Chapter 15
DERMATOLOGICAL EMERGENCIES

Learning Objectives:

- Develop clinical approach and evaluation of Dermatological emergencies.
- List key features of these conditions.
- Describe initial steps in the management.
- Learn the list of common drugs responsible for severe cutaneous drug reaction.
- Recognize when to refer a patient a dermatologist

INTRODUCTION

Dermatological emergences are not very common but they have high fatality rates if not recognized and treated early. Acute skin failure can lead to multiple system failure. Patients with extensive skin involvement need a high dependency management similar to burn patients. They need to be treated in aseptic environment with multidisciplinary consultation and life support facilities. Medical officers are expected to recognize these emergencies, provide supportive therapy and refer where relevant.

ACUTE SKIN FAILURE

Shuster was the first Dermatologist who described the systemic manifestation of severe cutaneous disorders in 1967. Skin being the largest organ in body has various functions, besides being the mechanical barrier, it also has complex roles like immunological and endocrine and has a major role in maintaining body homeostasis. Alteration of any of this function can have severe effect on internal organs especially renal.

Consequences of acute skin failure

- Fluid and electrolyte imbalance.
- Protein and calorie loss.
- Impaired thermoregulation.
- Alteration in Immunological function.
- Infection and sepsis.
- Endocrine - Vitamin D deficiency.
- Systemic- malabsorption, cardiac failure in people with poor cardiac reserve and renal failure.

Management

- Ensure oral intake, if poor NG tube feeding.
- Large bore IV cannula through unaffected skin.
- Maintain input output chart.
- Vital charting preferably by pulse oximeter.
- Investigation – CBC, CRP, RFT, LFT, FBS, S. proteins and Pus swab from multiple sites.
- IV fluid administration according to fluid loss. In first 24 hours IV fluid may be given according to extent of skin involvement.
- Electrolyte imbalance must be corrected.
- Temperature control by nursing in warm room to prevent shivering and calorie loss and use humidifiers or maintain a boiling kettle near the room.
- IV antibiotic if infection suspected. Some prefer broad spectrum IV antibiotic.
- Gentle handling of skin, strict barrier nursing.
- If erosions- wash with normal saline and apply Vaseline gauze dressing.
- Once patient is stabilized underlying cause needs to be treated. If drugs suspected stop the drug immediately.
- Refer to dermatologist for further management.

SEVERE CUTANEOUS ADVERSE DRUG REACTION (SCAR)

Steven Johnson and Toxic Epidermal Necrosis (Lyell Syndrome)

Cutaneous adverse reaction can occur at any age group and risk increases with age. Immunosuppressed patients and patient with HIV are more prone to develop SCAR. Steven

Johnson syndrome (SJS) and Toxic epidermal necrolysis (TEN) are severe muco-cutaneous drug reaction characterized by extensive skin and mucosal necrosis presenting as erythema, blisters and extensive skin sloughing. SJS and TEN are two spectrums of same disease differing in severity and mortality. SJS mortality varies from 5 to 12 % whereas TEN its more than 20 %. Increasing age, comorbid conditions and increasing extend of skin involvement is associated with poor prognosis.

Common drugs causing SJS and TEN:

There are many drugs that are implicated. There is a strong genetic susceptibility.
The common drugs that are responsible are:

- Sulfa antibiotics, Sulfasalazine
- Allopurinol
- Tetracyclines
- Anti-convulsant (carbamazepine, lamotrigine, phenobarbital, phenytoin).
- NSAIDS
- Nevirapine
- Thioacetazone

Rarely infection can cause, especially in children. *Mycoplasma pneumonia* has been implicated.

Clinical feature:

- Latency period from starting of drug to appearance of reaction is typically 7–10 days, but ranges from 5 to 28 days.
- Malaise, fever, prostration and upper respiratory tract symptom.
- Rash typically occurs on face and chest first and rapidly spread to other areas including palms and soles.
- Initial lesion is dusky to purpuric macules or atypical targets, which form flaccid blisters. Lesions rapidly progress and coalesces forming sheets of erythema.
- Skin lesions are painful.
- Nikolsky's sign is positive- gentle lateral pressure causes lesional intact epidermis to slide over the dermis indicating epidermal necrosis.
- Mucosal involvement can sometimes precede skin lesions.
- Eyes, mouth, nose and genitalia is usually an early feature and leads to painful erosive and hemorrhagic mucositis.
- Based on extend of skin lesion disease can be classified as:
    - SJS: BSA < 10%
    - SJS/TEN overlap: BSA 10-30%
    - TEN: BSA > 30%

Risk Assessment:

Table 15.1 SCORTEN. It represents prognostic scoring for patients with TEN. A SCORTEN of ≥ 5 has mortality rate of 90%.

Prognostic Factors		Points
Age	> 40 years	1
Heart rate	> 120 bpm	1
Cancer or hematological malignancy		1
BSA on day 1	> 10%	1

Serum urea	> 10mmol/l	1
Serum bicarbonate	< 20 mmol/l	1
Serum glucose	> 14 mmol/l	1

Differential Diagnosis:

- Erythema multiforme major.
- Pemphigus vulgaris.
- Bullous pemphigoid.
- Paraneoplastic pemphigus.
- Bullous lupus erythematosus.
- Acute bullous acute graft-versus-host disease.
- Staphylococcal scalded skin syndrome.

Complications:

Acute-

- Infection and sepsis commonly by Staphylococcus aureus and sometimes Pseudomonas aeruginosa.
- Acute skin failure and its complication.
- Hematological - anemia and leucopenia.
- Abnormal liver function is typical during the early stage of the disease.
- Epithelial necrolysis may occur in the bronchi during the acute phase of SJS/TEN resulting in bronchial erosions and airway obstruction.

Late complication-

- Corneal damage.
- Oral – Sicca syndrome, oral adhesions.
- GU –adhesions – vaginal, introital, urethral.

Management:

First line:

- Withdraw the culprit drug.
- Multi-disciplinary approach.
- If epidermal loss is >10% BSA will require treatment in a specialist unit with supportive care package, with particular attention to:
  - Heated environment
  - Fluid replacement
  - Nutritional regimen
  - Analgesia
  - Preventing/treating infection
- Analgesics- simple analgesia or may require opioids. Morphine 0.1 mg per kg 10 to 20 minutes before dressing can be given.
- Skin care- daily bathing; normal saline soaks may be soothing. Vaseline gauze dressing. Aspirate large blister keep the roof intact.
- Mucosal care-
  - Regular examination.
  - Oral hygiene/ mouthwash.
  - Remove oral and nasal debris.
  - Artificial tears for eyes, antibiotic drops.

  - Dressing and lubrication for vaginal erosions.
- Prompt intubation and ventilator support at the earliest sign of ARD.
- Gastric prophylaxis – PPI.
- Prevent DVT. May need LMW heparin.

Second line:
In the early stages of the acute phase consider using:
- IVIG (0.5–1 g/kg daily for 3–4 consecutive days), OR
- Systemic corticosteroid (e.g. prednisolone 0.5–1 mg/kg daily for 10 days, and tapered; or IV methylprednisolone500 mg on 3 consecutive days), OR
- Ciclosporin (3 or 4 mg/kg/day in divided doses for10 days, and tapered).

Offending drug should never be restarted. Remember that anticonvulsants can cross react (phenytoin, carbamazepine and phenobarbitone).

Drug Reaction with Eosinophilia and Systemic Symptoms (DRESS)

DRESS is multisystem idiosyncratic drug hypersensitivity reaction characterized by cutaneous eruption, eosinophilia and organ involvement typically hepatic. Genetic predisposition and viral reactivation (HHV6 and 7) have been postulated in patho-mechanism.

Causative Drugs:
- Allopurinol.
- Antiepileptics– carbamazepine, phenytoin, lamotrigine.
- Antibiotics– vancomycin, amoxicillin, minocycline, piperacillin-tazobactam.
- Sulpha drugs– sulphasalazine, dapsone, sulphadiazine.
- Furosemide.
- Omeprazole.
- Ibuprofen.

Clinical features:
- Onset typically from 7 to 28 days from start of the drug as compared to exanthematous drug reaction which is more common and onset 7 to 10 day.
- Patient present with fever, systemic upset and lymphadenopathy.
- Skin – Typically starts on face and trunk with urticated popular exanthema, morbilliform eruption. It may also present vesicles, tense bullae induced by dermal edema follicular as well as non-follicular pustules, erythroderma and purpuric lesions.
- Mucous membrane- cheilitis. Frank mucosal involvement is not common.
- Systemic:
  - Eosinophilia rarely pancytopenia.
  - Hepatitis, can lead to acute fulminant hepatitis.
  - Renal in about 10 %, Proteinuria, hematuria and urinary eosinophil (Interstitial nephritis).
  - Pericarditis, myocarditis.
  - Hypo- and hyperthyroidism during convalescent phase.
- RegiSCAR DRESS scoring system.

Complications:
- Fulminant hepatic failure.
- Interstitial nephritis.

- Autoimmune phenomenon following DRESS – LE, Thyroid.
- Erythroderma.

Differential Diagnosis:

- Sepsis.
- SJS/TEN.
- Acute generalized exanthematous pustulosis.

Investigation:

- Routine blood test/ PBS for atypical lymphocytes, LFT, RFT, Viral serology, ECG, Chest X – Ray, TFT.

Management:

- Supportive management.
- Withdraw the offending drug. Establish a drug timeline, if offending drug not known due to multiple drugs, then withdraw all medication.
- Start oral steroid prednisolone 1 mg per kg or IV methylprednisolone 1 gm for 3 consecutive days.
- If systemic steroid fails - Ciclosporin, cyclophosphamide, Rituximab, ganciclovir. If severe hepatitis – N-acetylcysteine.
- Offending drug should never be restarted.

Acute Generalized Exanthematous pustulosis (AGEP)

Severe cutaneous adverse reaction that is usually self-limiting but recognition is important to withdraw the offending drug and prevent systemic complication. Patient presents with fever and characteristic non-follicular pustules, which coalesce and form sheets of pus.

Drugs Associated with AGEP:

- Aminopenicillins.
- Quinolones.
- Chloroquine and hydroxychloroquine.
- Sulphonamides.
- Terbinafine.
- Diltiazem.
- Sometimes it can follow infection – Mycoplasma, Coxsackie virus, and parvovirus, spider bites.

Clinical features:

- Short latency period, can occur 2 to 5 days after the drug intake.
- Initially there is skin itching or burning followed by multiple non- follicular pustules typically on flexures but other areas can also be affected. There is background edema and erythema.
- Fever and mild renal, hepatic and pulmonary involvement.
- Self-limiting.
- Lesions resolve with superficial desquamation.

Differential diagnosis:

- Acute generalized pustular psoriasis.
- Sub corneal pustular dermatosis.
- Candida infection.

- Bacterial Infection.

Investigation:

- CBC- leukocytosis with neutrophilia, sometimes eosinophilia.
- CRP – to exclude infection.
- LFT, RFT, S. calcium.

Management:

- Withdraw the offending drug.
- Depending on severity topical steroid or oral steroid can be started.
- Offending drug should be avoided in future.

INFLAMMATORY CONDITIONS

Erythroderma

Erythroderma denotes inflammatory skin condition affecting more than 80- 90% body surface area. It is not a diagnosis but a manifestation of an underlying disease. Students are expected to recognize the morphology, learn the underlying causes and be aware of morbidity and mortality associated with erythroderma.

Clinical features:

- Generalized erythema of skin affecting 80 to 90% of BSA. When associated with scaling it is called exfoliative dermatitis.
- Fever with chills and malaise.
- Pruritus.
- Hepatomegaly, lymphadenopathy and peripheral edema.

Causes:

- Eczema – Atopic dermatitis, Allergic contact dermatitis, Seborrheic dermatitis.
- Psoriasis.
- Pityriasis rubra pilaris.
- Lymphoma and leukemias (CTCL, HL, NHL)
- Drugs (phenylbutazone, phenytoin, carbamazepine, gold salts, lithium, cimetidine).
- Hereditary disorders (ichthyosiform erythroderma).
- Pemphigus foliaceus.
- Other skin diseases (lichen planus, dermatophytosis, crusted scabies, dermatomyositis).
- Idiopathic.

Complication:

- Can be potentially fatal, mortality ranges from 16 to 64% and mostly due to cutaneous infection, sepsis and pulmonary Infection.
- Sepsis.
- Hypothermia.
- Hypo-proteinuria.
- High output cardiac failure (complication is more common in elderly)
- Cutaneous long-term complication – diffuse alopecia, keratoderma, nail dystrophy, ectropion.

Investigation:

- CBC, ESR, CRP, PBS, LFT, RFT/SE, S. proteins.
- Multiple skin biopsies.

- Lymph node biopsy if significant or malignancy suspected.

Management:

Initial management-

- Acute disease and acute flare up require hospital admission.
- Withdraw or switch medications that may be implicated as a cause.
- Monitor and correct fluid and electrolyte loss and control temperature instability.
- Treat any secondary infection.
- Nutrition and vitamin supplementation.
- Sedative antihistamine.
- Frequent application of greasy emollients.

Second line-

- Systemic therapy dependent on underlying cause.

Acute Generalized Pustular Psoriasis (Von Zumbusch)

Acute exaggerated form of psoriasis that affects all age groups and may be the first presentation of psoriasis or as a complication in patient with preexisting psoriatic plaques. There are many forms and it can be associated with multiple systemic manifestations.

Precipitating factors:

- Infection – bacterial and fungal
- Stress
- Pregnancy
- Hypocalcemia
- Irritating topical therapy- Coal tar, Dithranol
- Withdraw of systemic steroids, Cyclosporine

Clinical features:

- Fever with malaise. May have a prodromal of burning tender skin.
- Pre-existing plaques become redder and develop small pustules. Periphery of plaques can be studded with pustules.
- Can present for the first time with erythema and pustules.
- Pustules can appear in other areas of body including flexures and genital and form lakes of pus.
- Geographic tongue.
- Nails – thickened and sub-ungual pus.

Complication:

- Severe Hypoalbuminemia
- Hypocalcemia
- Acute tubular necrosis
- Malabsorption

Differential Diagnosis:

- Systemic infection
- (AGEP)

Management:

Long-term prognosis of the disease is not known but patient can die in absence of treatment. May lead to erythrodermic psoriasis.

- Investigation- CBC initially absolute lymphopenia later leukocytosis, raised ESR, Low albumin and serum calcium. FBS, RFT, FBS, LFT, Viral markers and exclude TB and other infection for systemic therapy. Skin biopsy to confirm disease.
- Rule out and manage accordingly any precipitating factors e.g. withdraw any irritating treatment.
- Symptomatic conservative treatment.
- Only bland emollients for the skin after bathing.
- Systemic therapy-
  - Acitretin – best evidence in recent studies. Contraindicated in pregnancy.
  - Ciclosporin - fast acting.
  - Methotrexate – easily available. Oral absorption may be erratic may have to give parental.
  - Prednisolone (special circumstances only)
  - Others – Biologics.

IMMUNE BULLOUS DISORDER

There are various causes of blisters in skin. Blistering disease can be broadly divided into genetic and immune- bullous disorders. Students should be able to identify severe blistering diseases, provide initial management and know when to refer to dermatologist.

Pemphigus Vulgaris

Pemphigus vulgaris is an autoimmune disorder, which present with skin and mucosal involvement and can be life threatening. Autoantibodies are directed against attachment complex in epidermis known as desmosomes. Breaks in these attachments that keeps the keratinocytes together result in blisters. Other autoimmune disorders can be associated particularly thyroid and rheumatoid arthritis. Morbidity and mortality are high due to inadequate treatment, chronic nature of the disease and medication side effects.

Clinical features:

Pemphigus can present at any age but it's more common in elderly. Severity can vary from patient to patient. Severe disease can present with extensive skin erosions with mucosal involvement. Distinct subgroups identified. Para-neoplastic pemphigus associated with internal malignancy and often have poor prognosis.

- Flaccid blisters with clear fluid that rupture and form painful erosion.
- Predilection for scalp, face, neck, chest and back.
- Nikolsky's sign is positive.
- Mucosal erosions, can involve oral, nasopharynx, larynx, esophagus, conjunctiva, vagina and cervix.
- Healing occurs without scarring.

Complication:

- Acute skin failure.
- Infection, sepsis.
- Drug side effects.

Differential diagnosis:

- Bullous pemphigoid - more common in elderly. Present with tense bullae over a normal skin or urticarial plaque. Mucosal involvement may or may not be present.
- Benign familial pemphigus.

Management:

- Routine investigation with special attention to rule out infection.
- Systemic steroid is main stay of treatment preferably with a steroid sparring agent.
- Manage as for acute skin failure.
- Oral and eye care.
- Skin:
    - Daily bathing. Soak with normal saline, it will be soothing.
    - Aspirate large bullae and leave the roof intact as a natural dressing.
    - Cover large erosion with sterile paraffin gauze.
    - Loose cotton clothing.
    - Avoid adhesive tapes; instead roller bandages can be used.
- High protein diet and vitamin supplementation.
- Systemic antibiotic should be guided by culture and sensitivity report.
- Drugs- Prednisolone 0.5 to 1 mg per kg IV methyl prednisolone (1gm) or IV dexamethasone pulse (100mg)
- Other drugs- Azathioprine, Cyclophosphamide, MMF, IV Ig and Rituximab.

DRUG INDUCED ACUTE URTICARIA or ANGIOEDEMA

Acute spontaneous urticarial can be allergic (food, drugs, following blood transfusion, bites and stings), non-allergic and due to infection. Allergic urticarial can be sometimes severe. Dug induced urticaria may be presenting symptom of anaphylaxis or acute serum sickness. ACEI can sometimes cause life-threatening angioedema.

Clinical Presentation:

- Temporal relation to drug.
- In most patients with ACEI induced angioedema, it occurs during first week of therapy but it can occur years later too.
- ACEI induced angioedema is usually without wheals. They present with painful swelling affecting head, neck and mucosa. Laryngeal edema can be life threatening. It is due to reduced bradykinin metabolism.
- Differentiate from hereditary angioedema.

Causes of acute Urticaria:

- Idiopathic.
- Infections-Viral, e.g. upper respiratory tract infections, hepatitis B and C. Bacterial, e.g. Streptococcus pyogenes.
- Parasitic.
- Foods, e.g. cow's milk, hen's egg, nuts, seeds
- Drugs, e.g. β-lactam antibiotics
- Stings, e.g. bee, wasp venoms
- Blood products
- Vaccines
- Contactant, e.g. latex

Investigation:

- Allergen specific IgE can be tested, e.g. peanut allergy which can be life threatening.
- If allergen not known investigation usually not useful.
- Prick test can be risky.

Management:

- Treat the cause if known.
- Immediate withdrawal of drug.
- If Urticaria with angioedema– 2nd generation H1 antihistamine, systemic steroids. Epinephrine as for anaphylaxis.
- ACEI induced angioedema is usually refractory to antihistamine, steroids and epinephrine.
- FFP during acute attack, Icatibant (bradykinin 2 receptor antagonist)
- Other ACEI should be avoided (class effect), ARBS can sometimes cause.

When to refer to Dermatologist?

- Individual lesion last more than 48 hours and/or lesions are painful and burning rather than itchy or presence of petechial lesions.
- Lesions resolve with pigmentation.
- Poor response to antihistamines.
- Associated systemic symptoms (More likely to be Urticarial Vasculitis).

INFECTIONS

Necrotizing subcutaneous Infection

These are group of infection with primary focus of infection on dermis, adipose and subcutaneous fascia. Hallmark is extensive necrosis with accompanying cellulitis. There can be severe local and systemic complication and can be life threatening. **Necrotizing fasciitis** is discussed here.

Causes:

- Predisposing factors- surgical procedures and diabetes.
- Two groups of organisms:
  - *β- Hemolytic* group A *Streptococci*
  - Mixed organism– *Staphylococcus aureus, Aeromonas hydrophilia*, *Vibrio vulnificus* and anaerobes.
  - Sometimes cultures may not grow any organism.

Clinical Presentation:

- Extremities and perineum are commonly affected sites.
- Patient present with erythematous to dusky hot tender swelling.
- Woody induration extending beyond the area of apparent involvement.
- Pain out of proportion to physical finding (early).
- Develop bullae and necrosis and may become anesthetic.
- Fever and severe constitutional symptoms. As disease progresses may go into septicemia.

Complication:

- Extensive local tissue destruction, deeper infection (myositis, polymyositis).
- Extensive skin sloughing that can be mutilating.
- Septicemia with multi-organ failure.

Investigation:

- Pus swabs are often negative.
- Diagnosis mainly clinical.
- Surgical exploration for diagnosis and management.
- MRI or CT- reveals edema fluid along fascial planes.

Management:

- Acute emergency requires ICU management.
- Surgical debridement of the affected tissue including surrounding area.
- Surgical debridement may need to be repeated every 2 to 3 days.
- Antibiotic – Broad-spectrum antimicrobial therapy (gram positive, negative and anaerobes) or according to culture report.

Meningococcal Infection

Acute meningococcal septicemia is a severe illness caused by *Neisseria meningitides.* Although it is not a dermatological disease, the rash is characteristic and early recognition and institution of adequate treatment may be lifesaving. Outbreaks can occur in communities and it's often through naso-pharynx carriage.
90% of disease caused by serogroups A, B, C, W-135, X and Y.

Clinical features:

- More common in winter months.
- Children less than 10 years are more commonly affected.
- Present with fever, malaise, headache and joint pain with or without features of meningismus and altered sensorium.
- Purpuric rash on limbs and trunk. Starts as a small pink macules or papules that can occur anywhere including palms and soles. May form petechial rash.
- These lesions become larger with irregular border (angulated margins), can become hemorrhagic with bullous formation or develop areas of necrosis.
- "Stellate purpura with typical gunmetal gray central hue"

Complications:

- Meningitis, encephalitis
- Sepsis, DIC
- Myocardial dysfunction
- Adrenal hemorrhage

Differential Diagnosis:

- Bacterial sepsis
- Systemic vasculitis
- Drug hypersensitivity reaction
- Endocarditis
- Acute viral hemorrhagic fevers
- Rocky mountain spotted fever

Investigation:

- Polymorphonuclear leukocytosis with band forms, thrombocytopenia (DIC)
- Raised ESR, CRP

- Proteinuria, hematuria
- Coagulation profile if DIC suspected
- Blood cultures
- Gram stain of aspirate from skin lesion (positive in 50 to 80%)
- Diagnosis confirmed by isolation of organism from blood or CSF or by PCR.

Management:

- May need ICU management.
- Circulatory support and intravenous fluids.
- Intravenous benzylpenicillin in high doses is the first treatment of choice.
- Ceftriaxone or cefotaxime are suitable alternatives.

Prophylaxis:

- Rifampicin for 2 days is recommended as prophylaxis for close family contacts, with clinical monitoring.
- Vaccines are available and given in high-risk cases.

TOXIN MEDIATED

Toxic Shock Syndrome

TSS is a multisystem disorder caused by staphylococcal or streptococcal exotoxin which as a super antigen. Menstrual TSS is rare now but usually associated with staph. Non-menstrual type may be undiagnosed. Streptococcal TSS is associated with invasive soft tissue infection.

Causes:

Toxic shock syndrome toxin 1 (pyrogenic exotoxin C) is the main toxin and more recently, staphylococcal enterotoxin B. It can also be associated with severe infections with Streptococcus pyogenes (scarlet fever toxin A).

Clinical features:

- Disease is rare and certain infection like cellulitis, underlying HIV or internal malignancy, alcohol abuse and diabetes are associated with an increased risk of TSS.
- Acute onset fever and rash. Vomiting and diarrhea are common.
- Circulatory shock (which does not respond to intravenous fluid replacement) and acute renal impairment frequently coexists.
- Multi-organ failure follows soon afterwards.
- For diagnosis at least three systems need to be involved (gastrointestinal, renal, hepatic, central nervous system, muscular, hematological)
- Skin rash may be the presenting feature or may develop within the first day. A widespread non-itchy macular erythema, or sometimes scarlatiniform and papulopustular eruptions can occur.
- Edema of the hands and feet.
- There is generalized mucous membrane erythema.
- Desquamation occurs 10–21 days after onset, and initially it may be only fingertips.

Complications:

- Septicemia, shock, DIC
- ARDS
- ARF and other organ failure

Differential diagnosis:

- Septic shock and other infections
- Adult onset Kawasaki

Investigation:

- Clinical diagnosis.
- Blood cultures- several sets, swabs from wounds, vagina (menstruating female or postpartum)
- Routine blood test.

Management:

- Hemodynamic resuscitation.
- Actively look for cause.
- Antibiotic- IV clindamycin +/– benzylpenicillin or vancomycin.
- For severe cases- intravenous immunoglobulin (initial dose 2 g/kg, then 4 days of 0.4 g/kg).

Staphylococcal Scalded Skin Syndrome (SSSS)

Toxin mediated disease that presents with tender erythematous skin followed by skin peeling. First described in children but sometimes adults can also be affected.

Causes:

- Exfoliative toxin (ETA and ETB) produced by Staphylococcal aureus phage group II.
- It can follow trivial infection such as impetigo and throat or GI infection.
- Toxin cleaves desmoglien-1 (skin adhesion molecule)
- Predisposing factors – children less than 6 years, neonates, renal impairment and Immunosuppression.

Clinical features:

- Child presents with fever, irritability, diffuse erythema and rash.
- Skin tenderness.
- Can form flaccid bullae, which rupture easily to leave raw tender area. Nikolsky's sign is positive. Paper like wrinkling of skin.
- Perioral radial fissures and sparing of mucosa.

Complications:

- Can be fatal in premature infants.
- Secondary gram-negative infection *(P. Aeruginosa*) and septicemia.

Differential diagnosis:

- TEN
- Hereditary mechano -bullous disorders

Investigation:

- Diagnosis is clinical mainly
- Swabs from suspected areas, blood cultures
- Tzanck smear from freshly denuded area – acantholytic cells without inflammatory cells.
- Complete blood test.

Management:

- Mortality rate is higher in adult (around 60%), in children it's about 4 %. Children usually recover within 7 days if antibiotic started early.

- Antibiotic –
  - IV penicillinase-resistant antibiotics such as flucloxacillin, should be used as first line therapy.
  - First generation cephalosporins, penicillin G or erythromycin– if susceptible strain of S. aureus on culture.
  - Supportive therapy if features of skin failure (especially children).

NEONATAL INFECTION

Recognition of these infections is important so that early treatment can be started. Sometimes it may be confused with transient neonatal dermatosis, which often requires no treatment.

Neonatal HSV

Herpes simplex infection of babies less than 28 days of life. Risk is highest with primary genital HSV infection in mother. Outcome depends on which trimester mother gets the infection. Maternal infection one week before and 10 days after delivery has the highest risk to the baby.

Early Intrauterine infection can result in fetal demise; nearly 100% of congenital HSV survivors have significant neurologic delay.

Three distinct patterns of transmission:

- Intrauterine, Perinatal and Post-natal.

Clinical features:

- 30% have symptoms at birth; the rest appear in the first 6 weeks of life.
- Characteristic patterns:
  a. Skin, mouth and eye lesions (SEM)– vesicles, erosions or ulcers typically on scalp or presenting part; oral vesicles, erosions or ulcers; conjunctivitis or keratitis; CNS involvement symptoms are often delayed or absent.
  b. CNS disease – cutaneous lesions, focal encephalitis or meningoencephalitis with irritability, lethargy, fever or seizures.
  c. Disseminated disease – cutaneous lesions, affects liver, adrenals, lung and brain with symptoms suggestive of neonatal sepsis.

Differential Diagnosis:

- Other congenital infection
- Erythema toxicum neonatorum
- Transient neonatal pustular melanosis
- Langerhans cell histiocytosis

Investigation:

- Viral cultures from skin and oral mucosal lesions, nasopharynx, conjunctivae, urine, blood and cerebrospinal fluid (CSF).
- Tzanck smear of vesicles: multinucleated epithelial giant cells.
- Serology for HSV antibodies: rising IgG titers are sensitive and specific for infection, whereas IgM titers are not sensitive in the newborn.
- CSF: lymphocytic pleocytosis, elevated protein and red blood cell count indicative of CNS involvement.
- Imaging: CT or magnetic resonance imaging (MRI) of the brain if indicated.

Management:

- Active primary herpes infection in the mother at the time of delivery, Caesarean section is indicated and prophylactic acyclovir for the neonate.
- Primary genital herpes during the third trimester - oral acyclovir should be started for mother although it is unclear whether this reduces the risk of neonatal herpes.
- For a baby born to a mother with a previous history of genital herpes but no active lesions during pregnancy or at delivery - the baby should be monitored and tested for the presence of herpes on the skin, since asymptomatic shedding may be a route for infection
- Rapid treatment of neonatal herpes, even if confined to skin and mucosae, is essential to reduce the risk of progressive spread of infection.
- Neonatal herpes is treated with high-dose intravenous acyclovir (60 mg/kg/day in three divided doses for 2–3 weeks) followed by oral acyclovir for 6 months as a suppressive therapy.

Neonatal Varicella

Maternal infection during pregnancy may lead to pneumonitis in mother. Infection in pregnancy, especially during 8 to 20 weeks, there is risk of fetal damage (congenital varicella syndrome) with high mortality. Maternal primary infection at the time of delivery can result in very severe infection of the baby, with a mortality of about 30%.

Clinical features

- Cutaneous findings from 1 to 16 days postpartum.
- Papulo-vesicular lesions which can become hemorrhagic or necrotic.
- High risk for disseminated disease: pneumonitis, hepatitis, encephalitis.

Differential Diagnosis:

- Neonatal infections: HSV, candidiasis, *Staphylococcus aureus.*
- Erythema toxicum neonatorum.
- Transient neonatal pustular melanosis.

Investigation:

- Tzanck smear of vesicle shows multinucleated epithelial giant cells.
- Direct fluorescent antibody for VZV.
- Viral culture.

Management:

- Specific zoster immune globulin (ZIG) – for neonates whose mothers develop varicella within the period from 5 days before to 2 days after delivery.
- Oral acyclovir for pregnant mother with uncomplicated varicella infection.
- Post exposure prophylaxis to mothers without a history of varicella and with significant exposure (should be given within 10 days)
- Newborn with severe disseminated varicella requires IV acyclovir.
- Isolation from other infants.

Congenital Candidiasis

Candidiasis is the infection of the skin and birth membranes caused by candida albicans, which present at the time of birth, and following intrauterine infection.

Risk factors:

- Low birth weight babies.
- Cervical sutures and retained IU devices.

Clinical features:

- Amniotic fluid is turbid.
- Lesions typically appear on the first day of life, presenting with generalized erythematous papulo-vesicular eruption leading to desquamation.
- Often involves palms, soles and oral mucosa (thrush).
- Can have respiratory and gastrointestinal involvement from aspiration of infected amniotic fluid.
- Systemic involvement is rare.

Differential Diagnosis:

- As above
- Malaria
- SSSS

Investigation:

- Potassium hydroxide preparation from skin scrapings reveals budding yeasts and pseudo-hyphae.

Management:

- Topical: Imidazole cream BID for cutaneous disease is typically sufficient.
- Severe skin involvement - Fluconazole 3–6 mg/kg per day PO.
- Disseminated disease: Fluconazole 5 mg/kg per day IV.

SCLEREMA NEONATORUM

There is widespread skin thickening and induration in a critically ill neonate and associated with high mortality.

Clinical features:

- It can occur up to 4 months of age.
- Rapid onset skin hardening that typically starts distally and progress to involve other areas.
- Skin is bound down with mask like facies and immobile joints and no pitting on pressure.
- Palms, soles and genitalia are sparred.
- Associated with sepsis, hypoglycemic, hypothermia and metabolic acidosis.

Pathogenesis:

- Unknown; hypothesis of cold injury and secondary solidification of the tissue due to enzymatic dysfunction.

Differential Diagnosis:

- Subcutaneous fat necrosis of newborn
- Restrictive dermopathy
- Stiff skin syndrome

Treatment:

- Supportive measures to reverse the underlying disorder can revert the skin to normal.
- Careful monitoring, correction of electrolyte abnormalities, respiratory support,

correction of hypovolemia, and control of hypothermia.

- Exchange transfusions.

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