

## A 12-Year-Old Girl with a Scaly Papular Rash

KAARE WEISMANN

*The Skin Clinic, Private Hospital Hamlet, Gyngemose Parkvej 66, DK-2860 Søborg, Denmark.  
E-mail: kwe@hamlet.dk*

**The author, Kaare Weismann, reports a case of pityriasis rubra pilaris in a 12-year-old girl. In his experience this disease is best treated with a good emollient and not with any aggressive medication.**



Pityriasis rubra pilaris (PRP) is a heterogeneous group of disorders that have in common circumscribed follicular keratoses, keratoderma, branny scales and a characteristic orange-red erythema with islands of unaffected skin. The aetiology of PRP is unknown. It is seen in adults and, rarely, in prepubertal children (1). The juvenile form is usually misdiagnosed as atopic eczema, psoriasis, lichen spinulosus, or erythrokeratoderma. Different treatment modalities are often tried with a poor or no response (2, 3).

### Case report

A 12-year-old girl was referred to the outpatient's clinic with a widespread exanthema, which started 7–10 days after she had been given a combined vaccine against measles, mumps and rubella. She developed pink scaly areas on the face and neck, followed by multiple follicular papules on the upper

extremities and trunk, with a cephalocaudal spread (Figs 1 and 2). A punch biopsy showed acanthosis, hyper- and parakeratosis, and dilated follicles with characteristic horny plugs (Fig. 3). A diagnosis of PRP was established. Prior to referral she was suspected to have atopic dermatitis and psoriasis and had been treated with various topical corticosteroids, pimecrolimus, tacrolimus, coal-tar and retinoids, oral prednisolone 15 mg daily for 2 weeks, and 20 narrow-band ultraviolet B (UVB) exposures. None of these treatments had had any effect.

In agreement with the patient and her mother, we abstained from further efforts to combat the disease. We recommended an emollient, and told her to return if the rash changed. Three months later she attended the clinic again. All skin changes had disappeared in the meantime, and there has been no recurrence in the following 2.5 years.



Fig. 1. Well-demarcated scaly plaques on the face with small areas of unaffected skin.



Fig. 2. Follicular eruption on the trunk and upper extremities.

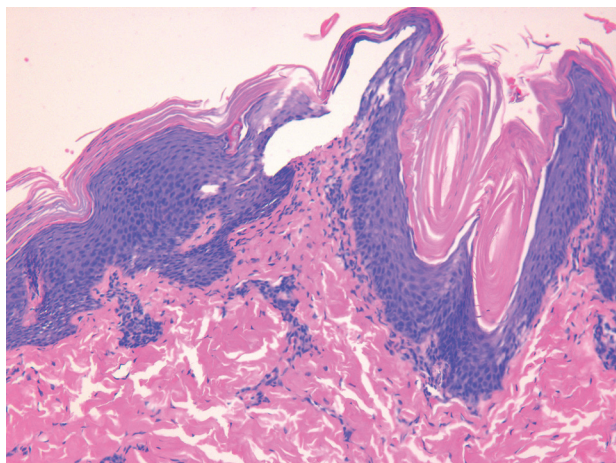


Fig. 3. Biopsy from abdominal skin shows dilated hair follicles with horny plugs. Epidermis is acanthotic with varying hyper- and parakeratosis (haematoxylin and eosin (H&E), original magnification  $\times 100$ ).

## Discussion

Different clinical varieties of PRP have been described (1), but it is often difficult to classify the patients, especially if they are young (2). The typical prepubertal PRP patient has pink or orange-coloured well-demarcated plaques with follicular plugging on the knees and elbows, widespread follicular papules on the trunk and extremities, keratoderma on the palms and soles, and scaly changes on the face with a sharp margin. A typical age is 12 years, as was the case in our patient (4). Juvenile PRP clears spontaneously in more than 50% of the patients within 6 months, and more than 60% within one year. In rare cases it may persist for years (2, 3).

There is no recommended treatment for juvenile PRP. Casuistic reports have shown effect of a wide range of treatments, such as coal-tar, vitamins A and D, ultraviolet B (UVB), psoralen plus

UVA (PUVA), methotrexate, prednisolone, cyclosporine, antibiotics, TNF-alpha inhibitors and systemic retinoids. In most cases the treatment lasted more than 6 months (4), thus spontaneous resolution of the rash might explain the reported success.

The histology of classical PRP is distinctive, but varies with the stage of the disease and may differ from site to site. The presence of dense horny follicular plugs is essential for the diagnosis.

It is speculative whether the vaccination somehow triggered the eruption in this case. It has been suggested that juvenile PRP is mediated by bacterial superantigens (5).

Due to its good prognosis, juvenile PRP should not be treated with aggressive medication, since no treatment seems to work, and there is always a risk of side-effects. A positive attitude and a good emollient is often the best option for the patient.

## Conflict of interest

None declared.

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