Pityriasis Rubra Pilaris

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Key Points

- Pityriasis rubra pilaris (PRP) is a chronic papulosquamous disorder of keratinization. The etiology is unknown. PRP shows similarities with psoriasis, but it is a distinct disease.
- According to Griffiths, there are six types of PRP. It appears in adults but also in a juvenile form. It typically affects extensor parts of the body, starting usually from the face. Typical lesion is hyperkeratotic papule. Spearing islands of the non-affected skin are a pathognomonic sign. After some time, salmon-like discoloration of the skin occurs. Palmoplantar keratoderma is almost always present.
- Histology is not confirmatory.
- PRP should be distinguished from psoriasis and other papulosquamous disorders.
- Treatment is topical or systemic. Retinoids are first-line treatment. Biologics are a new class of available medications for the treatment of PRP, basically used with success in psoriasis.

Definition and Epidemiology

Pityriasis rubra pilaris (PRP) is a chronic, inflammatory, papulosquamous disorder of keratinization with unknown etiology, characterized with affection of the body and palmoplantar ("sandal") affection. It appears both in hereditary and in acquired form. The first description of PRP is described by Claudius Tarral who, in 1835, published a case report in Rayer's *A Theoretical and Practical Treatise on the Diseases of the Skin* of a patient he had seen 7 years earlier. Tarral did not recognize the dermatosis as a distinct entity, and it was listed under the title of "general psoriasis."

It affects both sex male and female equally without sexual predilection. The highest incidence is in the first and sixth decade. Incidence is around 1:5,000. There may be racial variation as the incidence of classical PRP was reported to be closer to 1 in 50,000 in India.

The etiology is unknown. There are some similarities with psoriasis, but it is a distinct disease, which is more of a disorder of keratinization than an inflammatory disease. There are some clinical and histological evidence as well about vitamin A deficiency. Infective etiology and familial distribution are also suggested. PRP could appear also after drug therapy and the influence of some triggering factors, such as severe infection, especially in juvenile form. Simultaneously PRP may appear with other immunological disorders, like rheumatoid

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Table 77.1 Classification of pityriasis rubra pilaris. Griffiths' six types of PRP

Type	% of cases	Distribution	Prognosis
I. Classical adult	55	Generalized	Most clear in 3 years
II. Atypical adult	5	Generalized	Chronic
III. Classical juvenile	10	Generalized	Most clear in 1 year
IV. Circumscribed juvenile	25	Focal	Uncertain
V. Atypical juvenile	5	Generalized	Chronic

From Albert and Mackool (1999) modified

arthritis, and some malignancies like leukemia or Sezary syndrome, which indicates association with immunosuppression.

6. HIV related. Symmetrical, pruritic eruption composed of erythematous papules is associated with late-onset acne conglobata.

Classification

According to Griffiths there are six types of PRP (Table 77.1) and the classical adult type is the most common, and it has the best prognosis. Remission is appearing after several years. Type 2 PRP is the atypical adult form with a tendency to become chronic. The type 3 classical inherited variant seems to have also good prognosis. Type 6 is described recently in which PRP is associated with HIV and this type has a poor prognosis.

1. Classical adult. Commonest type (50 % of cases). Spreads caudally. The patient is usually erythrodermic with diffuse thickening of the palms and soles. Ectropion often is present.
2. Atypical adult. Long duration. Involves a more ichthyosiform pattern in association with hair loss and areas of eczematous changes.
3. Classical juvenile. Similar to type 1 but affects children in the first decade.
4. Circumscribed juvenile. Affects children, with sharply demarcated areas of follicular hyperkeratosis and erythema on the knees and elbows. Usually does not progress.
5. Atypical juvenile. Appears in the first few years of life and is chronic. Characterized by follicular hyperkeratosis, whereas erythema is not a prominent feature. The skin on the hands and feet can appear scleroderma-like.

Clinical Presentation

Clinical picture is the basis for establishing the diagnosis of PRP.

Classical disease begins on the face, with scaling and erythema, and on lateral parts of the neck and trunk. Also, the disease may appear on extensor surface of the extremities and especially on the back of the first and second phalange. PRP is starting with small plaque lesions which spread all over the body. Typical lesion is follicular hyperkeratotic papule. Spearing "islands" of not affected skin are a pathognomonic sign. In 20 % of the patients, mild pruritus and burning sensation are present. Also Koebner phenomenon may appear like in psoriasis. After some time orange-red "salmon-like" discoloration of the skin is present. This is the hallmark for PRP.

Palms and soles are almost always involved, often showing an orange color. Palmoplantar waxy keratoderma appears after some period, with typical side spreading, producing the so-called sandal pattern. Changes of the nails, thickened nails with a distal discoloration, and subungual hyperkeratosis are often present, but pitting of the nail and dystrophy like in psoriasis do not exist.

Erythroderma can occur after 2–3 months. Erythroderma is a very serious development of the disease and should be monitored carefully. However, usually PRP remains static and disappears after 12 months. Mild pruritus is present.

Histology

Histopathological examination is showing hyperkeratosis, acanthosis, and parakeratosis, both horizontal and vertical, and superficial perivascular lymphocytic infiltrate. There is prominent follicular plugging with dense keratotic material. Histopathological findings are not pathognomonic; they can exclude other papulosquamous diseases.

Differential Diagnosis

PRP has to be distinguished from psoriasis (which is sometimes very difficult) and other papulosquamous disorders in adults and atopic dermatitis in childhood. Other reasons of erythroderma should be excluded. Cutaneous T-cell lymphoma and drug skin reactions should be taken in consideration.

There is no diagnostic laboratory test to confirm PRP. Distinguishing clinical features of PRP include islands of normal, non-affected skin, follicular keratotic plugs, and orange-red discoloration of affected skin.

General Principles of Treatment

Because of the variable clinical course and low incidence, assessment of the treatment modalities is difficult. Treatment modalities should include topical application of medications and systemic therapy.

Topical Treatments

Emollients are effective treatments for pityriasis rubra pilaris. Conservative approach is advised in juvenile cases. Keratolytics, like salicylic acid and tar, are important in the treatment. Calcipotriol can also be effective in children. Urea is very helpful. Topical corticosteroids are not very effective. All irritating factors should be avoided. Palmoplantar keratoderma is difficult to treat. Keratolytics and corticosteroids under occlusion are advised. Calcipotriene should be considered. Tazarotene is also available in the treatment of PRP.

Systemic Treatments

Oral synthetic retinoids are currently the treatment of choice for PRP and have largely replaced vitamin A therapy. Oral retinoids are the first choice in the treatment of widespread disease. Acitretin 0.5–1 mg/kg (in adults) or isotretinoin 0.5–2 mg/kg is advised. Duration of treatment is from 3 to 6 months. The main disadvantage of retinoids is teratogenesis in females. Appropriate contraceptive measures are mandatory and pregnancy is absolutely contraindicated during treatment with acitretin and for 2–3 years after discontinuation of acitretin and during treatment with isotretinoin and for 1 month after discontinuation of isotretinoin. So, oral acitretin is not indicated for the treatment of women of reproductive potential.

Methotrexate is an alternative agent for refractory PRP. Methotrexate is a second line of treatment given orally 15–25 mg/weekly.

Use of cyclosporine in the treatment of pityriasis rubra pilaris is still controversial.

UV-light treatment uses the ultraviolet light spectrum of 290–400 nm to treat a variety of skin diseases. It can be used alone or in combination with other medications applied directly to the skin or taken orally. PUVA, Re-PUVA, is also applied in PRP with less success than in psoriasis vulgaris. UVB therapy used in psoriasis vulgaris could have negative influence and aggravate pityriasis rubra pilaris.

Oral antihistamines can be successful to decrease itching.

Oral megadoses of vitamin seem to be not so effective.

Biologics are new classes of medications that target immune system responses. The beneficial use of biologics in psoriasis is evidence based and tumor necrosis factor alpha (TNF-α) has a central role in psoriasis. There are some data emphasizing that blockade of tumor necrosis factor alpha with antagonists of TNF-α also could

have positive effects in PRP. They are administered by subcutaneous injection (adalimumab, ustekinumab, etanercept) or by intravenous infusion (infliximab).

In case of erythroderma, intensive supportive therapy is imperative.

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