Pityriasis rubra pilaris is a skin condition with many different clinical presentations. History, histology, clinical presentation, its different classified forms, treatments, and differential diagnoses are reviewed.

Pityriasis rubra pilaris (PRP) is a red scaly eruption characterized by the association of palmoplantar keratoderma, follicular plugging, and erythematous peri-follicular papules which may progress to plaques or erythroderma (White, 2003). Most cases are sporadic and acquired but a familial form may exist which is transmitted by an autosomal dominant or autosomal recessive mechanism.

This disease was first described by Devergie in 1856 who characterized PRP as a follicular eruption and noted its association with three other cutaneous disorders: psoriasis palmaris, pityriasis capitis, and pityriasis rubra. In 1877, Richaud recognized that all of the manifestations were of the same disorder. In 1889, Besner added the epithet rubra (White, 2003).

Onset
PRP exhibits a bimodal distribution of age of onset. It first peaks in early childhood and the second peak comes after the 5th decade. Onset in children frequently follows trauma or infection such as streptococcal. There is spontaneous resolution of PRP in at least 75% of cases within 3 months to 7 years of onset. Relapses are uncommon, but may occur. Familial PRP does not resolve spontaneously, although many cases that have been so classified may represent other ichthyosiform disorders. PRP affects both male and females equally, but it is uncommon in the general population (Arnold & Buechner, 2004). PRP is a rare disease, but one study reports a frequency of about 1 case in every 500 new pediatric patients with a dermatologic disease (Vijayalakshmi & Mallika, 2003).

The causes of PRP remain unknown. The epidermis is in a hyperkinetic state, with a rapid turnover rate of the follicular keratinocytes, and may approach the rate found in psoriasis. There is a resemblance of the histology to that found in phrynoderma, a distinctive form of follicular hyperkeratosis associated with nutritional deficiency with characteristic hyperkeratotic papules that first appear on the extensor surfaces of the extremities, shoulders, buttocks. The finding of clinical follicular occlusion suggests that a deficiency or malfunction of vitamin A might be involved (White, 2003). A decrease in retinol binding protein is also reported. Retinol binding protein is the specific carrier of vitamin A and its decreased synthesis is a biochemical marker for PRP. Although decreased retinol binding protein is a marker for PRP, it is not proven that decreased vitamin A is a cause. It is more likely due to the decreased serum level of the carrier of vitamin A (Evangelou, Murdoch, Palamaras, & Rhodes, 2005).

Clinical Findings
The initial clinical findings of PRP may be redness and scaling of the face and scalp. Skin on the palms and soles may become quite thickened, painful, and orange in color. The patient may experience debilitating hyperkeratosis. The
The skin becomes progressively redder, and painful fissuring of the palms and soles may occur. While an exfoliative erythroderma may ensue, characteristic “skip” areas of non-involvement are usually seen. Rarely patients may complain of fever, chills, and malaise (Arnold & Buechner, 2004). The most common affected sites are the palmo-plantar surfaces, elbows, and knees (75%-80%); dorsa of the hands and feet (60%); and the face (40%) (Vijayalakshmi & Mallika, 2003).

The most characteristic clinical feature is a follicular verrucous papule about 1 mm in diameter with a central keratotic plug. A yellow-orange ring surrounds the lesion (Vijayalakshmi & Mallika, 2003). The second most important diagnostic element is a palmoplantar keratoderma, which is salmon colored and edematous. The Achilles tendon area is frequently involved. As stated previously, there is a cephalic rash. The hair and teeth are not involved but occasionally psoriasiform nail changes may occur. White lacy plaques may be observed on the buccal mucosa (Vijayalakshmi & Mallika, 2003).

The histological changes seen in PRP are consistent but not specific. There is acanthosis with blunting of rete pegs and suprabasal perinuclear vacuolation in isolated cells. Liquefactive degeneration of the basal layer is most likely to be seen overlying the dermal papillae. The granular cell layer is attenuated in some areas and thickened in others. Mild superficial vasodilatation of capillaries with a slight to moderate lymphohistiocytic perivascular infiltrate is found (White, 2003). Examination of a follicular papule is most helpful in the diagnosis. Parakeratosis of the follicular ostea associated with follicular hyperkeratosis is seen. Hypertrophy of the erector pili is a helpful diagnostic feature when present. A dermal perivascular mononuclear cell infiltrate is present and there are occasional mononuclear cells (see Figures 1 & 2) (Arnold & Buechner, 2004).

The presence of PRP has occasionally been associated with immunodeficient states, such as HIV. In the juvenile forms of this disease, PRP may be associated with T-helper cell impairment, hypogammaglobulinemia, and decreased IgA. Underlying diseases are unusual, but PRP may also be associated with HIV, hypothyroidism, myasthenia gravis, celiac disease, and acute stem cell leukemia.

Patients with pityriasis rubra pilaris can be classified into five types, according to Griffith’s Classification (Griffiths, 1975), which differ from each other on the basis of clinical features, age of onset, and prognosis (see Table 1).

**Type I**

Classical adult onset PRP accounts for over half of the patients. The disease starts out with a single erythematosus patch on the upper half of the body and within a few weeks or months, there are large zones of follicular hyperkeratosis each with an erythematous perifollicular halo. Islands of unaffected skin, one to several centimeters in diameter remain scattered over the sheet of erythema. The scaling is fine and powdery. On the lower body the scaling is coarser (see Figures 3, 4, & 5) (Griffiths, 1975).

On the face and scalp, oral vitamin A (150,000 to 300,000 IU daily) has minimal improvement among patients (White, 2003). Vitamin A storage in the liver has the potential side effects of teratogenicity and elevated triglycerides (Schachner & Hansen, 2003). The retinoids, isotretin, etretinate, and acitretin appear to be more effective than vitamin A with over 80% successful clearance in 3 years (Schachner & Hansen, 2003). It seems that early

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**Figure 1.** There is psoriasiform hyperplasia of the epidermis with alternating areas of orthokeratosis and parakeratosis and plugs of the follicular infundibulum.

**Figure 2.** A sparse, superficial, perivascular lymphocytic infiltrate is also noted.
Diagnosis and early treatment with retinoids leads to the best chance for clearing. If there is a lack of response or contraindications to the use of retinoids, methotrexate could be used (Schachner & Hansen, 2003).

Type II

Atypical adult onset affects about 5% of patients. This disease is atypical because of its long duration of 20 years or more and atypical morphologic features. The scaling is more ichthyosiform, especially on the legs. Sparseness of the scalp hair is sometimes present. Palmpoplantar hyperkeratosis is coarse and lamellated (Griffiths, 1975). Patients must be treated on a case by case basis.

Type III

Classical juvenile PRP affects 10% of patients. It resembles type I in its mode of onset and appearance, but it affects children in the first year of 2 or life (see Figures 6-7) (Griffiths, 1975). Sixty percent of cases self-limit in 1 year, while 90% resolve in 3 years. Topical treatments with keratolytics, mild topical steroids, and emollients are preferred over systemic treatments in pediatric patients with PRP (Schachner & Hansen, 2003).

Type IV

Circumscribed PRP affects approximately 25% of patients. It affects prepubertal children and it is characterized by sharply demarcated areas of follicular hyperkeratosis and erythema on the knees and elbows. Hyperkeratosis may be found overlying bony prominences. Circumscribed PRP shows no tendency towards progression (Griffiths, 1975). Systemic retinoid acid works quite well with type IV, especially when alternated with a topical steroid (Schachner & Hansen, 2004).
of PRP have the capability to transform into another form of the disease. In one study in particular, a patient presented with type III but then later went on to develop type IV. This may suggest a common etiological mechanism for all types of PRP (Gelmetti, Schiuma, Cerri, & Gianotti, 1986; Shahidullah & Aldridge, 1994).

The classic adult type I has the best prognosis with 80% clearing within 3 years. Most children with PRP may be treated with topical therapy alone. Some forms of topical therapy include emollients, calcipotriene, retinoic acid, keratolytics, topical steroids, and hydrating agents. Systemic therapies include retinoids (acitretin, etretinate, isotretinoin), methotrexate, cyclosporin, and psoralen ultraviolet A (PUVA). Other agents being used, but none that can be recommended, include vitamins C and E, statinolozol, arsenic, and pilocarpine (Cohen & Prystowsky, 1989; Finzi, Altomare, Bergamaschini, & Tucci, 1981).

Two other classifications systems are also used for PRP. Gelmetti’s Classification (1986) is based on the duration of the disease. Piamphogsant’s Classification (1994) is based on physical findings and it is divided into four types.

Conclusion

Pityriasis rubra pilaris is an important disease to be aware of and understand. The differential diagnosis includes psoriasis, seborrheic dermatitis, phrynoderma, atypical keratosis pilaris, follicular eczema, and erythrokeratoderma (for example, Unna-Thost disease and Papillon-Lefevre syndrome). PRP appears in different clinical presentations but there are treatments available for all of them. When treating a patient with PRP, it is important to identify which type is presenting and treat accordingly. Fortunately, the majority of patients presenting with PRP have a favorable prognosis.

References


