Treatments for Pityriasis Rubra Pilaris

Therapeutic intervention and may allow prevention of protracted illness and serious complications. Suicide remains a risk in patients with generalized disease.

PREVENTION

Emollients and antihistamines provide significant benefit. Topical therapy with calcipotriol is effective and may be used alone or in combination with other therapies.

When conventional treatment strategies fail, new therapeutic approaches may include the use of narrowband ultraviolet B phototherapy or methotrexate. In patients with severe symptoms, effective treatment with retinoids or methotrexate may suffice. In cases where retinoids are ineffective, ultraviolet A1 phototherapy may be a satisfactory alternative.

Concurrent therapeutic approaches include the use of cyclosporine, fumaric acid esters, or tumor necrosis factor-α antagonists. Systemic retinoids, usually acitretin, are effective in clearing lesions. Topical therapies such as vitamin A analogues (tazarotene) or keratolytics (salicylic acid, urea) may also be used.

Considerations for treatment:
- First line: Oral retinoids (acitretin) are the current treatment of choice.
- Alternatives: Methotrexate or cyclosporine may be used as second-line therapies.
- Additional options include ultraviolet A1 phototherapy or tumor necrosis factor-α antagonists.

Always Rule Out

- Differential Diagnosis of Pityriasis Rubra Pilaris
  - Other skin conditions such as psoriasis, follicular ichthyosis, keratosis pilaris, and lichen ruber acuminatus.
  - Cutaneous T-cell lymphoma.
  - Systemic disorders such as systemic lupus erythematosus (SLE), chronic renal failure, and human immunodeficiency virus (HIV) infection.

Differential Diagnosis

- Type I (classic adult disease): Erythroderma with islands of normal skin (“nappes claires”), follicular hyperkeratosis, waxy diffuse palmoplantar keratoderma, and hyperkeratosis with only minimal erythema and a scleroderma-like appearance of the hands and feet.
- Type II (atypical adult disease): Often resolves within an average of 1-2 years.
- Type III (classic juvenile variant): Similar to type I but appears in year 1 or 2 of life and often resolves within an average of 3 years.
- Type IV (atypical variant): The atypical variants (type II and IV) have a less favorable prognosis for remission, although some cases of type IV improve in the late teens.
- Type V (erythrodermic variant): Often resolves within an average of 1-2 years.
- Type VI (HIV infection-associated type VI): Commonly observed in 1 to 2 years. The atypical variants (type II and IV) have a less favorable prognosis for remission, although some cases of type IV improve in the late teens.
- Type VII (circumscribed juvenile type): Often resolves within an average of 1-2 years.
- Type VIII (generalized type): Rare, with an average duration of less than 10 years.

Pathology

Pathologic findings in pityriasis rubra pilaris vary according to the duration of the disease. The features of type I are characterized by a hyperkeratotic plaques and erosions. Hair and teeth are normal. Type II may have a diffuse, yellowish keratoderma of the palms and soles. Type III is characterized by a reddish orange, scaling plaque on the scalp and face, and a diffuse, yellowish keratoderma of the palms and soles. Type IV may have a diffuse, yellowish keratoderma of the palms and soles. Type V is characterized by a diffuse, yellowish keratoderma of the palms and soles. Type VI is characterized by a diffuse, yellowish keratoderma of the palms and soles. Type VII is characterized by a diffuse, yellowish keratoderma of the palms and soles. Type VIII is characterized by a diffuse, yellowish keratoderma of the palms and soles.

Systemic symptoms are uncommon except when generalized erythroderma occurs, and then they may develop when the face becomes uniformly erythematous. Although they are rare, moderate to severe systemic symptoms may be observed in patients with generalized disease. The systemic symptoms include fever, malaise, lymphadenopathy, and arthritis. In addition, rheumatologic disorders, mainly inflammatory polyarthritis, have also been reported. In addition, increased severity and additional manifestations of acne conglobata, hidradenitis suppurativa, and folliculitis have been reported.

CLINICAL FINDINGS

Systemic findings in pityriasis rubra pilaris include fever, malaise, lymphadenopathy, and arthritis. In addition, rheumatologic disorders, mainly inflammatory polyarthritis, have also been reported. In addition, increased severity and additional manifestations of acne conglobata, hidradenitis suppurativa, and folliculitis have been reported.

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