

## Photosensitive pityriasis rubra pilaris

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Pityriasis rubra pilaris (PRP) is a rare, chronic papulosquamous skin disorder. The skin lesions rarely have a predilection to sun-exposed areas although it is known that natural sunlight and phototherapy can exacerbate PRP [1–4]. We present a case of a patient suffering from pityriasis rubra pilaris with photoexposed distribution and photosensitivity exclusively to UVB objectively detected by phototesting, successfully treated with methotrexate.

A 43-year-old man of Polish origin with no concomitant diseases was referred to our Department with a 3-week history of skin lesions with a predilection to sun-exposed areas. Medical history was unremarkable. The skin lesions occurred after excessive sun exposure. The patient was initially treated in the Outpatient Clinic with methylprednisolone in a dose of 32 mg/day and topical glucocorticoids with no clinical effect. On admission to the Clinic the patient presented extensive, confluent erythrosquamous and annular skin lesions, mostly on the skin of upper limbs, shoulders, trunk, upper back, neck and face. The peripheral blood laboratory tests showed no abnormalities. HCV antibodies, HBs antigen, HIV Ag/Ab and anti-nuclear antibodies tests were negative. Because of the unspecific clinical presentation clinically suggesting subacute cutaneous lupus erythematosus (SCLE) we performed phototesting with UVA and UVB on the perilesional skin. Phototesting showed the initial minimal erythema dose (MED) at 0.07 J/cm<sup>2</sup> after 24 h and after 48 h in all UVB test fields photoreproduction was observed (Figure 1). After 3 days of hospitalization we observed follicular papules on thighs and abdomen, islands of normal skin and orange, hyperkeratotic plaques with extensive desquamation on feet and hands. The clinical presentation was consistent with classical adult onset PRP (type 1) (Figures 2 A, B). The lesional histopathological examination showed perifollicu-

lar parakeratosis and acanthosis (Figure 3). The acitretin therapy in a dose of 25 mg twice a day was initiated with progressive improvement, but after a gradual dosage reduction to 20 mg/day the skin lesions started to exacerbate. Moreover, the patient suffered from depressed



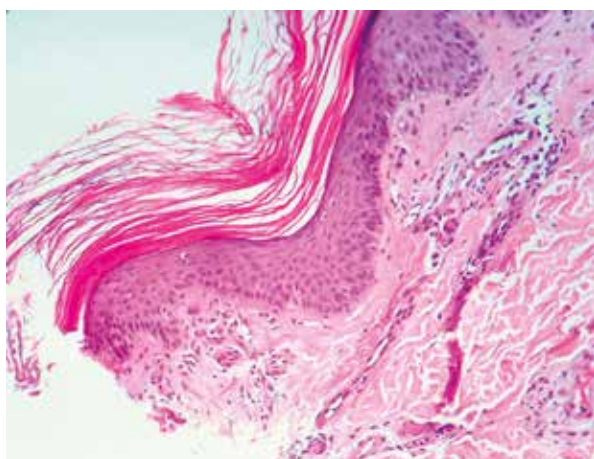
**Figure 1.** Phototesting after 48 h: photoreproduction in all UVB test fields

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**Figure 2.** A – Confluent erythrosquamous, annular skin lesions, follicular papules and islands of normal skin on the back, B – orange, hyperkeratotic plaques with extensive desquamation of palms

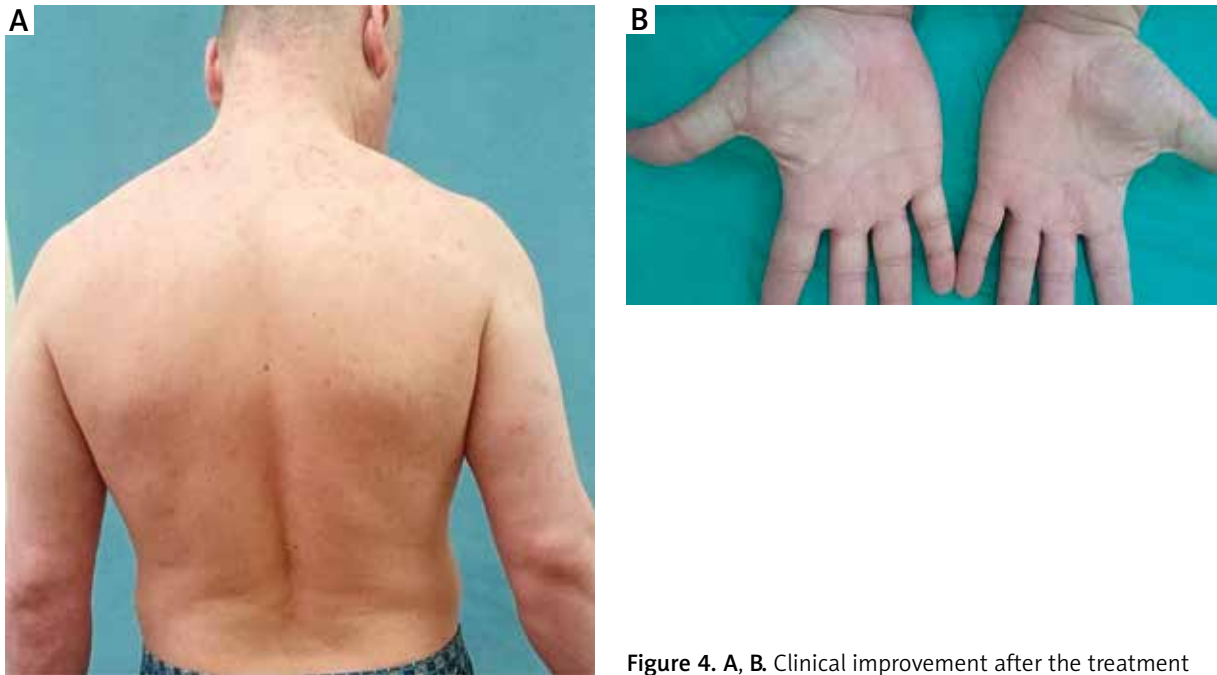


**Figure 3.** Horny plugs blocking the hair follicles, perifollicular parakeratosis and acanthosis with wide pouches of epidermis and dermal papillae

mood, severe skin and oral mucosal dryness, increased tendency to skin injuries and muscle and joints pain during the treatment. As recommended by the psychiatrist, the acitretin therapy was discontinued and the treatment with methotrexate in a dose of 15 mg (*p.o.*) per week with folic acid supplementation was initiated. The treatment did not lead to satisfactory effects after 6 weeks and subcutaneous injections of methotrexate in a dose of 20 mg/week were initiated with good clinical results (Figure 4). The treatment was conducted in combination with topical glucocorticoids, cooling ointments and emollients. We believe that the patient’s emotional problems and mental condition could contribute to initial unsuccessful

treatment. The patient eventually attended the psychotherapy.

Pityriasis rubra pilaris is a chronic disease, of which diagnosis and treatment can be difficult even for experienced dermatologists. Often it takes several weeks or months from the occurrence of first symptoms to determination of proper diagnosis. Pityriasis rubra pilaris is a heterogeneous dermatosis, characterized by erythrosquamous lesions with areas of normal skin, follicular papules and salmon-colored lesions with hyperkeratosis located on the palms and soles. The disease is divided according to clinical symptoms, age and prognosis into six types [5]. The most common type I PRP was diagnosed in the described patient. The etiopathogenesis of PRP remains unknown, although the trigger factors may include severe trauma, stress or drug reactions [6]. Our case is a principal example that the disease may be triggered by overexposure to ultraviolet radiation. So far only few cases of patients with photoexposed distribution were described and in most cases photosensitivity was exclusively to UVB [1–4]. Phototesting is rarely performed in PRP and phototherapy is debatable. Patients often report side effects associated with inflammation, irritation and itching of the skin. However, there have been cases of patients treated with UVB radiation with good effects in addition to a greater immunosuppressive effect of narrow-band UVB compared with PUVA [7, 8]. Retinoids have been proved to be the first choice therapy of PRP [9] and methotrexate is considered to be the drug of second choice [10], both in similar doses as in psoriasis. In our case we initially used acitretin. Because of the side effects and unsatisfactory clinical response, we decided to change the treatment to oral methotrexate at an initial dose of 15 mg weekly. The dose was eventu-



**Figure 4. A, B.** Clinical improvement after the treatment

ally increased to 20 mg weekly in subcutaneous injections. If methotrexate does not lead to improvement, the treatment with fumaric acid can be introduced [11]. Described methods are not always effective. However, biological drugs give great opportunities to patients with PRP. Petrof *et al.* analyzed 15 cases of patients with PRP and obtained complete response in 80% after the administration of anti-TNF- $\alpha$  [12]. One reported case was successfully treated with infliximab after the first dose [13]. Infliximab and methotrexate can be combined to achieve higher levels of infliximab and prevent infliximab antibodies formation [6]. We considered this method of treatment in our patient, but the response to subcutaneous injections of methotrexate alone was satisfying. The beneficial effects of ustekinumab were also reported [14]. Clinicians should not forget to choose the right topical treatment, especially the right emollients. Factors irritating the skin must be avoided. In our patient, we used topical glucocorticoids, cooling ointments and emollients with a good effect and reduction in itching of the skin.

Pityriasis rubra pilaris treatment options are complex and individual as the mental attitude of the patient seems to be crucial. Patients with chronic skin diseases, like PRP are exposed to much higher levels of stress and have reduced quality of life [15]. Treatment with retinoids and methotrexate leads to far worse effects than in the case of psoriasis. New hope for patients is a biological treatment. Phototherapy is still debatable, as both UVA and UVB radiation can exacerbate the disease. As shown in our case, phototesting should always be performed in every patient before initiating phototherapy and an appropriate photoprotection should be recommended as

it turns out, the UV radiation may be a factor triggering and aggravating PRP.

#### Conflict of interest

The authors declare no conflict of interest.

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