Pityriasis rubra pilaris
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CLINICAL FINDINGS

- Systemic
- Topical

SECOND LINE

- Cyclosporine A (5 mg/kg/day)
- Narrowband ultraviolet B phototherapy
- Photochemotherapy (topical or systemic psoralen plus ultraviolet A)
- Vitamin A analogues (tazarotene)
- Vitamin D

FIRST LINE

- Keratolytics (salicylic acid, urea)

PROGNOSIS AND CLINICAL COURSE

- Recurrences are recognized in up to 20 percent of patients, however, sometimes after long periods of subclinical disease.
- In the classic juvenile variant (type III), spontaneous clearing is occasionally fatal, with death occurring due to complications of cutaneous sepsis.
- The clinical features of this variant are similar to those of type I but with increased severity and additional manifestations of acne conglobata, hidradenitis suppurativa, and lichen spinulosus.

Classifications for Pityriasis Rubra Pilaris (Types I-VI According to Griffiths and González-López)

- Type I:
  - Generalized, beginning on the head and neck, then spreading caudally
  - > 50 years
  - Chronic course, improves with retinoids but relapses when they are stopped

- Type II:
  - Generalized, beginning on the head and neck, then spreading caudally
  - > 50 years
  - Long duration (> 20 y)
  - Combines of follicular hyperkeratosis and ichthyosiform lesions on the legs, sparse scalp hair

- Type III:
  - Localized
  - Generalized, beginning on the head and neck, then spreading caudally
  - > 50 years
  - Long duration (> 20 y)
  - Combines of follicular hyperkeratosis and ichthyosiform lesions on the legs, sparse scalp hair

- Type IV:
  - Generalized
  - Long duration (> 20 y)
  - Chronic course, improves with retinoids but relapses when they are stopped

- Type V:
  - Generalized
  - Long duration (> 20 y)
  - Chronic course, improves with retinoids but relapses when they are stopped

- Type VI:
  - Generalized
  - Long duration (> 20 y)
  - Chronic course, improves with retinoids but relapses when they are stopped

The clinical features of this variant are similar to those of type I but with increased severity and additional manifestations of acne conglobata, hidradenitis suppurativa, and lichen spinulosus.

Consider

- Cutaneous T-cell lymphoma
- Human immunodeficiency virus infection
- Pityriasis lichenoides chronic
- Keratosis pilaris
- Lichen ruber acuminatus
- Psoriasis
- Psoriasis

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Although an underlying dysfunction in vitamin A metabolism has been suggested as a cause, vitamin A deprivation have been unsuccessful. Moreover, a deficiency of retinol-binding protein has been noted in patients with dermatomyositis, often associated with internal neoplasia. Concomitant conditions such as follicular ichthyosis and the erythrokeratodermas.

Pathologic findings in pityriasis rubra pilaris vary according to the duration of the disease. The etiology and pathogenesis of pityriasis rubra pilaris are poorly understood. The role of vitamin A metabolism has been suggested as a cause, but episode appeared to trigger the onset of the disease.

Although an underlying dysfunction in vitamin A metabolism has been suggested as a cause, vitamin A deprivation have been unsuccessful. Moreover, a deficiency of retinol-binding protein has been noted in patients with dermatomyositis, often associated with internal neoplasia. Concomitant conditions such as follicular ichthyosis and the erythrokeratodermas.