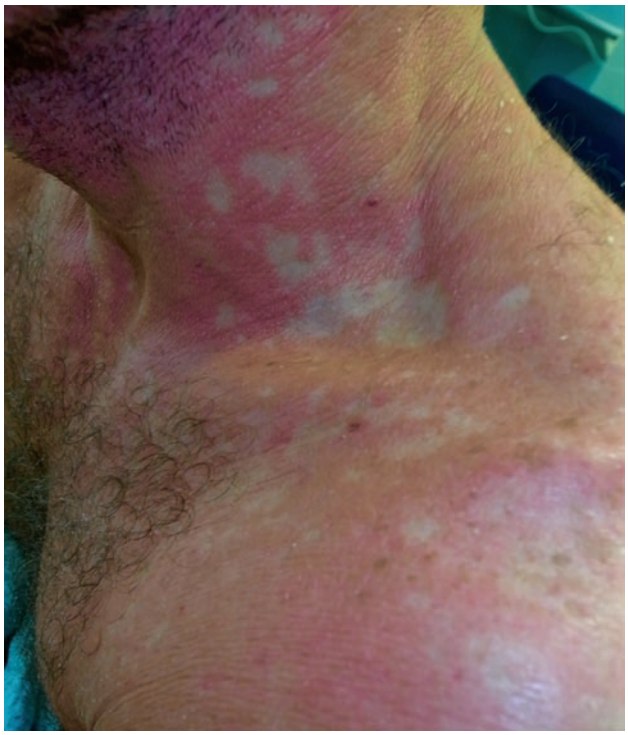

Figure 3. Islands of sparing seen in this patient with PRP.

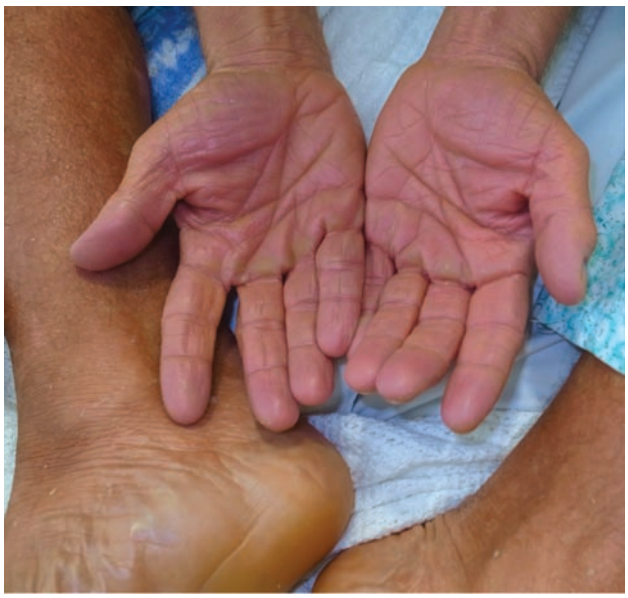

Figure 4. Characteristic colour of the thickened skin on the palms and soles.

Acitretin 10 mg daily and emollients were commenced in June 2014. It was weaned down to 10 mg five days per week due to xerosis. Acitretin was ceased in July 2015 with mostly scaling and post-inflammatory hyperpigmentation remaining. The patient applies an emollient containing liquor picis carbonis and salicyclic acid to residual scaly areas. He has not had clinical depression but further monitoring will be undertaken as he is increasingly worried about the risk of his PRP developing in the groin areas.

Discussion

Pityriasis rubra pilaris, an idiopathic uncommon dermatosis, can be hereditary or acquired. It affects both genders equally. Clinically it is characterised by follicular and palmoplantar hyperkeratosis, orange-red scaling plaques and islands of sparing in most active cases. However, it would also be useful to note that islands of sparing can be seen in cutaneous T cell lymphoma and psoriasis. The clinical course of PRP is variable, as demonstrated by the above cases. In adults it typically starts on the scalp and face and spread in a caudal direction. The plaques may become widespread over the entire body and are often pruritic. Histopathology shows an acanthotic epidermis with alternating orthokeratosis and parakeratosis in both vertical and horizontal directions, hypergranulosis, a thick suprapapillary plate, broad rete ridges and dilated hair follicles with keratotic plugs. However, the diagnosis of PRP is largely made clinically.[1]

Therapeutically, there is no curative solution as such unfortunately. If there are no contraindications, systemic retinoids may be considered. Other agents including methotrexate, phototherapy, azathioprine, cyclosporin, mycophenolate mofetil, fumaric acid and biologics have been used with varying success. Topical treatments are often used in adjunct. Commonly

used topical therapies include steroids, keratolytics, emollients, calcipotriol and tretinoin. Overall, however, treatment can be very challenging as PRP can be resistant to both topical and systemic therapies.[1]

On a more positive note, PRP may resolve spontaneously in up to 80% of patients within 1-3 years. By contrast, there are cases which last as long as 20 years. Experience indicates that individual prognosis is difficult to predict. Given the uncertain course of the disease and the deleterious effects of the disease PRP can markedly impair the quality of life of affected patients.[2]

Depression has been previously reported in PRP patients. In a series of 10 patients at a single center in Germany, 2 patients developed symptoms of depression including suicidal thoughts and required psychiatric care.[2] A questionnaire of 123 PRP patients in the United States reported a mean baseline DLQI score of 8.6.[3] PRP patients are burdened by disfigurement from the disease, pain, itching, malodour, fatigue, heat loss from exfoliative disease and time required for doctor appointments and treatments. Some patients experience hearing loss due to keratin build-up in the ear canals or are unable to use their hands or walk normally due to hyperkeratosis. These factors influence clothing choices, ability to participate in activities daily life activities and employment. There are often emotional and financial stresses for not only PRP sufferers but their families as well.[4-6]

Amongst our patients in this series, four of our five patients had mood changes related to their PRP, and one required psychiatric review and antidepressants. The patient requiring antidepressants had the longest duration of disease, appeared most refractory to treatment, had the highest number of hospitalisations and experienced complications from therapy. He attributed his depression to the appearance and malodour of his skin, continual itch, the unpredictable and relapsing course of PRP, and multiple hospital admissions.

It is possible that depression and stress can precipitate PRP through psychosomatic mechanisms. Stress can trigger the appearance of, or exacerbate, dermatological conditions including psoriasis[7] and vitiligo.[8] Stress has also been shown to worsen the intensity of itching in patients with atopic dermatitis.[9] Whilst our patients' depression can be attributed to their PRP, the converse may also be true. Perhaps patients who are predisposed to depression are more likely to develop PRP? There may be a genetic component which increases the risk of both depression and PRP. Time will tell.

When caring for patients with PRP it is important to keep in mind concurrent psychiatric disorders such as depression. Patients may feel more comfortable revealing how their disease impacts on quality of life if their dermatological reviews are not rushed and if reassuring eye-contact is maintained. Moreover, the oft forgotten human touch through palpating the skin may reduce feeling of alienation or unattractiveness. Even online patient support groups may be beneficial. To achieve the best outcome for our PRP patients, collaboration with psychiatry or clinical psychology may be required.[4,5,10]

Conclusion

PRP is an uncommon dermatosis with a variable clinical course. Patients often report mood disturbances due to their appearance, odour, itching and uncertain disease course. It is important to assess the impact of PRP on mood and quality of life during clinical reviews. Further research may be required to determine the prevalence of depression in PRP sufferers and to analyse the not insignificant damage on the patients' quality of life. Helping a patient with PRP can indeed be life saving!

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