***no patient handout*

Pityriasis rubra pilaris - Skin

Contributors: Belinda Tan MD, PhD, Noah Craft MD, PhD, Lindy P. Fox MD, Lowell A. Goldsmith MD, MPH, Michael D. Tharp MD

Synopsis

This summary discusses pityriasis rubra pilaris in adults. **Pityriasis rubra pilaris in children** is addressed separately.

Pityriasis rubra pilaris (PRP) is characterized by an acute cutaneous eruption that is often accompanied by pruritus and/or pain. Classic cutaneous lesions include follicular papules on an erythematous base coalescing to form large orange-red plaques but with characteristic islands of sparing. PRP commonly begins on the scalp and rapidly spreads in a craniocaudal direction and has the potential to quickly progress to erythroderma over several weeks' time. Additional features include an orange-red palmoplantar keratoderma and sparing of the mucous membranes. PRP can be classified into 5 clinical types based on age of onset, cutaneous features, and clinical course. In classic adult PRP, more than 80% of patients will experience spontaneous remission within 3 years.

The etiology of PRP has not been clearly defined. The onset of disease has been associated with myositis, myasthenia gravis, hypothyroidism, HIV, infection, and malignancy. In addition, UV exposure and minor skin trauma prior to the onset of PRP have been reported. While there are reports of heritable forms, the large majority of PRP cases are acquired and without sex predilection. The incidence of the acquired form occurs in 2 peaks: during the first and second decades and the sixth decade.

For more information, see **OMIM**.

Codes

ICD10CM:
L44.0 – Pityriasis rubra pilaris

SNOMEDCT:
3755001 – Pityriasis rubra pilaris

Look For

Discrete, follicular, scaling papules coalescing in areas to confluent, orange-red plaques. These often begin on the scalp and may expand to involve most of the body. There is orange hyperkeratosis of the palms and soles, sometimes with painful fissuring.

Nail changes include a thickened yellow-brown nail plate with subungual debris.

Oral mucosal changes are very rare, but when present can include gray or whitish papules or plaques, erythema, or erosions.

Diagnostic Pearls

Several features facilitate the differentiation of PRP from psoriasis, and they include the following:

- Islands of normal skin within larger plaques are characteristically seen in PRP
- Orange-red, waxy-like keratoderma of the palms and soles are also features seen in PRP
- Nail oil-drop changes and nail pitting are characteristic of psoriasis
- A family history of psoriasis is often seen in psoriatic patients
- PRP and psoriasis are histologically different, and a biopsy will aid in the diagnosis

Differential Diagnosis & Pitfalls

Skin biopsy will greatly aid in the diagnosis.

- **Psoriasis** – See Diagnostic Pearls.
- **Seborrheic dermatitis** – Much more responsive to standard therapies. PRP is often resistant to conventional therapies for seborrheic dermatitis. In addition, the progression of body surface involvement will distinguish PRP from seborrheic dermatitis.
- **Mycosis fungoides** / Sezary syndrome – Generalized lymphadenopathy, circulating malignant lymphocytes as determined by flow cytometry, leonine facies, a CD4/CD8 ratio greater than 10 as determined by flow cytometry.
- Erythrokeratoderma variabilis – More than 90% of patients present within the first year of life.
- **Drug eruption** – Often present with urticarial, **exanthematous**, or vesicular / **bullous** lesions. In addition, systemic symptoms are more pronounced, including fever, lymphadenopathy, and facial edema. Eosinophilia on CBC and histology are often seen (but not an invariable finding). Medication history will help.
- **Atopic dermatitis** – Patients are often aware of their atopic history, and the condition commonly starts in childhood. Look for lichenified plaques on the flexural surfaces and neck.
- **Erythrokeratodermia variabilis**

Best Tests

Skin biopsy will usually aid in confirming the diagnosis and differentiating PRP from psoriasis.

Histopathology Findings:

- Follicular plugging
- "Shoulder parakeratosis" adjacent to follicular plugs or "checkerboard parakeratosis" alternating with orthokeratosis
- Irregular acanthosis, may be psoriasiform
- Focal acantholysis especially at periphery
- Perivascular lymphocytes, may be lichenoid

Other laboratory and radiologic investigations are not routinely necessary. Depending on the clinical situation, a search for HIV infection or underlying malignancy may be indicated.

Patients who develop erythroderma should be evaluated for electrolyte abnormalities, hypoalbuminemia, and secondary infection.

Management Pearls

Some immediate relief can be gained from the application of potent topical steroids under occlusion, alternating with a solution of propylene glycol and lactic acid under occlusion. However, these modalities have little long-term therapeutic benefits. Heavy emollients, such as petroleum jelly or Aquaphor, may relieve fissuring and help erythrodermic skin retain moisture. Emollients such as 12% lactic acid cream or lotion (Lac-Hydrin 12%) may be helpful on areas of keratoderma.

The morbidity of PRP is due to the erythroderma. Measures should be taken to prevent the progression of PRP to erythroderma. Complications of erythroderma include volume shifts (ie, lower extremity edema) secondary to loss of fluids and proteins, tachycardia-induced high output cardiac failure, and thermoregulatory dysregulation (hypo- and hyperthermia).

Therapy

No large controlled trials have been performed. Prednisone 40-60 mg every 24 hours is effective in controlling the disease and can be tapered while instituting one of the following:

- Retinoids (isotretinoin, acitretin) are effective but must be used cautiously in women of child-bearing age due to their teratogenic effects. Physicians prescribing these medications must closely supervise the therapy, including monthly urine HCG, CBC, triglyceride level, liver function tests, and 2 forms of contraception in women, or
- Acitretin (10, 25 mg) – Begin at 25 mg every 24 hours, advance to 50 mg and possibly 75 mg every 24 hours depending on weight, or
- Isotretinoin (20, 40 mg) 0.5-1.5 mg/kg every 24 hours with food, or

- Methotrexate (10-30 mg weekly) – Well-known side effects of methotrexate include hepatoxicity, myelosuppression, and conversion from latent to active tuberculosis or histoplasmosis. Take with folic acid 1 mg every 24 hours or leucovorin 5 mg weekly. The FDA approved glucarpidase to treat toxic levels of methotrexate caused by impaired kidney function in 2012, or
- Azathioprine 1 mg/kg every 24 hours for 6-8 weeks, and increase by 0.5 mg/kg every 2-4 weeks; do not to exceed 2.5 mg/kg every 24 hourse, or
- Cyclosporin A 2.5 mg/kg every 12 hours for 4 weeks. Increase by 0.5 mg/kg every 24 hours every 2-4 weeks. Do not to exceed 5 mg/kg every 24 hours. Infliximab for refractory cases. Administer 5 mg/kg IV infusion followed by 5 mg/kg at 2 and 6 weeks after the first infusion, then subsequently every 2 months, or
- Phototherapy can exacerbate PRP or induce remission. Success has been reported using treatment with narrow-band UVB (TL-01) in combination with acitretin. Extracorporeal photochemotherapy has also demonstrated success.

Drug Reaction Data

Below is a list of drugs with literature evidence indicating an adverse association with this diagnosis. The list is continually updated through ongoing research and new medication approvals. Click on Citations to sort by number of citations or click on Medication to sort the medications alphabetically.

Medication	Citations
Beta blockers	1
imiquimod	1
labetalol	1
protease inhibitors	1
simvastatin	1
statin	1

Medication	Citations
telaprevir	1