Dermatological Emergencies

Objectives: were not given

Note: doctor went over ALL tables so make sure to go over them

Color index:

- Main text
- Important
- Dr's explanation
- Golden notes
- Extra

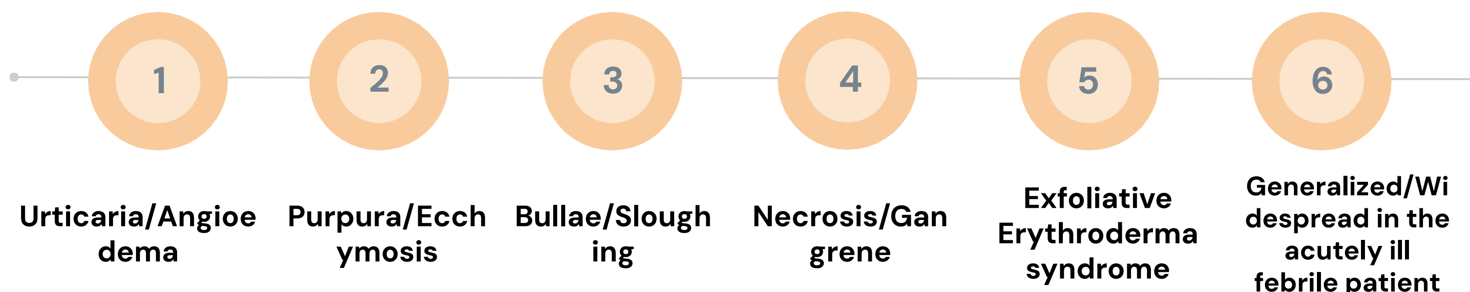
DERMATOLOGY TEAM 438

This lecture was originally done by both 438 & 439 teams.
So great thanks to them

Dermatological Emergencies

SJS/TEN | **Erythroderma** | **Purpura** | **Bullous disease** | **Urticaria/angioedema/anaphylaxis**

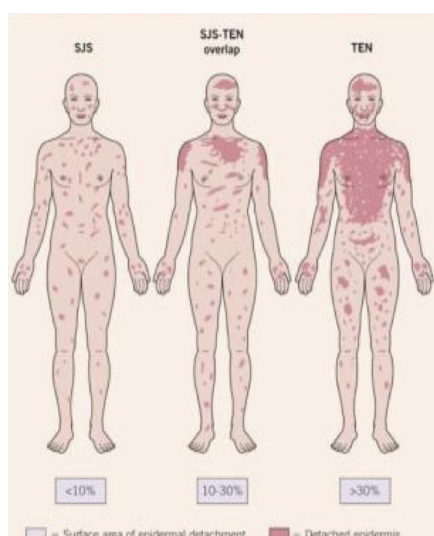
Alarming morphological patterns



Steven Johnson Syndrome (SJS) & Toxic Epidermal Necrolysis (TEN)

- Rare, acute, life-threatening **mucocutaneous** disease.
- Nearly always drug-related.
- Keratinocyte death lead to separation of skin at the dermal epidermal junction.
- **Characteristic symptoms: High fever, skin pain, anxiety and asthenia** (which is abnormal physical weakness or lack of energy).
- It is crucial to diagnose it early so the causal drug can be **discontinued.**
- SJS and its variant, TEN, involve the skin as well as mucus membranes.
- **Mortality:**
  - 5% for patients with SJS.
  - **25%-50% for patients with TEN.**

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.

Causes

1. **Medications**

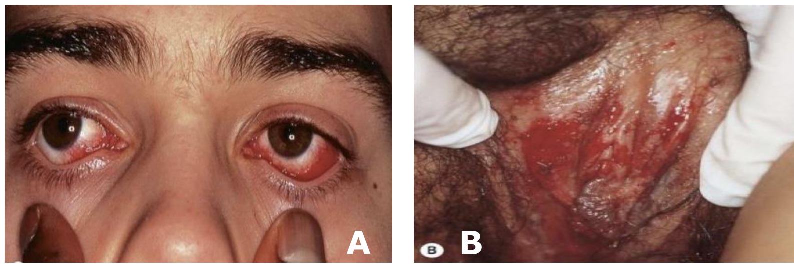
- More than 100 drugs have been identified to date as being associated with SJS/TEN! (we have to be updated).
- **Most common:**
  - Allopurinol
  - Antibiotics: (Trim-sulfa, Sulfonamides)
  - NSAIDs
  - Anticonvulsants.

2. **Infections and Immunizations (rare):** (Covid-19 vaccine-induced SJS TEN).

SJS/TEN

Clinical features of SJS/TEN

- Initially: Fever, Stinging eyes, and pain upon swallowing.
- These symptoms precede cutaneous manifestations by 1 to 3 days (you have to catch it early on).
- 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).
- Erythema/erosions of the buccal, ocular and genital mucosa are present in more than 90% of patients (if this is absent it is unlikely to be SJS).
- TEN involvement of epithelium of the respiratory and G.I tract can also occur.
- Skin lesions are usually tender & mucosal erosions are very painful.

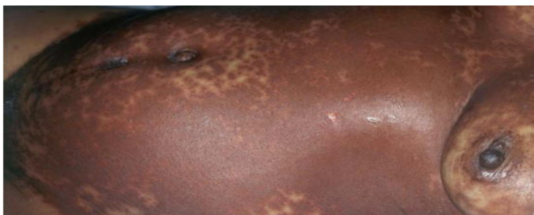


- Mucosal involvement in Stevens Johnson syndrome.

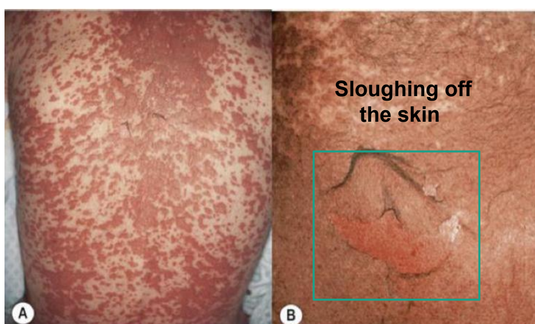
A) Erythema and conjunctival erosions.
B) Erosions of the genital mucosa.

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** (It is 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) you can see erosions as these blisters break easily.
- **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. (sloughing of skin)

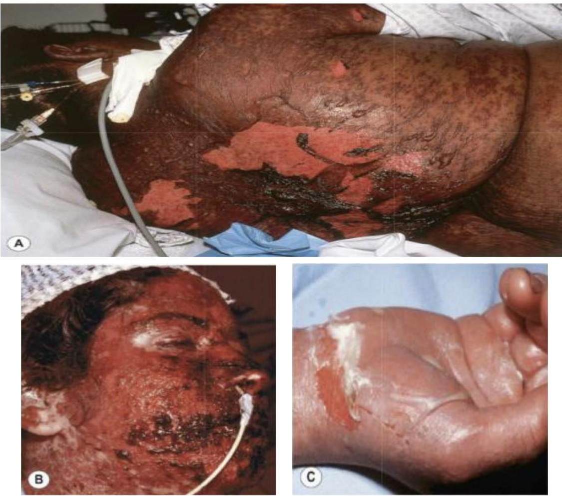


Stevens–Johnson syndrome (SJS) versus SJS–TEN overlap.

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.

SJS/TEN



Later on changes.
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

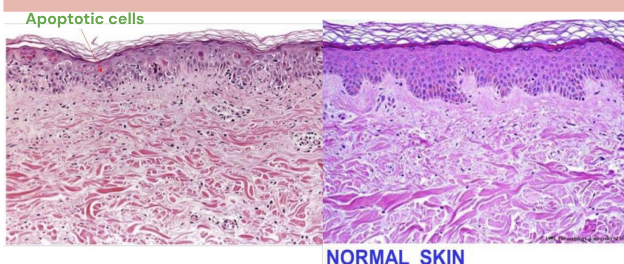
- Age > 40 years.
- HR > 120 bpm.
- Cancer or hematologic malignancy.
- BSA involved on day 1 above 10%.
- Serum urea level > 10 mmol/l.
- Serum bicarbonate level < 20 mmol/.l
- Serum glucose level > 14 mmol/l.

Mortality rate	
0-1	**3.2%**
2	**12.1%**
3	**35.8%**
4	**58.3%**
5 or more	**90%**

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 days** 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 duration to exclude medications outside that period.

Histology

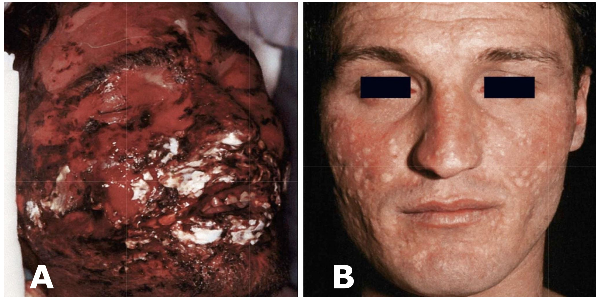


Toxic epidermal necrolysis – histopathologic features. Apoptotic keratinocytes are present individually and in clusters within the epidermis. Subtle vacuolar changes along the basal layer are accompanied by minimal inflammation, with scattered lymphocytes within the epidermis.

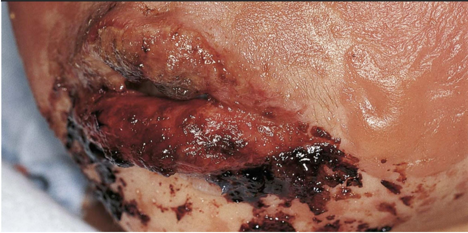
Treatment

- Supportive care in a burn unit: wound care, hydration, nutritional support..etc
- Regular examination by an ophthalmologist (for eye manifestations) 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 treatment)

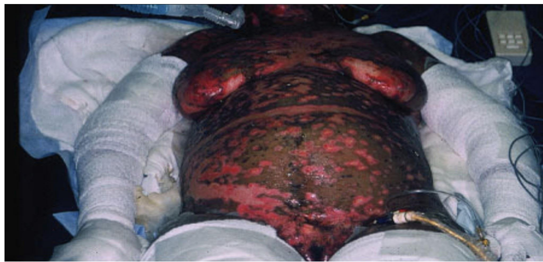
SJS/TEN & Erythroderma

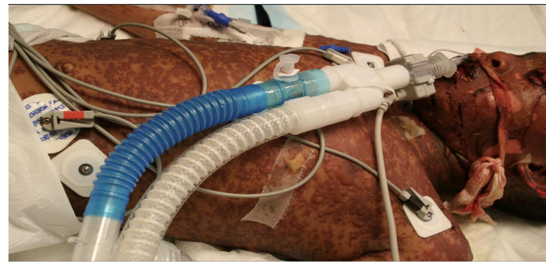


- Treatment of toxic epidermal necrolysis (TEN).
- Facial involvement of a patient with TEN (50% body surface area involvement) before (A) and 3 weeks after (B) treatment with IVIG(0.75 g/kg/day for 4 days).
- This a picture of a patient treated with IVIG.



- 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. It's a clinical presentation
- **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 (average age is ~50 yrs).**

Causes Of Erythroderma :

1) **Pre-existing dermatosis (psoriasis, eczema) 50% Most common.**
2) Undetermined 25%.
3) Drugs 15%.
4) Lymphoma, leukemia 10%.

Causes in adults VS **Causes in children**

Causes in adults:

- **Idiopathic.**
- **Atopic Dermatitis.**
- **Psoriasis.**
- **Drug reaction.**
- **Cutaneous T cell lymphoma (CTCL).**
- **Pityriasis rubra pilaris (PRP).**

Causes in children:

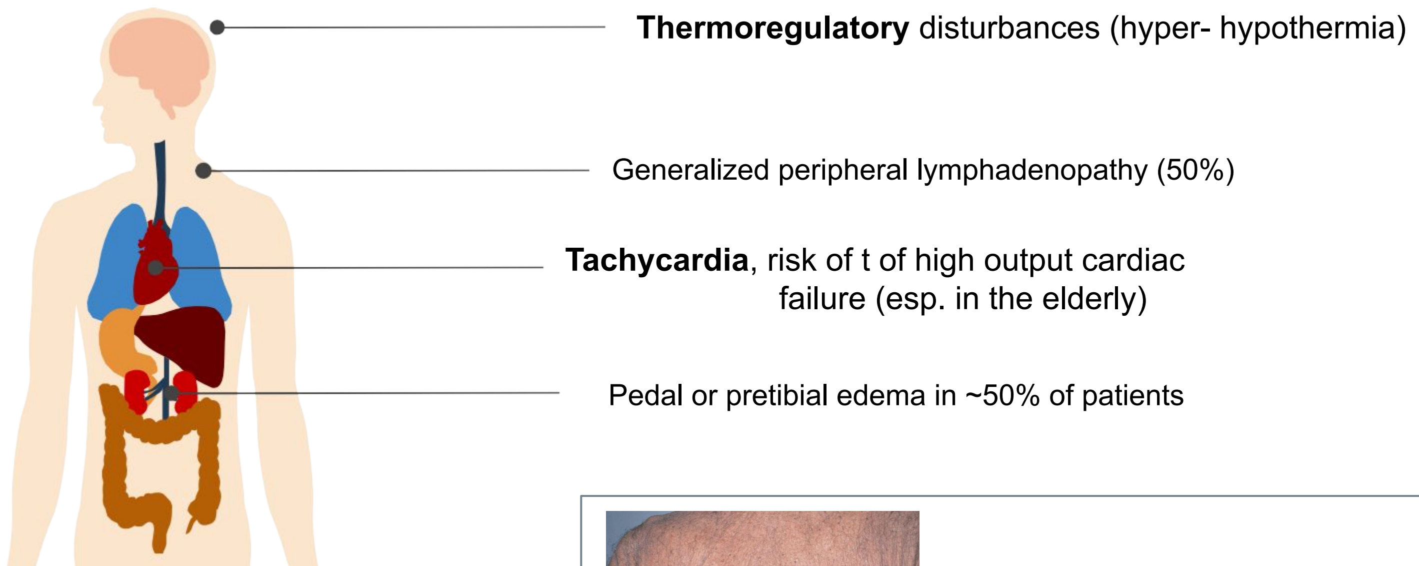
- Ichthyoses
- Immunodeficiencies , infections
- Dermatitis, Psoriasis.

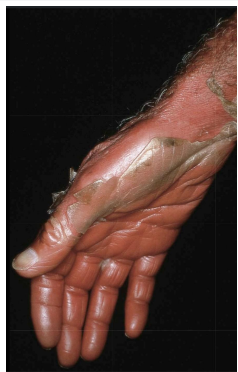
Erythroderma

Clinical features of erythroderma

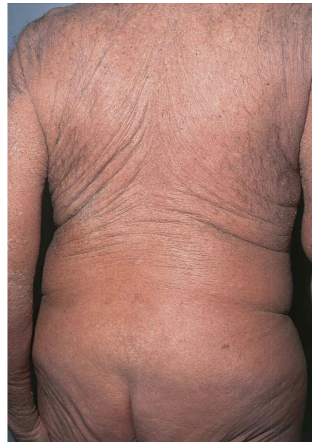
- Erythema precedes exfoliation by 2-6 days.
- Pruritis in 90% of patients.
- Palmoplantar keratoderma. (Thickness and redness of palms and soles)
- Nail changes in 40%.
- Diffuse non-scarring alopecia.

Systemic manifestations:





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



- Redness all over not normal skin.
- 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

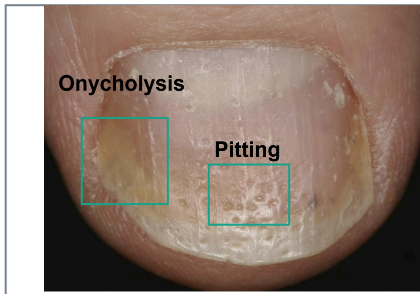
1) Psoriasis:
- **Nail changes** (Oil-drop, onycholysis, nail pits).

2) Atopic dermatitis:
- **Pruritus 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:
- Morbilliform or scarlatiniform exanthem (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.

Erythroderma

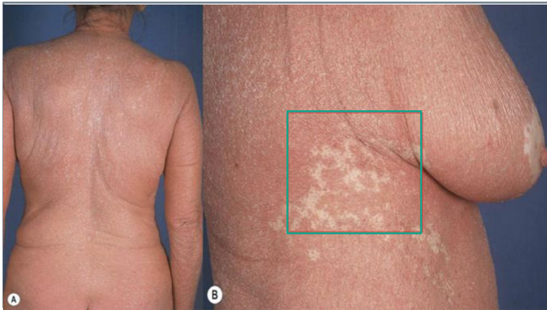


- Psoriatic erythroderma.
- Nail findings (pitting and onycholysis with a proximal rim of inflammation) point to the diagnosis of psoriasis.

Manifestations based on causative disease (cont'd):

5) CTCL (cutaneous T-cell lymphoma) : CTCL (More Benign), Sezary syndrome (malignant)
   - Sezary syndrome: Erythroderma, Malignant T lymphocytes and generalized lymphadenopathy.
     If you see CTCL and Erythroderma think about Sezary syndrome.
   - **Painful fissured keratoderma, diffuse alopecia, leonine facies.**

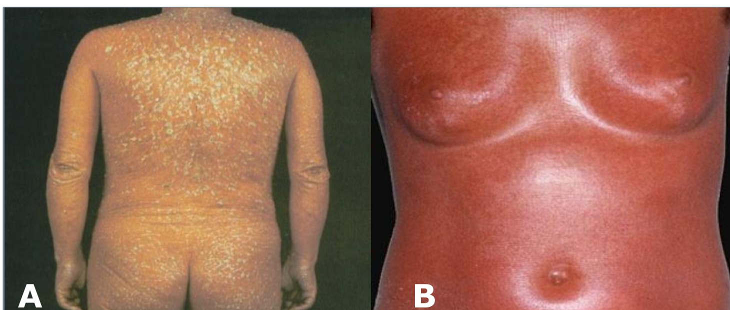
5) PRP (Pityriasis rubra pilaris) :
   - **Salmon to orange color.**
   - Follicular keratotic papules on the knees, elbows and dorsal fingers.
   - Islands of sparing.



- 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.(orange-like)**

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.**



- **(A)** A patient with Psoriasis.
- **(B)** 100% Erythroderma involvement of the skin.

Quiz!

1- In the pathogenesis of Stevens Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) which of the following is the best answer?

A) The circulating autoantibodies are directed against keratinocyte cell surfaces.
B) Impaired capacity to detoxify reactive intermediate drug metabolites.
C) Staphylococcus aureus producing epidermolytic toxin A and epidermolytic toxin B
D) It is Type I immunoglobulin E (IgE)–dependent drug reaction

2- Which of the following is/are clinical features of toxic epidermal necrolysis (TEN)?

A) Auspitz sign is positive
B) Tense blisters over the trunk and flaccid bullae over palms
C) Nikolsky sign is positive
D) The eruption starts at the distal portions of the arms and legs.

3- Regarding prognosis of toxic epidermal necrolysis (TEN) which of the following factors indicate poor outcome?

A) Children and young adults
B) Low serum glucose level
C) Low neutrophils count
D) Mucous membranes erosions

4- In the treatment of Stevens–Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) which of the following is the best answer?

A) Early use of systemic antibiotic as prophylactic for skin and systemic infections
B) Thalidomide is effective in blocking epidermal detachment and decrease mortality rate
C) High dose of intravenous immunoglobulins (IVIg) is associated with high mortality rate
D) Early diagnosis, immediate discontinuation of the causative drug(s) and supportive care

Answers:
1: A 2:C 3:C 4:D

Quiz!

1- A 20 year old lady who is epileptic present with 2 days history of fever, sore throat, malaise & painful cutaneous eruptions with dusky red color, 40% of epidermal detachment & hemorrhagic crusts of the lips. One month back, the epileptic medication was changed from valproate to carbamazepine. What is your diagnosis?

A) TEN secondary to carbamazepine
B) SJS secondary to carbamazepine
C) SJS secondary to valproate
D) TEN secondary to valproate

2- A 38 years old man referred to the on-call dermatologist with a 2-day history of sore throat, malaise and rash. Three weeks previously his antiepileptic medication had been changed to carbamazepine, on examination less than 10% of the skin surface is involved with erythematous, a typical target lesion on trunk, limbs and face, cheilitis, oral ulcers, conjunctivitis and erosions of the urethra. What is the most likely diagnosis?

A) Pemphigus vulgaris
B) Toxic epidermal necrolysis
C) Stevens-Johnson syndrome
D) Erythema multiforme

3- Acute erythroderma is caused by:

A) Bacterial infection
B) Drugs
C) Psoriasis
D) Herpes simplex

4- All are provoking stimuli of Erythema Multiforme (EM) except:

A) Bacterial infection
B) Drugs
C) Psoriasis
D) Herpes simplex

5- 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

Answers:
1 : A, 2 : C, 3 : B, 4 : C, 5 : B

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