

A red man with islands of normal skin

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A man has red scaling skin all over his body, with islands of normal skin. What is this dermatosis?

Over a four-week period, a 48-year-old man developed almost universal red scaling skin (erythroderma) with islands of sparing (Figure 1). The process had started on the face and scalp and progressed over his trunk and arms and subsequently to his legs. His palms (Figure 2) and soles were covered by a thick layer of keratin (keratoderma) with an orange hue. There was no previous history of a similar episode. The patient was not on any medications. The skin biopsies showed a thickened epidermis and a loose laminated stratum corneum with retained nuclei (parakeratosis). The underlying dermis showed a scant infiltrate of lymphocytes lacking atypia (Figure 3).

Differential diagnosis

Universal red scaling skin (erythroderma, exfoliative dermatitis) is often associated with pruritus and constitutional symptoms such as fever or malaise. Lymphadenopathy may be present. The differential diagnosis includes the following disorders.

- **Dermatitis**, particularly atopic, contact allergic and photosensitive dermatitis, may progress to erythroderma. A detailed history – enquiring about previous episodes of dermatitis, asthma or allergic rhinitis, occupational

exposure to chemicals, and mode of onset – may offer clues to diagnosis. Skin biopsy in these disorders is characterised by keratinocytes within the epidermis that are separated by fluid (spongiosis) and lymphocytic inflammation.

- **Drug reactions** may evolve to erythroderma, particularly as part of the hypersensitivity syndrome. Lymphadenopathy, peripheral hypereosinophilia and systemic abnormalities such as hepatitis may be found. Skin biopsy may reveal hypereosinophilia with lymphocytes and necrotic keratinocytes.

- **Psoriasis** may be complicated by erythroderma. A prior history of the plaque form of psoriasis, psoriatic arthritis or nail changes may point to the diagnosis. Skin biopsy is helpful as psoriasis is characterised by neutrophil microabscesses in the stratum corneum and a hyperplastic epidermis.

- **Cutaneous lymphoma** may present as an erythroderma and keratoderma. Lymphadenopathy is often present. Abnormal lymphocytes (Sezary cells) may be found in the peripheral blood or on skin biopsy.

- **Pityriasis rubra pilaris** is the correct diagnosis. The term refers to red, scaling, follicular papules that are often seen at the edge of the erythrodermic skin, and the condition may be complicated by hair loss.

Pityriasis rubra pilaris is an uncommon dermatosis and the cause is unknown. A variant associated with HIV infection has been described. The diagnosis can be recognised by the typical history as seen in this patient, clinical features and biopsy findings. The condition may persist for six months or more.

Treatment

Treatments include oral retinoids, methotrexate, topical corticosteroids and phototherapy, but the condition is difficult to treat.



Figure 1. Red skin with islands of normal skin.



Figure 2. Thick keratoderma of the patient's palms, with an orange hue.

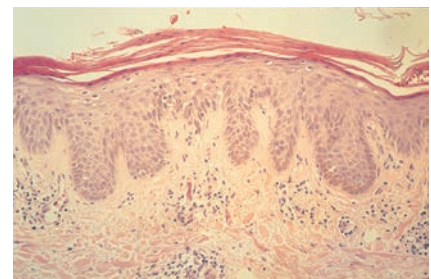


Figure 3. Skin biopsy demonstrating loose stratum corneum with retained nuclei (parakeratosis).

Keypoint

Erythroderma (exfoliative dermatitis) refers to a group of skin dermatoses in which there is severe widespread red skin associated with scaling and constitutional symptoms. **MT**

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