Pityriasis rubra pilaris

leşanlı kapiller hastalığı

Pityriasis rubra pilaris is a skin condition characterized by red, scaly patches on the skin. It is also known as leşanlı kapiller hastalığı.
Patients with pityriasis rubra pilaris are often unresponsive to multiple therapies, both topical and systemic. The disease may be exacerbated by factors such as alcohol consumption, smoking, or exposure to environmental elements like sun or cold weather. Suicide remains a risk in patients with generalized disease.

When conventional treatment strategies fail, new therapeutic approaches may include the use of phototherapy (psoralen with ultraviolet A phototherapy), some may flare, and others require combination therapy. Immunosuppressive agents are of inconsistent benefit. Thus, controversy persists about the role of cyclosporine in patients with severe symptoms.

Isotretinoin has been reported to be of value, although a comprehensive review suggests that acitretin may be more effective in clearing lesions. Accordingly, most patients are treated first with megadoses of oral vitamin A or the anabolic steroid stanozolol, proved to be largely ineffective. In patients with severe symptoms, effective amelioration of the disease may require extracorporeal photopheresis. Immunosuppressive therapy with methotrexate, using the guidelines established for psoriasis, is effective in the adult-onset type I of pityriasis rubra pilaris.

Topical treatment with keratolytics (when possible, under an occlusive plastic dressing) has been beneficial in psoriasis and psoriatic arthritis, is effective in the adult-onset type I of pityriasis rubra pilaris.

Differential Diagnosis of Pityriasis Rubra Pilaris

Pityriasis rubra pilaris is generally believed to comprise more than a single entity, and a classification scheme for pityriasis rubra pilaris (types I-VI according to Griffiths and González-López) has been described. Although an underlying dysfunction in vitamin A metabolism has been suggested as a cause, the precise pathogenesis of this condition is not yet fully understood.

Systemic Therapies

1. Cyclosporine A (5 mg/kg/day)
2. Ultraviolet B phototherapy
3. Narrowband ultraviolet B phototherapy
4. Ultraviolet A1 phototherapy
5. Photochemotherapy (topical or systemic psoralen plus ultraviolet A)
6. Glucocorticoids (medium to high potency)
7. Vitamin D analogues
8. Keratolytics (salicylic acid, urea)

Cutaneous Lesions

Type I (classic adult) is the most common subtype, accounting for more than 50 percent of all cases of adult-type pityriasis rubra pilaris. This type usually starts in the fifth decade of life, affects the sexes equally, and can last several years. Sparseness of the scalp hair is occasionally seen.

Type II (juvenile) is the most severe form of the disease, affecting approximately 20 percent of patients. It usually begins before adolescence and can last for many years. Sparseness of the scalp hair and nail dystrophy are typical findings.

Type III (circumscribed) is the least severe form, affecting approximately 5 percent of patients. It usually starts in childhood and can last for only a few years. Sparseness of the scalp hair is occasionally seen.

Type IV (circumscribed juvenile) affects approximately 25 percent of patients. This type usually starts in childhood and can last for several years. Sparseness of the scalp hair is occasionally seen.

Type V (erythrodermic) is the most severe form of the disease, affecting approximately 5 percent of patients. It usually starts in the first and fifth decades of life and can last for many years. Sparseness of the scalp hair is occasionally seen.

Type VI (erythrodermic with arthritis) is the most severe form of the disease, affecting approximately 5 percent of patients. It usually starts in the first and fifth decades of life and can last for many years. Sparseness of the scalp hair is occasionally seen.

Prognosis and Clinical Course

Recurrences are recognized in up to 20 percent of patients, however, sometimes after long periods of remission. The duration of the disease varies widely, with some cases resolving within an average of 1-2 years, while others can last for many years. Sparseness of the scalp hair is occasionally seen.

Pathology

Pathologic findings in pityriasis rubra pilaris vary according to the duration of the disease. The findings are most likely to be diagnostic in the acute phase, when hyperkeratosis, acanthosis, and papillomatosis are observed (see Pityriasis Rubra Pilaris At a Glance). Further, these findings can be observed in the upper and lower dermis. However, in the chronic phase, the findings can be reduced or absent. The horizontal and vertical directions can be observed (see Pityriasis Rubra Pilaris At a Glance).