Derma Team 436

Dermatologic Emergencies

Objectives:

- Not given

الدكتور يقول السلايدات كافية لذلك تم اعتماد السلايدات مصدر للمحتوى

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Before you start.. CHECK THE EDITING FILE

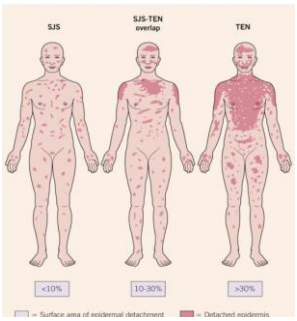
Sources:doctor's slides and notes
[Color index: Important | gold | doctor notes | Extr]

❖ Alarming Morphological patterns:

Urticaria / Angioedema	Purpura / Ecchymoses	Bullae / Sloughing
Necrosis / Gangrene	Exfoliative Erythroderma	Generalized/ widespread

❖ Steven's Johnson syndrome (SJS) Toxic epidermal necrolysis (TEN):

- Rare, acute, life-threatening mucocutaneous disease.
- Nearly always drug-related.
- Keratinocyte death à separation of skin at the dermal-epidermal junction.
- Characteristic symptoms: High fever, skin pain, anxiety and asthenia.
- It is crucial to diagnose it early so the causal drug can be discontinued.
- Asthenia: abnormal physical weakness or lack of energy
- SJS and its variant, TEN, involve the skin as well as mucus membranes.



- Spectrum of disease based on surface area involved
- SJS and TEN are variants of an identical pathologic disease and differ only in the percentage of body surface involved.

✓ Mortality:
- 5% for patients with SJS.
- 25%-50% for patients with TEN

✓ Cause:
- Medications (95%)
- Infections, Immunizations (rare)

Medications:
More than 100 drugs have been identified to date as being associated with SJS/TEN!

Most common:
- Allopurinol

Antibiotics (Sulfonamides)
NSAIDs
Anti-convulsants.

A lot of drugs can cause SJS, Allopurinol, Abx, NSAIDs and anti-convulsants are important causes that we should know.

Clinical features of SJS/TEN:

- Initially: Fever, Stinging eyes, and pain upon swallowing.
- These symptoms precede cutaneous manifestations by 1 to 3 days.
- Skin lesions first appear on the trunk, spreading to the neck, face and proximal upper extremities.
- Distal arms and legs are relatively spared (but not the palms/soles).
- ((Very important) if this is absent it is unlilkely to be SJS)) Erythema/erosions of the buccal, ocular and genital mucosae are present in more than 90% of patients.
- TEN -- epithelium of the respiratory and G.I tract can also occur.
- Skin lesions are usually tender & mucosal erosions are very painful.
- If Erythema/erosions of the mucus membranes is absent then its unlikely to be SJS (Very important).
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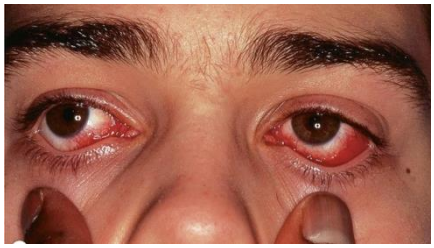
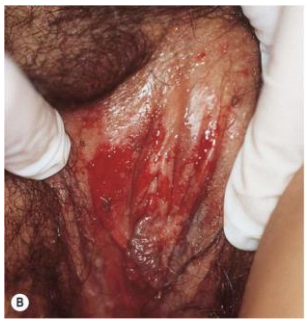
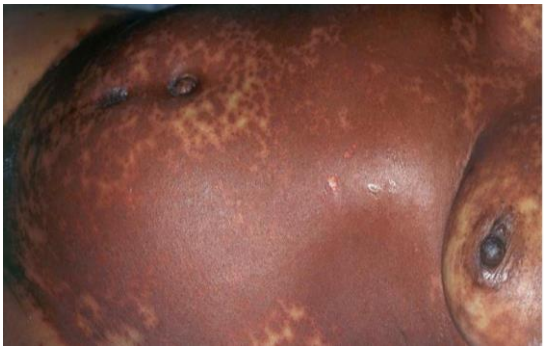




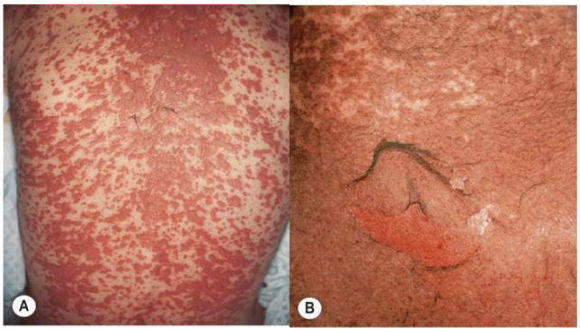
- Mucosal involvement in Stevens–Johnson syndrome.
- A Erythema and conjunctival erosions. B Erosions of the genital mucosa.

Morphology of skin lesions in SJS/TEN:

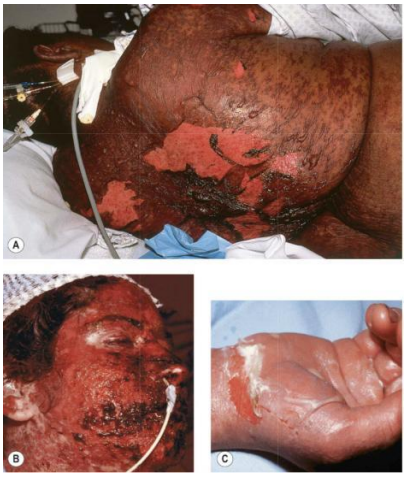
- First: erythematous, dusky red or purpuric macules of irregular size and shape, they have a tendency to coalesce. Coalesce is when multiple single lesions combine to form one big lesion.
- +ve Nikolsky sign. Nikolsky sign: dislodgement of intact superficial epidermis by a shearing force, indicating a plane of cleavage in the skin at the dermal-epidermal junction.
- Some lesions have a dusky center (Target-like appearance).
- Later: Full-thickness necrosis can develop (can be very rapid).
- The necrotic epidermis detaches from the dermis, fluid fills the space, giving rise to blisters (flaccid blisters).
- The blisters can be extended sideways by slight pressure of the thumb (Asboe-Hansen sign).
- The skin resembles wet cigarette paper.



Cutaneous features of toxic epidermal necrolysis (TEN). Characteristic dusky red color of the early macular eruption in TEN. Lesions with this color often progress to full-blown necrolytic lesions with dermal–epidermal detachment.



Stevens–Johnson syndrome (SJS) versus SJS–TEN overlap. A In addition to mucosal involvement and numerous dusky lesions with flaccid bullae, there are areas of coalescence and multiple sites of epidermal detachment. Because the latter involved >10% body surface area, the patient was classified as having SJS–TEN overlap. B Close-up of epidermal detachment, whose appearance has been likened to wet cigarette paper.



Clinical features of toxic epidermal necrolysis (TEN). A Detachment of large sheets of necrolytic epidermis (>30% body surface area), leading to extensive areas of denuded skin. A few intact bullae are still present. B Hemorrhagic crusts with mucosal involvement. C Epidermal detachment of palmar skin.

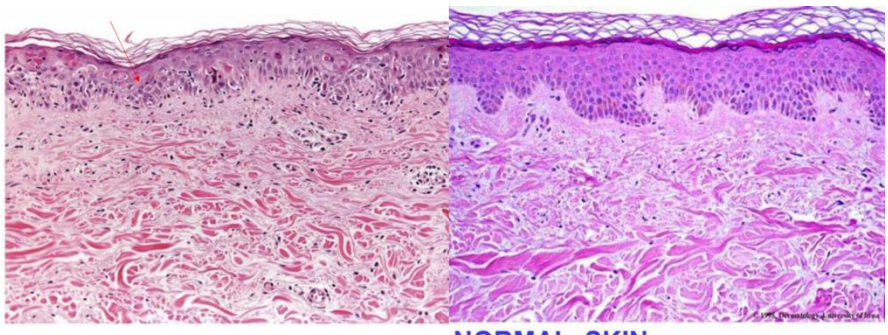
SCORTEN A prognostic scoring system for patients with TEN:

Age >40 years	Mortality rate:
HR >120 bpm	0-1 ----- 3.2%
Cancer or hematologic malignancy	2 ----- 12.1%
BSA involved on day 1 above 10%	3 ----- 35.8%
Serum urea level > 10 mmol/l	4 ----- 58.3%
Serum bicarbonate level <20 mmol/l	5 or more ---- 90%
Serum glucose level >14 mmol/l	

Toxic epidermal necrolysis (TEN):

- Death occurs in 1/3 of pts with TEN (mainly due to infections).
- Best managed in the ICU/Burn unit.
- Eliminating the culprit medication is the most important first step.
- SJS/TEN usually occurs 7-21 after the initiation of the drug (first exposure) and within 2 days in the case of re-exposure to a drug that previously caused SJS or TEN.
- We transfer the patients to a burn unit. Know the period to exclude medications outside that period.

Histology:

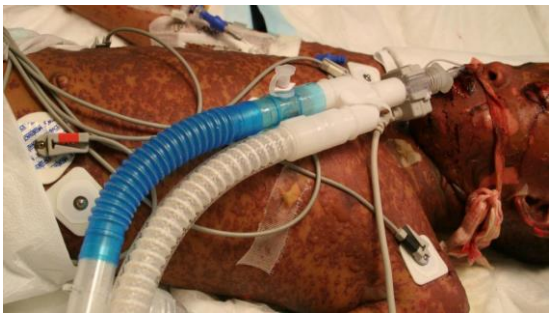


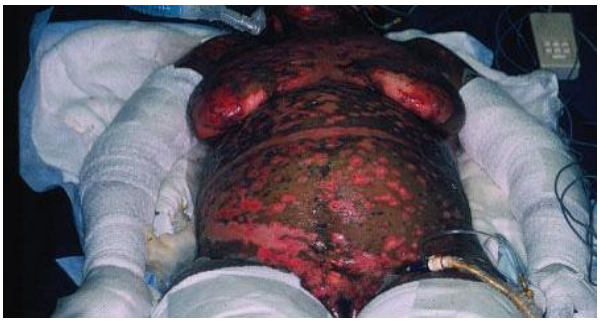
NORMAL SKIN

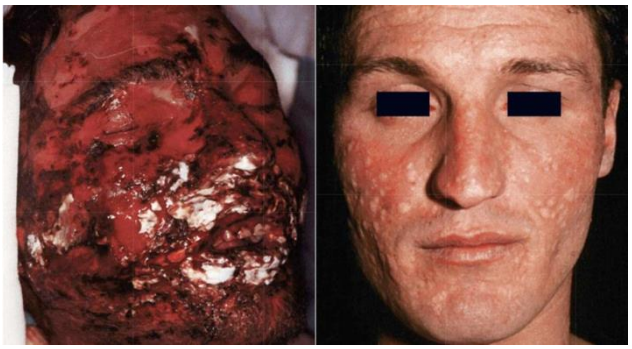
Toxic epidermal necrolysis – histopathologic features. Apoptotic keratinocytes are present individually and in clusters within the epidermis. Subtle vacuolar changes along the basal layer are accompanied by minimal inflammation, with scattered lymphocytes within the epidermis.

Treatment:

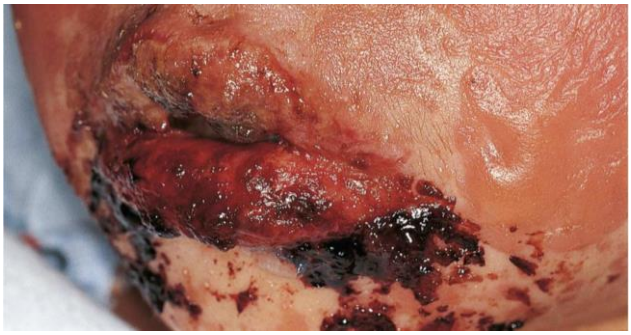
- Supportive care in a burn unit: wound care, hydration, nutritional support..etc
- Regular examination by an ophthalmologist (can have eye manifistations as well) it can cause blindness if left untreated.
- To date, no specific therapy has shown efficacy in prospective, controlled clinical trials.
- Cyclosporine
- Cyclophosphamide
- Systemic steroids
- IVIg (Good evidence on this one)







- Treatment of toxic epidermal necrolysis (TEN).
- Facial involvement of a patient with TEN (50% body surface area involvement) before (A) and 3 weeks after (B) treatment with IVIg (0.75 g/kg/day for 4 days).
- This a picture of a patient treated with IVIG.



- Childhood Stevens–Johnson syndrome secondary to trimethoprim–sulfamethoxazole therapy.
- Note the hemorrhagic crusts and denudation of the lips as well as bullous cutaneous lesions.

❖ **Erythroderma**: Erythroderma is not a diagnosis.

- Generalized redness and scaling of >90% of the skin surface.
- Considered a serious, at times life-threatening condition.
- It does not represent a disease but rather a clinical presentation of a variety of diseases.
- M > F (avg age is ~50 yrs)

Causes of Erythroderma:

Pre-existing dermatosis (psoriasis, eczema)

50%

Drugs

15%

Lymphoma, leukemia

10%

Undetermined

25%

Clinical features of erythroderma:

- Erythema precedes exfoliation by 2-6 days.
- Pruritis in 90% of patients.
- Palmoplantar keratoderma. (Thickness and redness of palms and soles)
- Nail changes in 40%.
- Diffuse non-scarring alopecia.

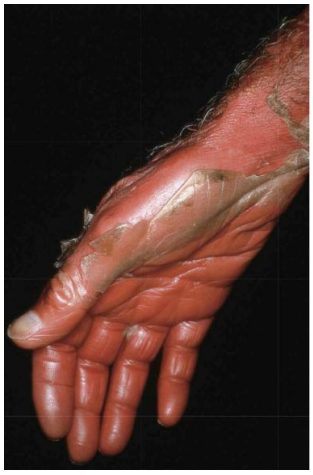
Systemic manifestations:

GENERALIZED PERIPHERAL LYMPHADENOPATHY (50%)

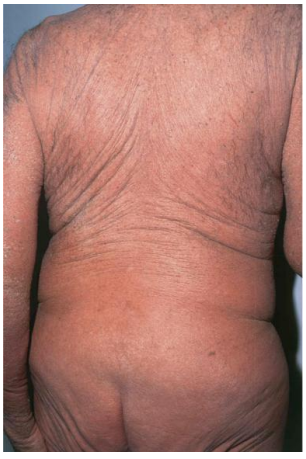
PEDAL OR PRETIBIAL EDEMA IN ~50% OF PATIENTS

TACHYCARDIA, RISK OF HIGH OUTPUT CARDIAC FAILURE (ESP. IN THE ELDERLY)

THERMOREGULATORY DISTURBANCES (HYPER-HYPO THERMIA)



Erythroderma with desquamation. Obvious exfoliation of scale with underlying erythema.



Idiopathic erythroderma. This is the type of patient that requires longitudinal evaluation to exclude the development of cutaneous T-cell lymphoma.

❖ Manifestations based on causative disease:

1) Psoriasis:

-Nail changes (Oil-drop, onycholysis, nail pits)

2) Atopic dermatitis:

- Pruritis is intense
- Lichenification (Lichenification is a skin condition that occurs in response to excessive itching or rubbing of the skin and results in thick, leathery patches of skin.)

3) Drug reactions:

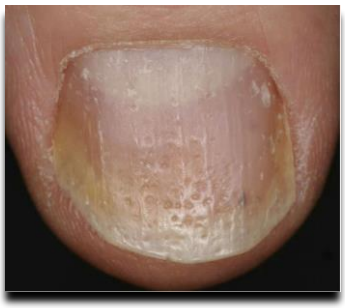
-Morbiliform or scarlatiniform exanthema (The term morbilliform refers to a rash that looks like measles. The rash consists of macular lesions that are red and usually 2–10 mm in diameter but may be confluent in places.)

4) Idiopathic erythroderma:

-Elderly men

-Lymphadenopathy and extensive palmoplantar keratoderma.

-Peripheral edema



Psoriatic erythroderma. Nail findings (pitting and onycholysis with a proximal rim of inflammation) point to the diagnosis of psoriasis.

5) CTCL:

-Sezary syndrome: Erythroderma, Malignant T lymphocytes and generalized lymphadenopathy.

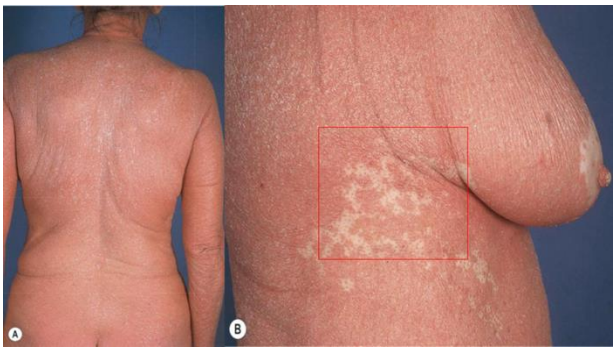
- Painful fissured keratoderma, diffuse alopecia, leonine facies.

6) PRP:

-Salmon to orange color.

-Follicular keratotic papules on the knees, elbows and dorsal fingers.

-Islands of sparing.

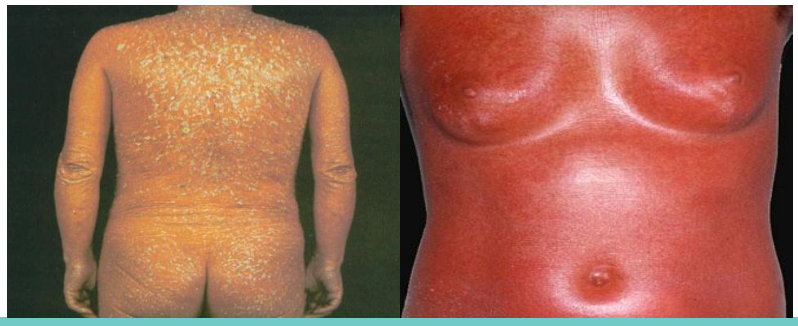


-Erythroderma secondary to pityriasis rubra pilaris. A few islands of sparing are noted on the upper back (A), but are more noticeable on the flank and breast (B). Note the salmon color.

-Islands of sparing is an important finding in PRP.

Treatment:

- Hospitalization may be required.
- Regardless of cause: Nutritional assessment, correction of fluid and electrolyte imbalance, prevention of hypothermia and tx of secondary infections.
- Idiopathic: Topical and systemic corticosteroids. Anti-histamines.
- Treat the cause of erythroderma.



Test your knowledge:

Angioedema Can be life threatening especially when associated with:
a) generalized lymphadenopathy
b) angioedema of the larynx
c) angioedema of the pharynx
d)fatigue anorexia

All are provoking stimuli of Erythema Multiforme (EM) except?
a) bacterial infection
b) drugs
c) Psoriasis
d) herpes simplex

Acute erythroderma is caused by:
a) bacterial infection
b) drugs
c) Psoriasis
d) herpes simplex

B
C
B