Emergency dermatology and need of dermatological intensive care unit (DICU)

Iffat Hassan, Parvaiz A Rather

Postgraduate Department of Dermatology, STD & Leprosy, Govt. Medical College, Srinagar, J & K India

Abstract

Dermatological emergencies comprise diseases with severe alterations in structure and function of the skin, with some of them leading to acute skin failure that demands early diagnosis, hospitalization, careful monitoring and multidisciplinary intensive care to minimize the associated morbidity and mortality. Prompt intensive management of acute skin failure in the ICU on the lines of 100% burns is mandatory; clearly establishing the necessity of a dedicated intensive care unit comprising of well synchronized team of dermatologist, internist, pediatrician, critical care physician and skilled nursing staff. In this article, we review the literature and discuss the major causes of dermatological emergencies, some of which lead to acute skin failure and lay stress for their management in ICU like set up attached to dermatology department itself, i.e., dermatological intensive care unit (DICU), so that such emergencies may be dealt with more effectively and without wastage of time. DICU should be equipped to such an extent that it provides initial, immediate and necessary support and it need not be as advanced and sophisticated as cardiac, surgical or neonatal ICU.

Key words
Dermatological emergencies; acute skin failure; dermatological intensive care unit (DICU).

Introduction

Dermatology is often thought of as a non-acute, outpatient-centered specialty. However, there are many dermatological conditions presenting as emergency situations (Table 1). It has been reported, however, that on an average, approximately 5% to 8% of all emergency department visits are due to dermatological conditions, with variations from 4.8% to 21% in different studies.

Dermatological emergencies can be primary, where involvement of skin is the primary cause and/or major manifestation or associated with medical/surgical emergencies, where cutaneous manifestations are the indicators of impending or underlying severe systemic involvement. Mortality and morbidity due to dermatological emergencies, among other things, is related to age at presentation, severity and preparedness to deal with the condition. Sudden severe alterations in the anatomy and physiology of skin consequent to some of the generalized dermatoses presenting as emergency situation, can lead to disabling complications eventuating in the potentially fatal condition of acute skin failure. With the availability of effective drugs, monitoring facilities and awareness of need for immediate care, there has been a significant decline in the fatality rate associated with dermatological emergencies. Understanding the etiopathogenesis of various systemic complications of acute skin failure and their prompt management in ICU on lines similar to that of burns can salvage many lives. In this
review, we describe major causes of dermatological emergencies, some of which lead to acute skin failure and outline the general management of acute skin failure in intensive care unit setting.

### Causes of dermatological emergencies

The major causes of emergencies due to dermatological conditions are listed in Table 1.

#### 1. Emergencies related to clinical dermatological conditions:

**i) Erythroderma (exfoliative dermatitis)**

Erythema and scaling involving most of the body surface area (90%), develops either de novo (primary or idiopathic) or as a progression of a pre-existing skin disease (secondary). It can be acute (few days duration) or chronic. In adults, causes include eczemas of various types (40%); psoriasis (25%); lymphomas (15%); drugs (10%) like sulphonamides, dapsone, NSAIDs, antiepileptics, penicillins etc; hereditary causes like ichthyosis, pityriasis rubra pilaris (1%); pemphigus foliaceus (0.5%); staphylococcal scalded skin syndrome (SSSS); crusted scabies; dermatomyositis; lichen planus (0.5%) etc. Other cases are idiopathic (8%).

In pediatric age group, causes of erythroderma (red scaly baby) are varied. In one of the studies, the causes indentified were infections (40%); psoriasis (25%); lymphomas (15%); drugs (10%) like sulphonamides, dapsone, NSAIDs, antiepileptics, penicillins etc; hereditary causes like ichthyosis, pityriasis rubra pilaris (1%); pemphigus foliaceus (0.5%); staphylococcal scalded skin syndrome (SSSS); crusted scabies; dermatomyositis; lichen planus (0.5%) etc. Other cases are idiopathic (8%).

### Table 1 Common causes of dermatological emergencies

| 1. Erythroderma (exfoliative dermatitis) |
| 2. Urticaria, angioedema and anaphylaxis |
| 3. Bullous disorders |
| 4. Infections |
| Staphylococcal scalded skin syndrome |
| Neonatal cutaneous infections |
| Necrotizing fasciitis |
| Neonatal varicella |
| Neonatal HSV infection |
| Candidiasis |
| Other |
| 5. Drug reactions |
| 6. Connective tissue diseases |
| 7. Metabolic conditions: |
| 8. Miscellaneous |
| Kasabach-Merritt phenomenon |
| Purpura fulminans |
| Kawasaki disease |
| Sclerema neonatorum |
| Erythromelalgia |
| Other |
| 9. Emergencies related to sexually transmitted diseases (STDs) |
| 10. Emergencies related to leprosy |
| 11. Emergencies related to dermatosurgery procedures |
| Anaphylaxis |
| Vasovagal syncope |
| Lidocaine allergy |
| Acute stroke |
| Status epilepticus |
| Electrosurgery and pacemakers/defibrillators related emergencies |

and amino acid deficiency. Drug induced erythroderma in children occurs commonly due to sulfonamides, antimalarials, penicillins, isoniazid, thioacetazone, streptomycin, nonsteroidal anti-inflammatory drugs (NSAIDS), topical tar, homeopathic and ayurvedic medicines, captopril, cinemidine and ampicillin. In neonatal erythroderma, ceftriaxone and vancomycin have been incriminated. Immunodeficiency was the leading cause (30%) of erythroderma in neonates and infants in western studies, as also Omenn's syndrome (erythroderma, failure to thrive, lymphadenopathy and recurrent infections) and graft-versus-host reaction.
2. Urticaria and angioedema

Urticaria and angioedema are common cutaneous vascular reaction patterns. Urticaria is characterized by transient, pruritic, edematous, lightly erythematous papules or wheals lasting less than 24 hours. Angioedema involves deeper subcutaneous structures. Life threatening reactions are associated with angioedema especially when the respiratory mucous membranes are involved leading to laryngeal edema. Severe attacks may be associated with abdominal pain, nausea, vomiting due to intestinal obstruction in children.

3. Bullous disorders

Immunobullous diseases like pemphigus, pemphigoid etc. and hereditary mechanobullous disorders like epidermolysis bullosa (EB) can be disabling and even life-threatening in some cases. EB is divided into three types: EB simplex (EBS), junctional EB (JEB) and dystrophic EB (DEB). Among several subtypes of EB, severe form of EBS Dowling-Meara (EBS-DM), Herlitz-type JEB (JEB-H) and recessive DEB (RDEB) can be lethal in neonatal period.

4. Infections

A) Staphylococcal scalded skin syndrome (Ritter’s disease) Commonly seen in infants and children, this condition is caused by Staphylococcus aureus phage type 71 due to liberation of exotoxin. The clinical features include diffuse erythema, fever, tender skin, large flaccid bullae with clear fluid which rupture soon after being formed. This may lead to extensive loss of the skin surface. The exfoliative toxins spread hematogenously from a localized source causing widespread epidermal damage at distant sites.

B) Neonatal cutaneous infections Neonates, especially premature and low birth weight infants are susceptible to various fatal infections like staphylococcal scalded skin syndrome (SSSS), necrotizing fasciitis, neonatal varicella, neonatal herpes simplex infection (HSV) and cutaneous candidiasis.

Necrotizing fasciitis, primarily a disease of adults and rare in neonates, is characterized by fulminant course. The infection is polymicrobial in 75% of cases usually caused by S. aureus. Immediate surgical intervention with antibiotics is required. Death usually occurs due to septicemic shock, disseminated intravascular coagulation and/or multiple organ failure.

Neonatal varicella, usually transmitted from maternal varicella during last 3 weeks of pregnancy. The manifestation of neonatal varicella during first 10-12 days of life suggests transplacental transmission of the disease. Postnatally acquired neonatal varicella presents after 12 days of life. The severity and mortality of neonatal varicella depends on the day of onset of rash in the mother and neonate.

Neonatal HSV infection is transmitted from mother during intrauterine (5%), peri-partum (85%) and postpartum (10%) periods. Infants born to mothers who have first episode of genital herpes near term are at increased risk of developing neonatal herpes than those born to mothers with recurrent infection. Skin vesicles at or soon after birth are the most common clinical presentation of neonatal HSV infection. Fever and lethargy are common in disseminated and CNS disease and vesicles may not develop.

Candidiasis in newborns occurs in two forms; congenital cutaneous candidiasis (CCC)
acquired in utero and neonatal candidiasis acquired during passage through infected birth canal. CCC classically presents as generalized erythematous macules, papules and/or pustules predominantly over back, extensor extremities, skin folds and almost always involving palms and soles. The diaper area is usually spared and oral mucosa rarely involved. The lesions generally resolve with desquamation within 1-2 weeks.35,36 Neonatal candidiasis manifests after 7 days of life and is localized to oral cavity and diaper area.

Other conditions with infectious etiology which can be fatal include eczema herpeticum, Waterhouse-Friderichsen syndrome, staphylococcal and streptococcal toxic shock syndromes, Rocky Mountain spotted fever, anthrax and ecthyma gangrenosum.

5. Drug reactions

Drugs may cause emergency situations like drug reaction, eosinophilia and systemic symptoms (DRESS) syndrome, anaphylaxis, toxic epidermal necrolysis (TEN) and Stevens-Johnson syndrome (SJS).

Both SJS and TEN are immune complex-mediated blistering conditions of the skin which may result in high morbidity and mortality. Drugs, infections and certain miscellaneous conditions are among the most important implicating factors. It is believed that they both represent different spectra of the same disease with TEN being the most severe form, with epidermal detachment greater than 30% and a mortality of 15-40%, while in SJS, epidermal detachment is less that 10% of body surface area.37,38

6. Connective tissue diseases

Acute lupus erythematosus, dermatomyositis, antiphospholipid antibody syndrome, eosinophilic fasciitis, scleredema, to mention a few, may present as emergency situation.

7. Metabolic conditions

Metabolic and nutritional disorders like multiple carboxylase (holocarboxylase synthetase and biotinidase), essential fatty and amino acid deficiency may be life threatening. Metabolic conditions like multiple carboxylase and essential fatty acid deficiency can present in infancy as erythematous-squamous rash progressing to involve whole body.

8. Miscellaneous

a) Kasabach-Merritt phenomenon (KMP), with mortality of 20-30%, is a clinical syndrome of thrombocytopenic coagulopathy in association with vascular tumor, tufted angioma and Kaposi's hemangioendothelioma, particularly the latter.39,40 It is usually seen in infants less than 3 months of age. It is caused by sequestration of platelets, accumulation of activated coagulation factors and local fibrinolysis in the tumor.

b) Purpura fulminans (PF) is an acute syndrome characterized by rapidly progressive skin necrosis and disseminated intravascular coagulation (DIC).41 Hereditary (congenital) protein C deficiency, an autosomal recessive disorder, manifests at birth. Acute infectious PF in neonates is commonly caused by group B streptococcal septicemia and also Gram negative septicemia.42

c) Kawasaki disease is a systemic vasculitis predominantly affecting younger children less than 4 years of age with peak age of onset of 6
to 11 months. Classical presentation of KD is high grade fever not relieved by antipyretics, generalized erythematous maculopapular rash, bilaterally symmetrical non-pitting edema of hands and feet, fissuring of lips, reddish discoloration of tongue and non-purulent bilateral bulbar conjunctivitis.

d) Sclerema neonatorum regarded as end stage of severe systemic disease, is an uncommon, life-threatening condition, usually of newborns, with a case-fatality rate ranging from 50 to 100%. It is characterized by sudden onset diffuse hardening of skin initially involving lower legs and later spreading to thighs, buttocks, trunk and cheeks. The palms, soles and genitalia are usually spared.

e) Erythromelalgia A condition of painful red extremities with burning associated with vasodilatation of the skin, with attacks lasting for few minutes to several hours, presents as an emergency situation.

f) Other conditions like calciphylaxis, vasculitis etc may present as emergency situations.

9. Emergencies related to sexually transmitted diseases (STDs)

Due to the prevalent misunderstanding and discrimination, the mere suspicion of acquiring STD especially HIV evokes extreme psychological anxiety that can lead to suicidal ideation and worsening illness by delay in logical course of action. Paraphimosis, phimosis, phagedenic ulceration, bubo formation, rupture of dorsal artery of penis etc. are common emergencies. Penicillin therapy in syphilis can cause Jarisch-Herxheimer reaction which can prove fatal.

10. Emergencies related to leprosy

The emergencies associated with leprosy especially acute neuritis of ulnar, common peroneal and facial nerves, eye and testicular involvement etc. due to acute inflammatory episodes (lepra reactions), should be actively looked for and treated promptly to obviate permanent damage. Dapsone syndrome, acute abdomen due to clofazimine and ‘flu’ like syndrome due to rifampicin are serious adverse effects of commonly used anti leprosy drugs.

11. Emergencies related to dermatosurgery procedures

Diagnostic and therapeutic procedures performed in dermatology rarely precipitate a crisis, some of the emergency situations that can arise include:

i) Anaphylaxis is a generalized multiorgan allergic reaction characterized by rapid evolution of cutaneous features like diffuse erythema, pruritus or urticaria, followed by inspiratory stridor, laryngeal edema, bronchospasm, hypotension, cardiac arrhythmias, or hyper peristalsis. Anaphylaxis is a potentially life threatening event. In classic anaphylaxis, the offending antigen binds to immunoglobulin E (IgE) on mast cells and basophils, initiating the release of inflammatory mediators. Anaphylactoid reactions are clinically similar, but are not IgE mediated. Therefore, they are not allergic reactions. They occur by directly stimulating mast cells and basophils, provoking the release of the same mediators as in anaphylaxis. Anaphylactoid reactions are most commonly caused by radiocontrast media, aspirin, non steroidal anti-inflammatory agents, opioids and muscle relaxants.
In dermatological set up, anaphylactic reactions could be due to preoperative antibiotic prophylaxis with penicillin or cephalosporin, local anesthesia infiltration with an ester anesthetic or lidocaine with methylparaben (see lidocaine “allergy” below), bacitracin, neomycin, topical nitrogen mustard, chlorhexidine, and natural rubber latex (surgical gloves). Prompt recognition is the key to anaphylaxis management.

ii) Vasovagal syncope Vasovagal syncope is the most common cause of acute brief unconsciousness. It is far more prevalent than anaphylaxis. There are often no associated cardiac or neurological abnormalities. Emotional stress, acute pain and fear are precipitating factors. The characteristic prodrome includes anxiety, diaphoresis, nausea, tachypnea, tachycardia and/or confusion. The skin becomes pale and cool. Vagal-induced bradycardia in the setting of decreased systemic vascular resistance can initiate collapse. Pseudo-seizure activity can occur. Blood pressure may initially decrease but is restored with recumbence.

iii) Lidocaine allergy Reactions mostly occur to the ester group of anesthetics like procaine, tetracaine, and benzocaine, derivatives of paraaminobenzoic acid (PABA), an established allergen. True allergic reactions to pure lidocaine are extremely rare. Lidocaine belongs to the amide class of anesthetics, which do not cross-react with ester anesthetics. Methyl- and propyl paraben, sulphite preservatives added to lidocaine bottles cross reaction with PABA causing type IV (delayed type) sensitivity to lidocaine, 2 days following exposure.

Toxic reactions to lidocaine resulting from overdosage (central nervous system or myocardial depression or excitation, perioral numbness, nausea, seizures, coma) and vasovagal reactions should be distinguished to avoid confusion with lidocaine allergy.

Anxiety regarding the use of needles and/or the effects of frequently added epinephrine in lidocaine vials can lead to palpitations, panic attacks and vasovagal events that the patient may long remember as an allergic reaction. Patch testing and intradermal challenge assist in the evaluation of type IV sensitivity.

iv) Acute stroke A stroke occurs when the blood supply to a portion of the brain is disrupted, resulting in a sudden neurologic deficit from inadequate oxygen delivery. Strokes may be ischemic (85%) or hemorrhagic. The dermatologist’s role chiefly concerns the detection phase of a sudden neurologic deficit. The time critical nature of stroke management means that patient should be transported rapidly for specialized care. In the interim, the dermatologist should attend to the ABCs (airway, breathing, circulation) of basic life support as needed.

v) Status epilepticus Dermatologists encounter patients whose cutaneous disease has potential epileptic manifestations such as tuberous sclerosis, neurofibromatosis, Sturge-Weber syndrome, lupus erythematosus. There is a risk of office seizures and also status epilepticus. The dermatologist can provide basic support like maintenance of a patent airway, deliver oxygen, monitor vitals, place intravenous line till patient gets specialized treatment.

vi) Electrosurgery and pacemakers/defibrillators The increasing prevalence of implantable pacemakers and defibrillators has raised questions regarding the safety during electrosurgical procedures. Electrodesposition, fulguration, coagulation and
cutting current involve a potentially significant transfer of electrical activity to patients with pacemakers, especially ventricular inhibited and ventricular triggered pacemaker, leading to Brady- and tachyarrythmias respectively.

Implantable cardioverter-defibrillators (ICDs) are implantable electronic devices that sense cardiac electrical activity and terminate ventricular fibrillation and ventricular tachyarrhythmias. Electromagnetic interference could potentially damage/deactivate the ICD device or trigger the device to deliver a defibrillatory discharge. Electrosurgery should be avoided in pacemaker patients if an alternative, equally effective modality existed. Prior consultation with a cardiologist, emergency backup, short bursts of electrosurgery (under 5 seconds) and good grounding away from the heart are all recommended.

Besides these, therapeutic procedures act as stress and may trigger complications of systemic diseases like hypertension (stroke, cardiac arrest), diabetes (ketoacidosis) etc.

**Acute skin failure and its consequences**

Some of the emergency dermatological conditions cause structural and functional alterations in the skin which leads to failure of skin to perform its multiple functions, which can subsequently lead to acute failure of heart, lung, kidney and other organs. Destruction of stratum corneum, the layer mainly responsible for the barrier function of the skin, can cause up to 40 times increase in fluid loss. 50% body surface area (BSA) involvement leads to daily fluid loss of up to 4-5 liters. Loss of proteins (40 gm/L), Na (120-150 mmol/L), Cl⁻ (10-90 mmol/L) and K⁺ (5-10 mmol/L) in the bullous fluid leads to decrease in intravascular volume. The resultant decrease in urinary output and increased blood nitrogen can lead to renal failure unless treated energetically. Damaged skin and its exudates along with altered immunological function support growth of a wide spectrum of endogenous and exogenous organisms leading to systemic infection, severe sepsis and shock. Impaired thermoregulation can cause either hyper or hypothermia depending on the surrounding environment. Hypercatabolic state increases energy expenditure by 2-4 times. Loss of proteins in the exudates leads to hypoalbuminemia. Inhibition of insulin secretion and insulin resistance lead to hyperglycemia and glycosuria, which cause amino acid breakdown leading to further worsening of hypercatabolic state. Increased cutaneous blood flow nearly doubles the cardiac output and may prove fatal, particularly in the elderly and in those with compromised cardiac reserve.

**Management of acute skin failure: need of dermatological intensive care unit**

Prompt initiation of appropriate treatment on the lines of a 100% burns patient and excellent double barrier nursing care are the twin principles of management that can salvage many lives. Management in dermatological ICU set up is must, though it is a team work which requires support of other health professionals as well. Management is being described under various headings:

**i) Cleaning measures** Barrier nursing is the key point in the management. Proper hand washing, gloves wear by doctors, nurses and attendants, floor cleaning, linen care, strict sterilization measures, separation of dirty and clean utility areas can go a long way in preventing transmission of cross infection.
ii) Proper nursing  Nursing in a room at temperature of 30-32°C, in an air fluidized bed is helpful. Care of mucous membranes like eyes, nose, mouth, genitals etc. is essential to prevent complications like corneal scarring, synechie, phimosis, meatal stricture, thus decreasing morbidity. Regular change of posture to prevent bed sores, physiotherapy and psychological counseling of patient and relatives are vital. Help from other specialists can be taken.

iii) Monitoring  Heart rate, pulse rate, urinary volume (50-100ml/hr) should be monitored hourly and urinary osmolarity, glycosuria, temperature and gastric contents monitored 3-4 hourly. Any change in extent of skin lesions and body weight should be noted daily along with calculation of fluid loss.

iv) Hemodynamic and electrolyte homeostasis  Correction and maintenance of hemodynamic and electrolyte equilibrium by fluid and electrolyte administration is of prime importance. Fluid requirement during first 24 hours is isotonic saline 0.7ml/ kg/ % of body surface area (BSA) affected and human albumin 1ml/ kg/ % BSA. Potassium phosphate is added to IV fluids to prevent insulin resistance. About 1500ml of nasogastric feed can be given in addition on first day. Subsequently depending on the progress, oral feeds are increased and IV fluids are reduced gradually.

v) Nutritional support  Aggressive nutritional support is required to compensate the hypercatabolic state and to promote tissue healing. Energy requirement in adults is 1500-2000 Kcal in first 24 hrs, with an increment of 500 Kcal daily up to 3500-4000 Kcal/day. Protein intake of 2-3 gm/ kg/ day (3-4 gm/kg in children) should help in faster healing. Micronutrient supplementation is also important.

vi) Topical medications and dressings  Topical antiseptics like silver sulphadiazine (contraindicated in patients sensitive to sulpha drugs) should be applied after proper bath/ soaks with potassium permanganate solution. Topical application of wet dressings and bland emollients such as petrolatum or white soft paraffin helps in maintenance of barrier function of stratum corneum.

vii) Investigations  Daily arterial blood gas analysis, complete blood count, blood urea, creatinine, glucose, electrolytes, albumin, LFT, complete urine examination, ECG and chest radiograph are essential. Culture from skin lesions and venous line is desirable for appropriate antibiotic selection. This may or may not be repeated depending on the clinical and microbiological response to antibiotic therapy.

viii) Sepsis screen  Pus, blood and urine cultures should be sent on every 3rd day to know antimicrobial and drug sensitivity pattern more so for deadly nosocomial infections.

ix) Systemic medication  Judicious use of antibiotics/ antimicrobials is a must to avoid strain selection, infection and drug reactions. Cover for secondary candidiasis is also required. Sudden rise or fall of temperature, deterioration of consciousness, oliguria, accelerated pulse, tachypnoea, increase in insulin requirement and gastric residual volume indicate need for antibiotics in absence of pus/blood culture results, otherwise wait for culture/sensitivity. We should consider judicious use of NSAIDs for adequate pain relief, especially in SJS/TEN, where opioids are the better choice.

x) Specific therapy  Specific therapy depends on the underlying cause.
ICU in a skin department has now been recognized as a necessity due to a large number of extensive skin diseases eventuating into potentially fatal syndrome of ‘acute skin failure’. Setting up dermatological ICU may be easier and different than that of conventional ICU. In a DICU, mainly acute skin failure cases will be managed. It should be properly designed by a multidisciplinary team consisting of ICU medical director, ICU nurse manager, architect experienced in ICU designing and engineering staff. Some of the characteristics of an ideal DICU, like other ICU, are:

- The heating, ventilation and air conditioning system should be properly designed to maintain indoor air temperature and humidity at comfortable level, control odor, remove contaminated air, facilitate air handling in order to minimize the transmission of air borne pathogens.
- There should be separate areas to deal with infectious and non-infectious conditions.
- Air cleaning by filtration and ultraviolet irradiation.
- Proper floor plan and design with separate patient area, properly located nursing station (preferably central), storage area, reception area, specialized procedure room, staff lounge, visitor/waiting room.
- Supply and professional traffic should be separated from public/ visitor traffic.
- There should be proper noise control measures; adequately visible patient cabinets; uninterrupt power, water, oxygen, compressed air, lighting services and well developed intercommunication system.
- There should be 24-hr clinical laboratory services and physician on call services.

In conclusion, there is need of a separate ICU attached to a dermatology department to deal with expertise and urgency all types of dermatological and dermatosurgical practice related emergency situations, especially acute skin failure, in order to decrease mortality and morbidity. The DICU need not be as advanced and sophisticated as cardiac, surgical or neonatal ICU. DICU should be equipped to such an extent that it provides initial, immediate and necessary support.

References


