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Dermatological emergencies: What the term encompasses and key features in their diagnosis

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BACKGROUND

Dermatology has traditionally been considered to be an outpatient-centered specialty, with very few emergencies and lethal conditions; so the term *dermatological emergencies* may seem like an oxymoron. However, there are several life-threatening dermatological conditions that may become lethal in about 8% of cases, unless adequate emergency care (prompt management and monitoring in the intensive care unit) is provided, contrary to popular belief [1]. Although dermatologists may encounter several such conditions in their career, proper awareness of them is still lacking in many parts of the world.

Recently, most of these conditions have been incorporated under the broad term *acute skin failure*, where there is extensive structural disruption and total functional impairment of the skin [2,3]. Acute skin failure is an acute emergency characterized by aberration of thermoregulation, fluid electrolyte imbalance, loss of proteins resulting in malnutrition, and failure of the mechanical barrier to prevent entry of infective organisms and other foreign materials. Thus, prompt diagnosis and emergency intervention are needed to revive the patient. Although this condition is as severe as other organ dysfunctions, viz. cardiac, renal, pulmonary, or hepatic failure, this condition is still not well known, and the need for intensive care in dermatology is often overlooked. This entity is discussed in detail in a separate chapter. The aim of the current chapter is to provide a brief introduction to these different dermatological emergencies.

ETIOLOGY

Several conditions may present as dermatological emergencies in both the pediatric (including neonates) and adult populations, which may pose diagnostic and therapeutic challenges. These conditions encompass both primary cutaneous disorders and dermatological manifestations of severe systemic disorders. Some of the important etiologies have been tabulated in Tables 1.1 and 1.2 [3–5].

Table 1.1 Causes of neonatal and pediatric dermatological emergencies

Disorders that can manifest as emergencies

A. Neonatal erythroderma
- Ichthyosiform erythrodermas (bullous/nonbullous/lamellar/X-linked/harlequin)
- Collodion baby
- Netherton syndrome
- Immunodeficiency disorders (Omenn syndrome, graft versus host disease, severe combined immunodeficiency disorder)
- Papulosquamous disorders (congenital erythrodermic psoriasis)
- Drug induced (vancomycin, ceftriaxone)
- Others (diffuse cutaneous mastocytosis, Leiner disease)

B. Neonatal cutaneous infections
- Bacterial (staphylococcal scalded skin syndrome, necrotizing fasciitis)
- Viral (neonatal varicella, neonatal herpes infection)
- Fungal (congenital/neonatal candidiasis)

C. Vesiculobullous disorders (epidermolysis bullosa, pemphigus)
D. Metabolic disorders (biotin deficiency, essential fatty acid deficiency)
E. Vascular complications (Kasabach-Merritt phenomenon)
F. Others

Purpura fulminans
Kawasaki disease
Sclerema neonatorum

Table 1.2 Causes of dermatological emergencies in adults

1. Erythroderma (idiopathic, dermatitis [atopic/seborrheic], psoriasis, pityriasis rubra pilaris, drug induced, other keratinization disorders, Sézary syndrome)
2. Drug reactions (Stevens-Johnson syndrome, toxic epidermal necrolysis, drug hypersensitivity syndrome)
3. Acute generalized pustular psoriasis
4. Immunobullous disorders (pemphigus vulgaris and foliaceus)
5. Infections (staphylococcal scalded skin syndrome, febrile viral exanthemas)
6. Emergencies where systemic involvement is seen in association with dermatological manifestations
   i. Connective tissue disorders
   ii. Vasculitis
   iii. Malignancies
7. Emergencies related to dermatosurgery:
   i. Anaphylaxis (adverse reactions to intravenous antibiotics, local anesthetics, and other allergens like latex rubber)
   ii. Syncope—vasovagal
   iii. Lignocaine-related emergencies like syncope, anaphylaxis, bleeding, cardiac arrhythmias, etc.
   iv. Bleeding
   v. Acute stroke, status epilepticus, cardiac arrest in the operating room
   vi. Hypotension
   vii. Hypoxia
8. Emergencies related to Hansen's disease or leprosy
   i. Type 1 reaction that can be upgrading/downgrading type
   ii. Type 2 reaction—multisystem involvement along with skin lesions called erythema nodosum leprosum
   iii. Lucio phenomenon
9. Sexually transmitted diseases associated emergencies

ETIOLOGY BASED ON CLINICAL PRESENTATION

Although dermatological emergencies have been classified according to the underlying cutaneous/systemic disorder, often such a diagnosis becomes difficult in the emergency. So we have provided a set of conditions that may lead to dermatological emergencies, based on their clinical presentation. Skin rash is the most common presentation in the dermatology emergency, with or without fever. We have highlighted several causes based on the characteristic of skin rash (Table

1.3) [6,7].

Table 1.3 Nature of skin rash and possible dermatological emergency

Nature of skin rash	Possible dermatological emergency
• Erythrodermic (redness with scaling >90% body surface area)	Causes listed in Tables 1.1 and 1.2
• Maculopapular	Viral exanthem (may be associated with hemorrhagic features and shock as exemplified by dengue hemorrhagic fever and dengue shock syndrome, respectively, to form a dermatological emergency)
• Purpuric/petechial	Vasculitides, purpura fulminans/disseminated intravascular coagulation, Rocky Mountain spotted fever, meningococcemia, scarlet fever
• Pustular	Disseminated candidiasis, pustular psoriasis, acute generalized exanthematous pustulosis
• Vesiculobullous	Neonatal herpes simplex, pemphigus, bullous pemphigoid, herpes zoster
• Hemorrhagic/erythematous	Vasculitis, Kawasaki disease, erythema multiforme, Stevens-Johnson syndrome, toxic epidermal necrolysis

APPROACH TO DIAGNOSIS

Whenever a patient presents to the dermatological emergency, prompt diagnosis and early management should be our aim to reduce morbidity and mortality. Like all other diseases, this process involves adequate history and physical examination to arrive at the correct diagnosis.

History

A detailed history regarding the nature of rash and other key aspects is of paramount importance to arrive at the accurate diagnosis. However, the first priority is to assess whether the patient is seriously ill and needs immediate management or is relatively stable to provide history. Some important points need to be evaluated during this process as given in Table 1.4. Though a detailed history is necessary, some facets may be curtailed depending on the situation and dermatologist's experience.

Table 1.4 Important elements of history for acute dermatological emergencies

- Mode of onset—acute onset or acute exacerbation of a chronic disorder
- Distribution/spread—localized (zoster, vasculitides)/generalized (varicella, staphylococcal scalded skin syndrome, erythroderma)
- Site of onset
- Family/contact history—to rule out contagious diseases
- Travel history—to exclude endemic conditions, e.g., Rocky Mountain spotted fever
- Animal exposure
- Drug history—to rule out drug reactions, e.g., erythroderma
- Any history of allergy (drug/chemical)
- Occupational history
- Vaccination status
- Sexual history
- Any exacerbating/relieving factor, e.g., climate, chemicals, etc.
- Associated symptom—fever/itching or pain

Physical examination

Physical examination should follow history taking in order to narrow down the list of differentials, involving general, dermatological, and systemic evaluation. A few aspects of general and systemic examination should be undertaken in all cases:

- Pallor, cyanosis, jaundice, edema, clubbing, lymph nodes, and vitals should be checked thoroughly to rule out any systemic disease or life-threatening condition like sepsis or anaphylaxis.
- If any systemic disorder is suspected, specific attention should be paid to some conditions:
  - Any murmur, particularly new onset
  - Generalized lymphadenopathy
  - Hepatosplenomegaly
  - Arthritis
  - Oral, conjunctival, or genital lesions
  - Any sign of neurological dysfunction like meningeal irritation

Dermatological examination

A proper dermatological examination is mandatory to arrive at the correct clinical diagnosis and initiate appropriate management at the earliest opportunity. Some important points need to be kept in mind during cutaneous examination:

- Dermatological examination involves checking the hair, skin, nails, and mucosae (oral, genital, ocular) apart from skin.

- Examination should be performed from head to toe after undressing the patient, in adequate illumination to appreciate any subtle color/textural change of the skin.
- Any cream/makeup should be removed, which may obscure the true nature of the lesion.
- Inspection and palpation are the components of cutaneous examination. A magnifying glass may be used if clarity is needed. An overall inspection of skin lesion/rash is necessary, as many skin diseases can be diagnosed by their characteristic appearance or morphology of the lesions [8].
- The entire lesion should be palpated with a gloved finger to assess its texture and detect any blanching or bleeding under pressure.

A focused approach is needed regarding the dermatological examination, to save time and arrive at the correct diagnosis promptly. We look out for the types of skin lesions and their other aspects during cutaneous examination.

TYPE OF SKIN LESION

Skin lesions can be broadly categorized into two types: primary and secondary. Primary lesions are basic physical changes in the skin that are caused directly by the disease itself, while these lesions mature over time to give rise to secondary changes. Primary and secondary skin lesions are presented in Tables 1.5 and 1.6.

Table 1.5 Primary skin lesions

Name of lesion	Characteristic feature
Macule	Circumscribed area of color change, flat lesion; <0.5 cm in diameter
Patch	Circumscribed area of color change, flat lesion; ≥0.5 cm in diameter
Papule	Solid, circumscribed raised skin lesion, diameter <0.5 cm; many additional features have been described like domed, flat-topped, umbilicated
Plaque	Solid, raised plateau-shaped skin lesion, ≥0.5 cm in diameter
Nodule	Raised solid lesion with deeper infiltration (height more than width)
Tumour	Skin swelling >2 cm in diameter
Vesicle	Well-circumscribed, clear fluid–filled lesion <0.5 cm in diameter
Bullae	Well-circumscribed, clear fluid–filled lesion ≥0.5 cm in diameter
Pustule	Circumscribed, elevated lesions containing pus; are commonly infected (as in folliculitis) but may be sterile (pustular psoriasis)
Wheal	Edema of skin, evanescent lesions

Table 1.6 Secondary skin lesions

Name of lesion	Characteristic feature
Erosion	Total or partial loss of epidermis; does not heal with scarring
Ulcer	Total loss of epidermis with or without dermal involvement, heals with scarring
Excoriation	Scratch marks, may result in erosions/ulcers
Fissures	Crevices in skin that may extend upto the dermis
Scale	Flakes of dead and dry skin
Crust	Dried-up purulent material, blood, or other exudate
Atrophy	Loss or thinning of epidermis, dermis, or subcutaneous tissues; skin appears white, papery, and translucent with loss of surface markings
Pigmentation	Hypo- or hyperpigmentation that may occur as part of disease process or after healing (post inflammatory)
Striae	Linear, atrophic lesions due to degeneration of connective tissue that may appear pink/red depending on stage; common site of topical corticosteroid abuse

OTHER IMPORTANT ASPECTS OF SKIN LESIONS

After determining the nature of a skin lesion (primary/secondary), we need to evaluate the other aspects of these lesions, such as morphology, color, arrangement, distribution, extent, and evolutionary changes, to obtain further clues regarding the disease.

Investigations

Investigations form an essential part of patient evaluation, as they help us to confirm the suspected diagnoses. They can be categorized under two heads:

- Bedside diagnostic procedures
- Laboratory investigations

BEDSIDE DIAGNOSTIC PROCEDURES

Some simple bedside diagnostic procedures may be performed to diagnose and differentiate different skin problems at the first instance [7]:

- *Diascopy*: Diascopy is a technique in which a glass slide is pressed firmly against the lesion to exert pressure on the lesion. It is primarily used to distinguish hemorrhagic and inflammatory lesions. While inflammatory lesions blanch with pressure (e.g., erythema), hemorrhagic lesions remain nonblanchable (petechiae

or purpura).

- *Nikolsky sign*: This sign refers to sloughing and erosion of skin when a shearing force is applied on normal skin in the perilesional area. This sign is positive in superficial blistering diseases like pemphigus, toxic epidermal necrolysis, and staphylococcal scalded skin syndrome (SSSS). It may be seen in severe cases of erythema multiforme, epidermolysis bullosa, pemphigoid, and variegate porphyria.
- *Bulla spread sign*: This is helpful in bullous disorders and confirms the location as epidermal like in pemphigus.
- *Grattage and Auspitz sign*: Taking out scales leads to positive grattage, while pinpoint bleeding after further scratching leads to the Auspitz sign, which is characteristic of psoriasis.
- *Dermatographism*: This sign refers to exaggeration of the physiologic triple response of Lewis (whealing tendency) on application of pressure. This is the hallmark of physical urticaria.
- *Wood lamp*: This is a useful clinical screening tool for several conditions like porphyria. It is an ultraviolet lamp with wavelength of 360–365 nm.
- *Dermoscopy*: Dermoscopy is a relatively new diagnostic maneuver often helping in the bedside diagnosis of several dermatological conditions, such as melanoma (atypical pigment network and vessels, blue-white veil, regression structures, irregular streaks, eccentric homogeneous pigmentation, and asymmetrical pigmented globules) and nonmelanoma skin cancers, such as squamous cell carcinoma (central yellow keratinous plugs with arborizing vessels centrally and hairpin vessels peripherally) and basal cell carcinoma (multiple blue-gray dots, globules and ovoid nests, leaf-like areas, arborizing vessels, erythema, erosions, and ulceration) [9]. These malignancies need a hasty diagnosis to avoid complications including metastasis and lethality.
- Swabs may be taken from discharge/ulceration for microbiological examination.
- Gram staining is a vital tool to detect and classify bacterial infections.
- Skin scrapings and microscopy may be done to confirm fungal infections (potassium hydroxide exam [KOH test]) or ectoparasite infestations such as scabies.
- Tzanck smear may be done if viral infection (herpes, varicella) or bullous disorder is suspected.

LABORATORY INVESTIGATIONS

Laboratory investigations include venereal disease research laboratory (VDRL), rapid plasma reagin (RPR), enzyme-linked immunosorbent assay (ELISA) for HIV,

antinuclear antibody (ANA), and other such investigations to confirm the various clinical diagnoses.

Differential diagnoses

When a patient presents to the dermatological emergency with skin rash, a plethora of differential diagnoses need to be considered. It is often a challenge for the emergency physician to differentiate benign skin disorders from the more serious, fatal conditions that require immediate intervention.

The authors classified five major types of skin rash that may be encountered in the emergency: (1) maculopapular, (2) erythematous, (3) petechial/purpuric, (4) pustular, and (5) vesiculobullous.

All of these varieties of rash have been discussed briefly along with the major differentials and possible diagnostic workup based on clinical presentation. After a suspected diagnosis has been made, efforts should be made to confirm the same (tests discussed above) and then stabilize the patient.

MACULOPAPULAR RASH

Maculopapular rash is the most common type of skin rash, which can be differentiated based on its distribution and systemic features as given in Table 1.7 [7].

Table 1.7 Differential diagnoses of maculopapular rash

Features of rash	Possible diagnosis
Centrally distributed rash with systemic toxicity	Viral exanthema Lyme disease Typhus fever
Centrally distributed rash without systemic toxicity	Cutaneous adverse drug reaction (history of drug intake positive)
Peripherally distributed rash with systemic toxicity	Secondary syphilis Meningococcemia Rocky Mountain spotted fever Erythema multiforme Stevens-Johnson syndrome (early)
Peripherally distributed rash without systemic toxicity	Scabies Eczema Psoriasis

ERYTHEMATOUS RASH