Erythroderma

Nick J Levell

Abstract

Erythroderma is a clinical syndrome producing generalized red skin. The presentation may be acute or chronic. In acute erythroderma there may be 'skin failure' leading to life-threatening systemic manifestations, requiring supportive care on an intensive care unit, whereas in chronic erythroderma patients systemic problems are usually absent. The condition may be due to inflammatory, or rarely, neoplastic processes. Common causes are psoriasis, eczema and drug eruptions. The presentation may be dramatic, and doctors must ensure they are not distracted from a diagnostic process based on careful history-taking and examination. Specific skin therapy will depend on the nature of the underlying cause and the severity and time course of the condition. Close collaboration between general physicians, intensive care physicians and dermatologists is necessary for successful management.

Keywords drug rash; eczema; erythroderma; psoriasis; sub-erythroderma

What's new?

- Anti-tumour necrosis factor biological therapy is effective in the treatment of severe psoriasis
- Drug rash syndromes include drug reaction with eosinophilia and systemic symptoms (DRESS) and acute generalized exanthematous pustulosis (AGEP)
- Erythroderma may be a feature of HIV infection

Common causes

Erythroderma (Table 1) is not a skin disease but a clinical syndrome that can be due to many different causes. The common causes presenting to doctors in secondary care are psoriasis, atopic eczema, seborrhoeic eczema, other forms of eczema and drug eruptions. There are numerous rarer causes.[1] Erythroderma in the neonate may be due to inherited disorders or to staphylococcal scalded skin syndrome.

Diagnosis

Diagnosis is based mainly on the history, examination and skin biopsy; other investigations are only sometimes of use.

History

How did the rash start? Was it a sudden-onset generalized rash with no preceding history — perhaps suggesting a reaction to a sudden toxic insult such as a medication, allergy or infection? Had it been present for years, fluctuating in severity — indicating chronic eczema or psoriasis? Did it start in a localized itchy rash and then disseminate — more typical of eczema? Did it start as multiple small patches that became confluent following a sore throat — pointing towards erythroderma following guttate psoriasis? Did it start gradually and insidiously spread to cover the body over months with a non-itchy rash — perhaps pointing towards cutaneous lymphoma?[2]

Has the patient a past history of eczema (dermatitis) or psoriasis? Many patients do not realize that they have mild psoriasis. A history of scaling of the scalp, elbows and knees may be important. Other patients may not realize that their dry, discoloured or sensitive skin is eczema.

Have any drugs been recently started? Specifically ask about all drugs, vitamins, herbal remedies and alternative medicines. Ask about remedies that are taken intermittently for bowels, cramps and analgesia. A new medicine typically would cause a rash within a few days but the rash may be delayed by many months or even years. Withdrawal from oral corticosteroids or initiation of lithium treatment may trigger erythrodermic psoriasis but many other drugs, including illegal sulfur mustard gas exposure in wars, have been implicated.[3,4,5]

Has the patient had previous allergic reactions to drugs? Ask about allergies to creams (some drugs are topical). Ask about application of cosmetics and sun creams.

Is the patient photosensitive? Ask about sun exposure and sunbeds. Consider phototoxic drug reactions, other photodermatoses and systemic causes such as systemic lupus and variegate porphyria.

General history: important systemic symptoms of skin failure include thirst, due to dehydration, and abnormal temperature perception with shivering. Ask about symptoms relating to underlying malignancy.[6]

Contacts: if family or friends are itching, consider scabies. Crusted (Norwegian) scabies is highly contagious.

Clinical examination

General examination must include measurement of core temperature to identify hypothermia, assessment for dehydration, and examination of respiratory and cardiovascular systems for signs of sepsis, adult respiratory distress syndrome and high output cardiac failure.[7]

Skin examination: full examination of skin, mucosae, scalp and nails is needed. Clues to the cause of the erythroderma may be

Nick J Levell *MD FRCP MBA is Clinical Director in Dermatology at the Norfolk and Norwich University Hospital, UK. He is a Senior Lecturer at Norwich Medical School and President of the British Society for Medical Dermatology. He has wide clinical research interests working with the UK Dermatology Clinical Trials Network to deliver studies through the NIHR. He also has research interests in epidemiology, health economics, health service delivery and history of medicine. Competing interests: none declared.*

Types of erythroderma

Erythroderma

- More than 90% of the skin is red

Sub-erythroderma

- 70–90% of the skin is red

Table 1

subtle. Severe skin disease may be associated with ocular disease, which can cause scarring and blindness.

Investigation

Skin biopsy: this may not be necessary if there is a clear history and signs of generalized spread of pre-existing eczema or psoriasis. Histology may be useful to confirm severe drug rashes (see Drug eruptions on pages 360–364 of this issue).

Pityriasis rubra pilaris (PRP) and mycosis fungoides (MF) produce characteristic histology and, in lymphoma, a T-cell clone may be isolated.

In all cases of erythroderma consult with a dermatologist before taking the biopsy to ensure that the samples are taken from the correct area and for appropriate tests. An inexperienced person may accidentally biopsy a coincidental benign cutaneous lesion, mistaking it for a florid area of the rash causing diagnostic confusion.

HIV infection: severe psoriasis and crusted scabies may be found in HIV. The seroconversion illness may produce generalized erythema.[8]

Treatment

General principles

Skin controls body temperature, retains fluid and acts as a barrier against infection. Erythrodermic patients may develop skin failure and lose these functions. Skin failure is characterized by erythroderma, oedema, tachycardia, hypothermia or hyperthermia and is more dangerous in neonates and in the elderly who are more susceptible to secondary infection. Intensive care monitoring of fluid balance and core temperature is sometimes needed in an acute severe erythroderma. In extreme cases, particularly in the elderly, high-output cardiac failure and acute adult respiratory distress syndrome have been reported.[9] Advice from multiple specialities may be needed. Ophthalmic care is needed for patients with associated ocular disease. However, many patients with chronic erythrodermic eczema and psoriasis cope surprisingly well as outpatients.

Good skin care is essential. Avoid adhesive tape on fragile skin to avoid tearing, hold dressings in place with circumferential bandaging adherent to itself, and use beds or mattresses that minimize pressure damage.

Pain relief is essential in acute erythroderma.

Swab the skin and affected mucosa regularly in acute patients to identify secondary infection early. Reverse barrier nurse to protect against hospital infections.

Patients are catabolic and serum albumin may fall considerably. Nutritional support may be needed and expert dietetic advice should be sought.

Specific conditions causing erythroderma (Table 2)

Eczema

Lichenification (thick skin due to scratching and rubbing) is characteristic of chronic eczema (Figure 1). Acute eczema tends to be weepy, crusty and may blister. Secondary infection with either bacteria (*Staphylococcus* or *Streptococcus*) or virus (herpes simplex) must be considered. If multiple tiny vesicular or crusted lesions are present consider eczema herpeticum (caused by herpes simplex).[8] Take swabs for bacterial and herpes viral culture (in the correct medium) and treat with oral antibiotics and/or aciclovir.

Acute weepy eczema may require antiseptic soaks such as potassium permanganate or saline. Dryer areas will require emollients such as a 50/50% mixture of liquid paraffin and white soft paraffin. An emulsifying ointment should be used for bathing. Very potent topical corticosteroids are needed in acute eczema.

Patients with chronic erythrodermic eczema require assessment by a consultant dermatologist to exclude underlying triggering factors such as allergy. Such patients may require systemic azathioprine or ciclosporin as longer-term corticosteroid-sparing agents.

Psoriasis

Erythrodermic psoriasis ranges in colour from deep red to salmon pink, often with a scale (Figure 2). There may be pustules or peeling (desquamation) if it is acute (Figure 3). In chronic psoriasis then there may be considerable scaling. Psoriasis is associated with arthritis, nail pits and onycholysis.

Acute unstable or pustular psoriasis is usually treated with twice-daily topical moderate-potency corticosteroids and bed rest. Treat any streptococcal infection. Systemic treatments with oral ciclosporin (3–5 mg/kg/day) or methotrexate (7.5–25 mg/week) are used. In patients who do not respond to these medications, anti-tumour necrosis factor and other biological treatments are used in the UK according to NICE guidelines.[10] Avoid systemic corticosteroids in psoriasis as withdrawal can be difficult leading to severe flares.

Drug reactions

The suspected offending drugs should be withdrawn, but it may take several months for the rash to settle completely (see Drug eruptions on pages 360–364 of this issue).[11] In toxic epidermal

Causes of erythroderma

- Psoriasis
- Eczema (e.g. atopic, seborrhoeic)
- Drug eruptions (e.g. drug reaction with eosinophilia and systemic symptoms, acute generalized exanthematous pustulosis, toxic epidermal necrolysis)
- Pityriasis rubra pilaris
- Cutaneous lymphoma (e.g. Sezary syndrome)
- Crusted scabies
- Staphylococcal scalded skin syndrome and toxic shock syndrome
- Congenital ichthyoses (many different diseases)
- Pemphigus and bullous pemphigoid
- Netherton syndrome and other rare genodermatoses

Table 2