Current Problems in Dermatology

Exfoliative Dermatitis (Erythroderma)
Cutaneous Manifestations of Toxoplasmosis
The Acid Mucopolysaccharides of Skin
The Langerhans Cell
Tolerance and Desensitization in Experimental Eczema
The Role of the Basophilic Leucocyte in Hypersensitivity
Dermabrasion

4

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J.W.H. Mali, Nijmegen

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Current Problems in Dermatology

Vol. 4

Edited by J. W. H. MALI, Nijmegen

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Current Problems in Dermatology

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Introduction

The fourth volume of Current Problems in Dermatology tries to establish the trend started with the apparition of the third volume. It is our purpose to inform the practising dermatologist about the new developments in his field. Experts in their domain give an exposition of the trend of their investigations and enlarge thus the background on which the practising dermatologist has to act in his every day work.

As we work in the clinical situation many times outside of the territory of knowledge it is important to be sure of its border. It is only by applying the new concepts of general medicine and biology that dermatology can hope to remain a speciality of its own. The present development of molecular biology gives an unique opportunity to connect dermatology with the main stream of advancing biological science. The first two contributions in this volume are reviews of clinical conditions. ADAM treats the exfoliative erythrodermia's which constitute always a challange to the dermatologist. Significantly JUSTUS, the writer of the review on toxoplasmosis is a pathologist. It is to be expected that as awareness of this disease increases, the diagnosis may be more often and earlier made. MIER and WOLFF give reviews on subjects which have been for a long time so obscured by technical difficulties that it was impossible for the practical dermatologist to obtain an insight into them. Now new technical developments have made it possible for both contributors to give a balanced opinion about their subjects. WOLF-JÜRGENSEN brings new information about the elusive basophil leucocyte and forms the transition to the contribution of POLAK and FREY about tolerance and desensitization in experimental eczema. Absorption of their ideas will give the clinician the opportunity to rearrange his clinical observations. It seems not unlikely that at this point cross-fertilization of experiment and clinic might bring real progress in the prevention of skin disease.

Finally, EPSTEIN gives a personal opinion about a very practical subject, one of the few parts of dermatology in which the dermatologist can satisfy his urge as an artisan: dermabrasion.

J. W. H. MALI, Nijmegen

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Exfoliative Dermatitis (Erythroderma)

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Contents

Exfoliative dermatitis may be defined as a persistent, generalized, indurated erythema of the skin (erythroderma), followed by scaling (exfoliation). Erythroderma is usually applied when the exfoliation is not a conspicuous

Table I. Exfoliative dermatitis

Primary etiology	Incidence %
Pre-existing skin disease	30–40
Atopic dermatitis	
Psoriasis	
Seborrheic dermatitis	
Pemphigus foliaceus	
"Id" reaction	
Other eczemas	
Senile dermatitis	
Drugs	10
Penicillin	
Barbiturates	
Gold	
Arsenic, etc.	
Malignancy	10–15
Lymphoreticulosis	
Other organs	
Contactants	1
Chrysarobin	
Mercury	
Hereditary	1
Pityriasis rubra pilaris	
Ichthyosiform erythroderma	
Infantile	1
Ritter's disease	
Leiner's disease	
Erythrodermic Candidiasis	
Toxic Epidermal Necrolysis	
Unknown	30–40

clinical feature, but for practical purposes, the terms should be regarded as synonymous.

Etiology

Exfoliative dermatitis should always be considered as a secondary diagnosis, i. e. a "reaction" to some other etiological factor. The primary etiology and its relative incidence is classified in table I [ADAM, 1968]. The ability to detect the underlying etiology will depend upon the determination of the

attending physician to investigate the numerous avenues necessary which will lead to the ultimate diagnosis.

Although exfoliative dermatitis is usually found to occur most commonly in middle to latter life (45–70 years), it may be found in all age groups. The debilitating effects of new illnesses and the drugs used to treat them; as well as the increased incidence of malignancy in the older age groups are factors which may be responsible for the increased incidence during these years.

By and large, the most frequent primary etiology found to be responsible for exfoliative dermatitis is generalization of a pre-existing skin disease. Atopic eczema, psoriasis and the other eczemas such as stasis eczema; seborrheic eczema; nummular eczema, etc. may extend to involve the entire body, or may develop so rapidly for no obvious reason, as to present initially as an exfoliative dermatitis. Rarely, even lichen planus and erythrodermic (Norwegian) scabies may present as an erythroderma. History of a pre-existing skin disease should not deter an investigation looking for other causes; i. e. a patient with long-term well-controlled psoriasis who suddenly developed exfoliative dermatitis was found to have a systemic malignancy [TELNER and ADAM, 1965]. Generalization of a pre-existing dermatosis may be a sequela of topical overtreatment or systemic drug therapy and should not be overlooked as the *modus operandi* in erythrodermic patients who had previously well-controlled localized skin disease.

Drugs alone may be the primary etiologic agent. There is no accurate test to determine drug allergy; and the diagnosis can only be established by careful detailed history, frequently rechecked, as well as a strong suspicion and the simultaneous exclusion of other possible causes. Alternatively, patients originally suspected of having drug-induced disease may, in time, demonstrate a more clearcut etiologic basis for their condition, such as pityriasis rubra pilaris or mycosis fungoides. Any drug from penicillin and the barbiturates, to arsenic and gold may be responsible for the erythroderma.

Malignancies of the reticulo-endothelial system and other organs such as lung, gastrointestinal tract and pancreas, etc. may develop exfoliative dermatitis during the course of their disease. A silent hidden malignancy should always be considered as a possible etiology among the older age groups who develop this condition.

Frequently, in a large group of patients, the primary basic etiology cannot be determined even with extensive detailed investigation. This may be due to the fact that the patient is a poor historian and drug ingestion was forgotten; or investigation failed to detect an occult malignancy; or the disease has not become fully developed such as in the premycotic stage of mycosis

fungoides or in early pityriasis rubra pilaris. In the elderly, the gradual progression of xerosis or asteotic (senile) dry skin, into exfoliative dermatitis may be overlooked as a possible etiologic source. Therefore, "unknown" does not fulfill a criteria for considering this to be a primary erythroderma. It means only that the basic etiology remains to be discovered and that the exfoliative dermatitis is still secondary but not catergorized; but, there are still some present-day authors who consider primary or idiopathic erythroderma as a definite entity [MONTGOMERY, 1967].

Sex incidence has shown that males are found to be affected between two to three times more frequently than females.

Clinical Features

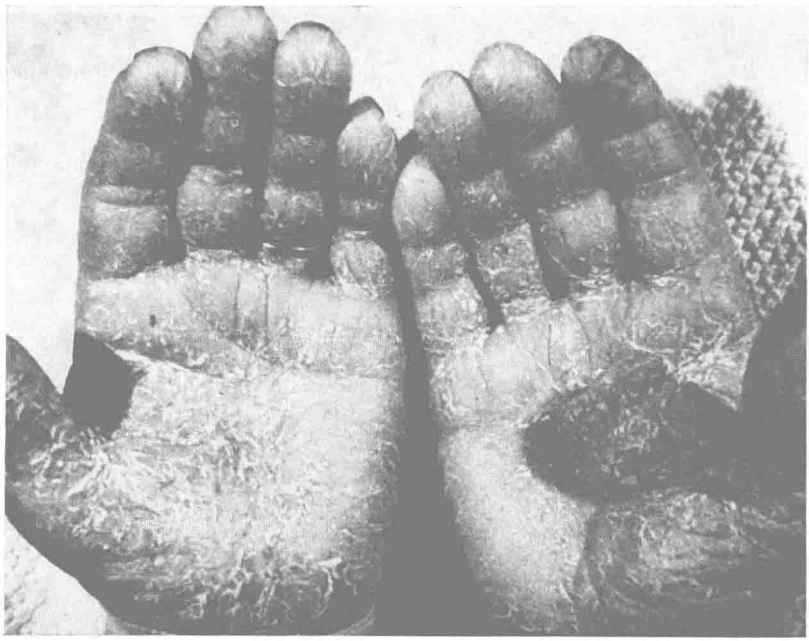
The condition may begin as a pre-existing cutaneous disease (psoriasis, eczema) which becomes widespread; or as a patchy eczema of sudden onset which rapidly generalizes in 12–48 h. The intensity of the erythema fluctuates, but the skin is generally hot, red, dry and indurated (l'homme rouge). Constitutional features of malaise, shivering, chills and fever frequently accompany the extensive erythema.

Scales appear after the first few days, varying from fine and bran-like, to profuse desquamation in large sheets. The patient's skin feels tight and dry, but the discomfort may become so severe that pain and tenderness limits all movements, and he lies motionless in his bed. Edema of the skin is often present and may be pronounced in dependent areas (legs, sacrum), sometimes leading to oozing in the large joint flexures. Small islands of normal skin are sometimes seen amid the general desquamation. Palms and soles may show thick peeling areas and/or fissuring, or may be normal (fig. 1). The character of the erythema and scaling usually obscures the previous underlying skin condition, although in psoriatic patients some features may occasionally persist.

Pruritus is usually pronounced, leading to lichenification (fig. 2) and excoriations, and occasionally to secondary infection with honey-colored yellow impetigo crusts or purulent discharge. Pyodermic nodules, infected biopsy sites and the presence of large numbers of *Staphylococcus aureus* on the peeling skin make these patients reservoirs of pathogenic organisms which can easily disseminate to adjacent hospital patients.

Fig. 1. Erythrodermatous hands showing fissuring, thickening and scaling.

Fig. 2. Back with lichenification, infiltration and scaling.



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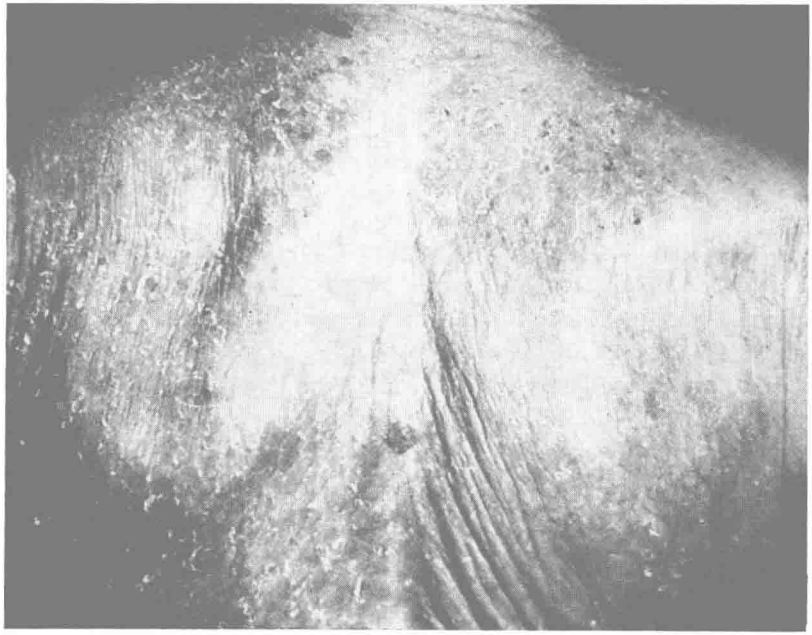

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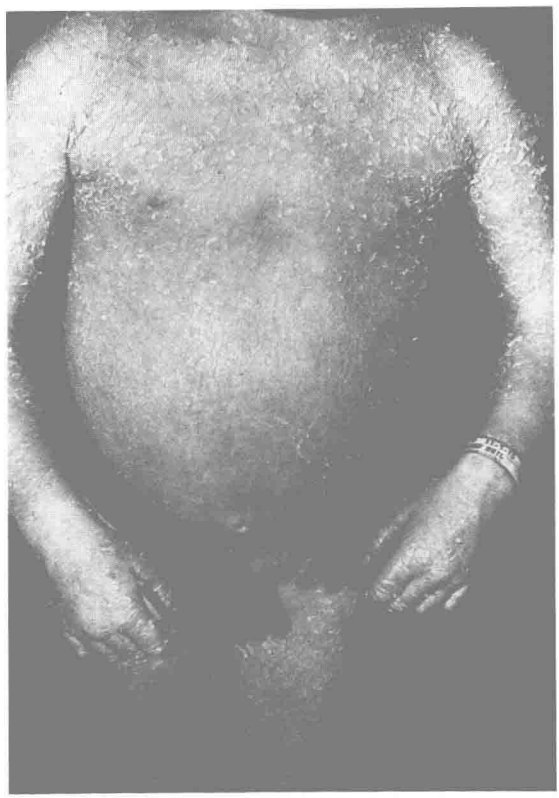

Fig. 3. Psoriatic erythroderma in a young child.

The rancid odor frequently associated with erythrodermatous patients is due to the decomposing action of the resident bacteria (gram-positive staphylococci) on the apocrine secretion and the proteinaceous scaling debris [HURLEY and SHELLEY, 1960; SHEHADEH and KLIGMAN, 1963].

If the erythroderma is present for some weeks, loss of scalp and body hair may occur. It is usually slight but may be diffuse, with complete alopecia occurring only rarely in prolonged cases. Dystrophic nail changes are often seen, especially in patients with hair loss. The nails may appear opaque, yellowish, thickened, pitted and assume linear grooves and ridges. The distal portion of the nails often appear shiny, as if polished; and is due to the frequent rubbing of pruritic skin.

Pigmentation may give the patient's skin a brown to slate-gray appear-

ance and can persist as postinflammatory hyperpigmentation. In the dark-skinned races the reverse is usually the case with widespread or patchy loss of pigment (fig. 1–3).

Lymphadenopathy is present in nearly all cases; firm, non-tender, freely movable nodes are found primarily in the groin and axillae. Suppuration does not occur and the glands return to normal size as the skin improves, unless reticulosis is present.

Hepatomegaly and splenomegaly are occasionally found to be present but this does not necessarily imply malignancy or a bad prognosis. Only a small proportion of patients have malignant reticulosis even with extensive hepatosplenomegaly.

Duration

Exfoliative dermatitis may persist for as little as a few months to more than 10 years, depending on the etiology. Erythroderma secondary to drugs, psoriasis or eczema can be expected to respond more readily to therapy than those due to other conditions; but complete clearing usually takes an average of over 3 months. Patients with malignancies may have erythroderma continuing throughout the entire course of their tumor, i. e. until they are cured or die from their malignancy. In patients with mycosis fungoides, an erythrodermatous phase may persist unabated for years before the diagnosis is confirmed histologically. Therefore, once the diagnosis of exfoliative dermatitis is made, active treatment will be necessary for months or even years in the majority of patients.

Investigations

Hematological examinations are usually not characteristic although anemia is sometimes found. Elevated white cell counts would be suggestive of either infection or leukemia; and differential counts often show an eosinophilia, sometimes up to 50% in atopic erythroderma. The erythrocyte sedimentation rate is elevated in only 50% of patients (a very rapid sedimentation rate may indicate lymphoma). Bone marrow findings are not diagnostic unless a specific lymphomatous process is present. Serum protein estimations are often less than 6 g/100 ml with a reversal of the albumin-globulin ratio. Other liver function tests are usually normal.