Lecture (9)
Dermatologic Emergencies

Objectives: not given.

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Color index: slides, doctor notes, extra explanation.

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Introduction

Emergency is an acute unexpected dangerous condition that requires quick action.

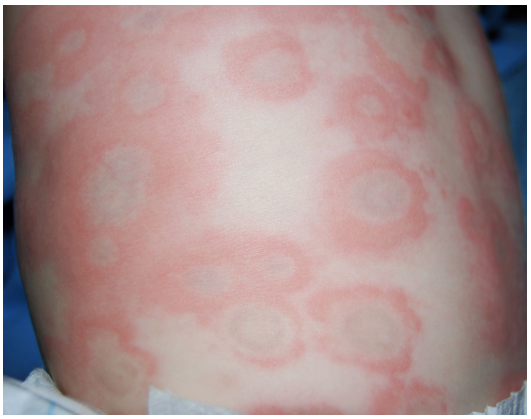
Some morphological patterns are alarming, meaning that this is an emergency condition, those morphological patterns include:

- Urticaria and angioedema.
- Purpura and ecchymosis.
- Bullae and sloughing.
- Necrosis and gangrene.
- Exfoliative Erythroderma Syndrome (EES).
- Generalized or widespread rashes in an acutely ill febrile patient.

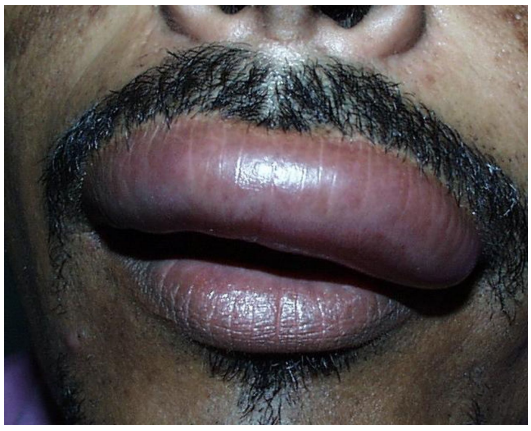
Urticaria\Angioedema

Transient swellings and erythema due to vasodilatation and fluid exudation. Manifest by elevated, evanescent annular wheals that develop rapidly and clear within hours. Can be life threatening especially when there is angioedema of the larynx. The condition may take years to resolve.

In angioedema, there is swelling of the eyes, lips and inside the airways that may cause stridor due to difficulty in breathing, this is a real emergency which is treated by epinephrine (patients with angioedema should carry a syringe that is inserted directly whenever they feel there is a swelling in the airways). A strange and rare presentation of angioedema is that they can develop inside the intestine leading to abdominal pain that mimic acute abdomen.



Annular lesions (Wheals) characteristic for Urticaria

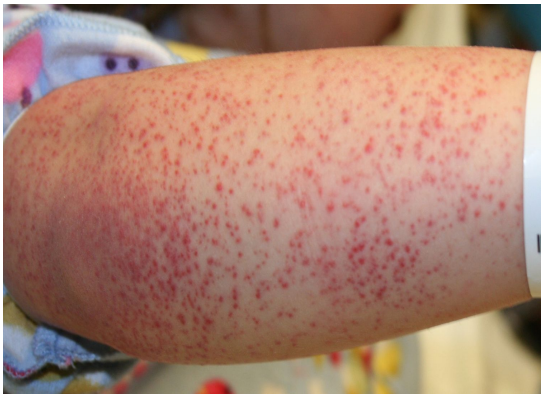


Swelling in the lips in Angioedema

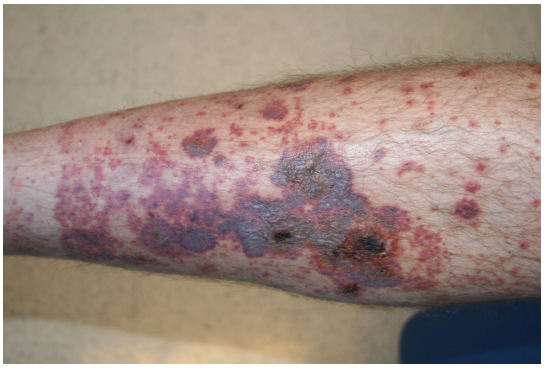
Purpura

Extravasated blood, purpura is mainly bleeding into the skin and it could be petechiae(small sized), purpura(medium sized) or ecchymoses(large sized).
This condition is caused by pathology in either of the following:
I - Inside blood vessel (disorders of coagulation)
II - Of blood vessel walls (Vasculitides)
III – Outside blood vessels (affecting supporting stroma due to: aging, drugs, Vit C deficiency, amyloidosis)

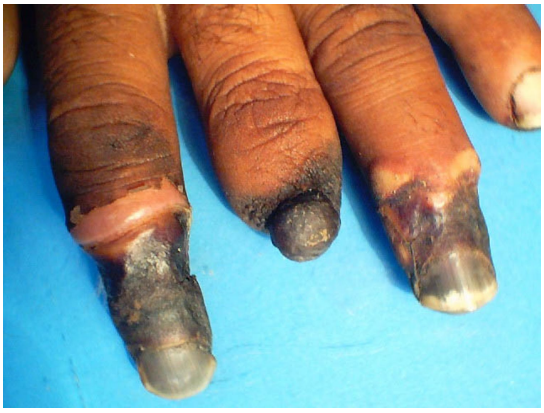
Purpura is important especially if it is palpable because it indicates vasculitis in which there is involvement of other body organs.



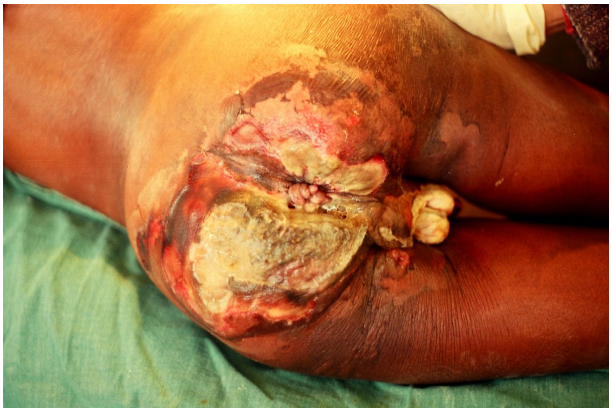
Petechiae



Palpable purpura and ecchymosis



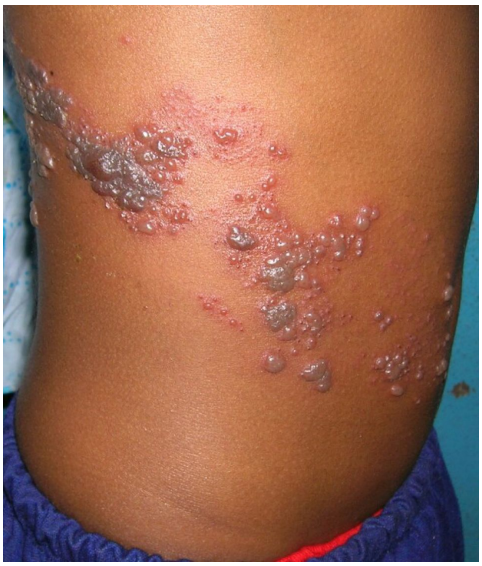
Blisters, digital infarction and necrosis at the tip of the fingers



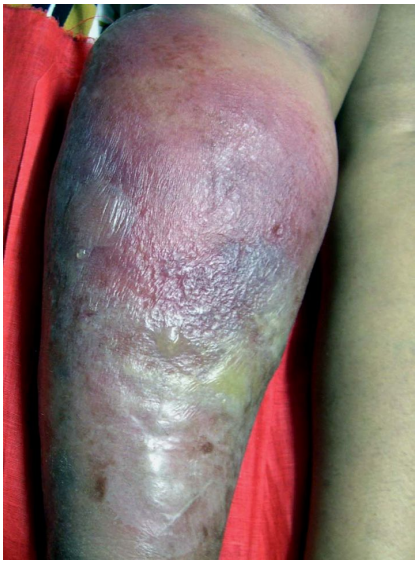
Sloughing of the skin

Bullous Diseases

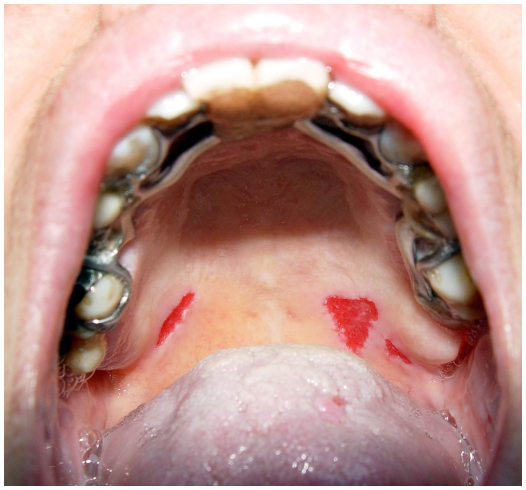
Blisters are circumscribed fluid filled skin lesions.
Burns, bullous impetigo, herpes simplex and zoster, severe contact dermatitis and insect bites are common examples.
Skin diseases presenting mainly with blisters are relatively rare but may be fatal such as autoimmune and mechanobullous diseases.
Flaccid blisters are blisters that are seen as an erosion because they are not intact(تتفجر بسرعة). They are associated with conditions such as pemphigus.



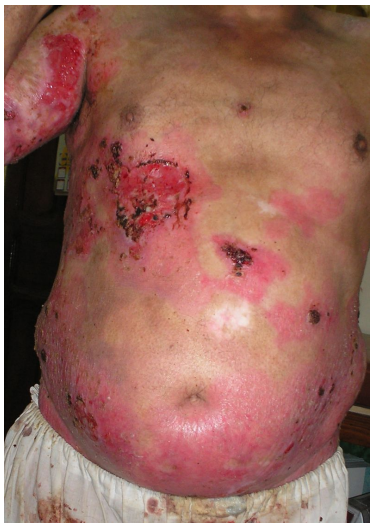
Blisters in dermatomal distribution seen in Herpes zoster



Very large vesicle



Flaccid blisters inside the mouth



Multiple erosions

Erythema Multiforme (EM)
Stevens Johnson Syndrome (SJS)
Toxic Epidermal Necrolysis (TEN) Spectrum

EM is a cutaneous reaction pattern to several provoking stimuli including infections and drugs or idiopathic. The classical lesion is targetoid that involve the hands and feet and less frequently the elbows and knees. There is now consensus that SJS and TEN are different from EM.

SJS and TEN are severe variants of an identical pathological process (apoptosis of keratinocytes induced by a cell-mediated cytotoxic reaction: Haptens vs. Cytokines) and differ only in the percentage of body surface involved (TEN is an extreme variant of SJS, it takes few days to develop TEN from SJS).

Both can start with macular and EM-like lesions; however about 50% of TEN evolve from diffuse erythema to necrosis and epidermal detachment. The condition is rare and life threatening.

Most common in adults more than 40 years with an equal male-female ratio. Risk factors include SLE, HIV. HLA –B12, it has multiple etiologies including: Drugs (sulfas, anticonvulsants, allopurinol, NSAIDS, antibiotics), infections, autoimmune, chemicals or idiopathic.

The rash starts on the face and extremities, it may generalize rapidly (few hours to days). The scalp, palms, and soles may be spared. Mucous membranes are involved(very specific feature of SJS and TEN), 85% have conjunctival lesions(which can lead to blindness due to adhesions). They may involve internal organs as well.
The condition later on evolve to:

- Confluent erythematous macules with crinkled surface.
- Raised flaccid blisters.
- Sheet like loss of epidermis.
- Red, oozing dermis resembling a second-degree burn.

The drug-induced type usually start with prodromal symptoms: fever, malaise, arthralgias 1-3 weeks after drug exposure and 1-3 days before mucocutaneous lesions. There may be tenderness, itching, burning, pain or paresthesia, photophobia, painful micturition, impaired alimentation and anxiety.

In histopathology there is full thickness necrosis of the epidermis and a sparse lymphocytic infiltrate. Recovery begins within days and completed in 3 weeks. Pressure points and periorificial sites take longer time to recover. Shedding of the nails and eyelashes may occur. It may lead to scarring and death. Mortality rate is 30% in TEN

and 10% in SJS, mortality usually occur due to sepsis and hemorrhage. Re-exposure to the offending agent is associated with more rapid and severe recurrence.

Differential diagnoses to this condition include:

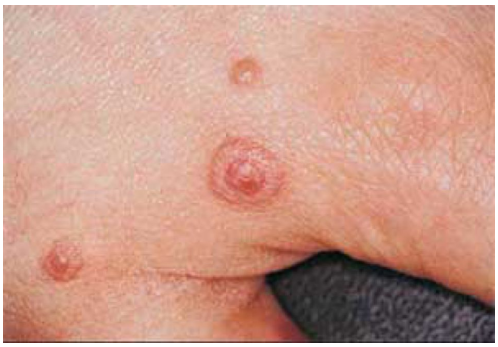
A. Exanthematous drug eruption.
B. Phototoxic eruptions.
C. Toxic shock syndrome.
D. Burns.
E. Generalized bullous.
F. Fixed drug eruption.
G. Exfoliative dermatitis.

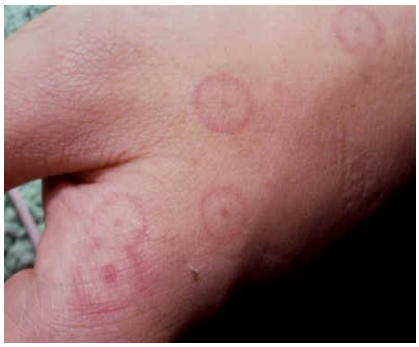
Bad prognostic factors include:

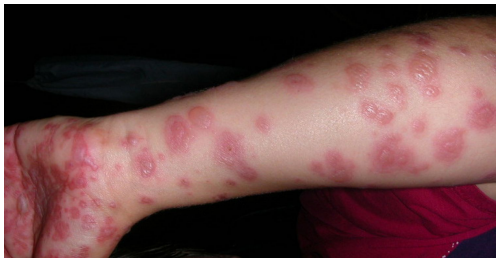
- Body surface area > 10%.
- Serum Urea >10mM.
- Age > 40 years.
- Heart rate >120.
- Serum glucose > 14mM.
- Serum Bicarbonate <20mM.
- Malignancy.

Management:

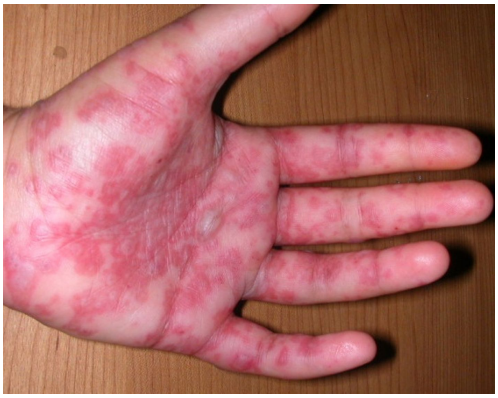
- ABC and STOP THE OFFENDING AGENT.
- Manage SJS and TEN patients as burn patients.
- Admission to ICU or burn unit.
- IV fluids and electrolytes as for a third degree burn, and temperature control of the skin.
- Symptomatic treatment.
- IV glucocorticoids/ immunoglobulins/ Pentoxifylline (usually expensive and not required).
- Treat eye lesions early to prevent blindness by referral to ophthalmologist (Very important).
- No surgical debridement.
- Daily skin dressing.



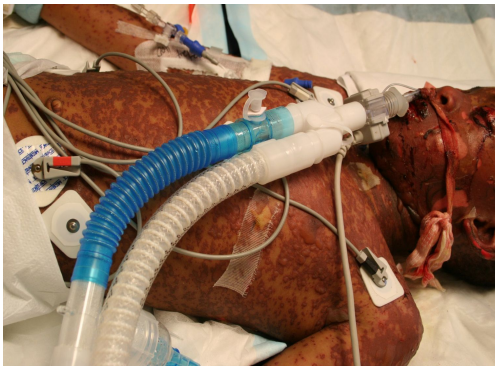




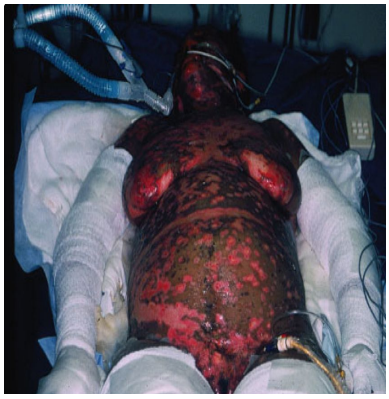
Targetoid lesions in SJS and TEN

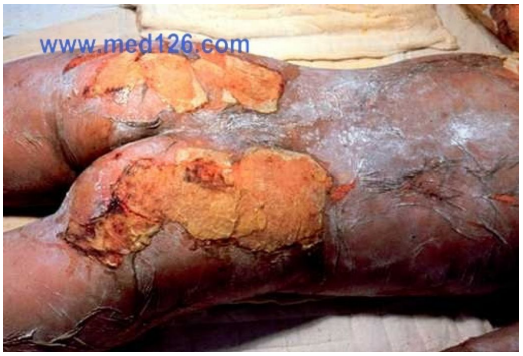


Mild form of TEN



Severe form of TEN with multiple blisters and erosions involving the whole skin. (notice mucosal membrane involvement as well)





Very severe forms of TEN, These patients usually die.

ERYTHRODERMA (Redness of the whole skin)

EXFOLIATIVE ERYTHRODERMA SYNDROME (EES):

EES is a serious, at times life-threatening reaction pattern of the skin characterized by: generalized and uniform redness, branny or lamellar scaling. Fever, malaise, shivers, pruritis, fatigue anorexia and generalized lymphadenopathy. Loss of scalp and body hair, nail thickening and onycholysis. It affects patients usually above 50 years with an increased risk in males. In children it mainly results from atopic dermatitis or pityriasis rubra pilaris.

Etiology:

- Following a pre existing dermatologic condition (psoriasis, eczema and others) in 50% of the cases.
- Drugs (eg. Allopurinol, carbamazepine, cimetidine, gold, lithium, quinidine) in 15% of the cases.
- Lymphoma, Leukemia in 10% of the cases.
- The etiology is undetermined in 25% of the cases.

Acute erythroderma

Erythroderma itself is not a disease, it is a sign indicating a severe disease.
Acute erythroderma is caused by drugs and it is potentially a fatal condition. It has profound effects on the entire body leading to poikilothermia, fluid and electrolyte imbalance, high output cardiac failure, increased basal metabolic rate,hypoproteinemia, anemia due to reduced levels of iron, folic acid and other vitamins, endocrine, hepatic and renal complications, effects on hair and nails. You don't need to worry about chronic erythroderma as much as the acute one, mortalities are usually associated with acute erythroderma.

The skin barrier in erythroderma is affected meaning that there is deterioration of skin function in maintaining body fluids, temperature and increased risk of infections.

When you see this condition you have two things: 1- How to look for the cause. 2- How to deal with erythroderma.

You have to look for the cause, there are some clinical clues about the underlying etiology: (doctor mentioned idiopathic, drugs and skin diseases and did not read the rest).

- It could be Idiopathic.
- If the condition is acute the etiology is usually drugs.
- If there are areas of sparing it is usually Pityriasis rubra pilaris(PRP).
- If there is massive hyperkeratosis and deep fissures of the palms/soles then it's usually: psoriasis, Cutaneous T-cell lymphoma(CTCL), PRP.
- If there is sparing of scalp hair it could be: psoriasis, eczema.
- If erythema is variable with scale thickness/ brownish hue and large lymph nodes then it is CTCL.
- When there is massive scaling of scalp with hair loss: CTCL, PRP.
- When it's dusky red : psoriasis.
- When it's Yellow/orange – red : PRP.
- If there is lichenification/erosions/excoriations : eczema.
- Sometime there are typical nail changes of psoriasis.
- Ectropion :CTCL, PRP.

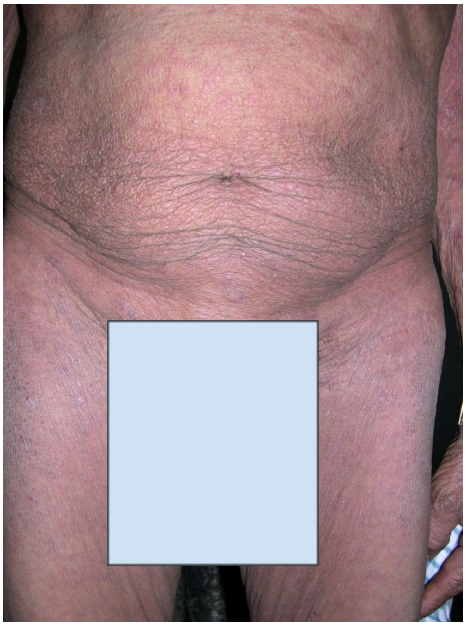
History(especially past medical and drug history) and physical examination for clues are important.
Histopathology is not always helpful
Investigations to identify the underlying disease are important, they include: chest X ray, immunoelectrophoresis, CT scans/ MRI and bone marrow aspiration, skin and lymph node biopsy, skin and blood bacterial cultures.

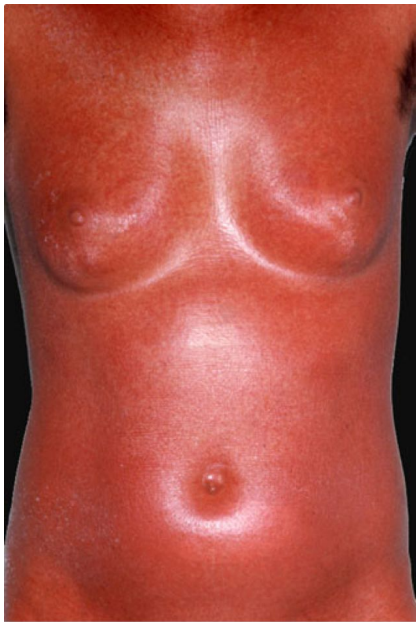
VITAL SIGNS ARE VERY IMPORTANT!
Sometime you may have to admit the patient due to hemodynamic instability, therefore checking vital signs is very important.

Management:
- Look for the cause and treat it.
- Order the proper investigations.
- Treatment is supportive by maintaining skin function, including fluid, electrolytes and albumin restoration, parenteral nutrition and temperature control.
- Look for signs of sepsis, renal and cardiac failure.
- Watch for deleterious adverse effects of prolonged glucocorticoid therapy.

Topical therapy:
Water baths, bland emollients ± topical steroids.
Beware of increase in absorption of topically applied medications eg: salicylism, methemoglobinemia.
Be cautious of irritant topicals eg: dithranol, tar.

Systemic therapy:
Oral glucocorticoids for remission induction but not for maintenance.
Specific systemic therapy for the underlying condition.





Very severe erythroderma