Dermatological Emergencies

Objectives :

NOT GIVEN

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Sources: *doctor's slides and notes +FITZPATRICK color atlas +433 team male + 434*

[Color index : Important | Notes | Extra]

Dermatological emergencies

An introduction mentioned by the doctor but not on the PPT:

Important emergencies in Dermatology From 433:

Anaphylaxis:

- Is one of the most important emergencies in medicine (from voltarine for example)
- It's a type 1 allergic reaction that happens within seconds.
- Angioedema can kill the patient.

TEN:

- Is the most killing dermatological emergency because risk of sepsis and an infection.
- Killing emergency that needs to be managed urgently.
- Its mortality is 30% in comparison to SJS which is 1-5% and is not as serious as TEN.

Pemphigus vulgaris:

- Is an important emergency (killing) and before the era of steroids.

Necrotizing fasciitis:

(Most commonly by Group A beta-hemolytic streptococcus, **Group B** strept)

- You acquire it post trauma or surgery and also seen in diabetics.
- Violaceous and dusky red skin is an early presentation which can later extend to the fascia and deeper.
- It's treated by surgical debridement and antibiotics.

Cellulitis:

- Is a superficial infection with a well defined tender erythema.
- Predisposing factors: Portal for entry (wounds "especially between the toes", tinea pedis), diabetes, immunocompromised, etc.
- You must treat the underlying cause to avoid consequences such as sepsis and death.

Pustules:

- In neonates needs to be investigated, you cannot depend on your clinical judgment as it could be a presentation of (**Candida, herpes**, etc).
- The presentation is not typical in neonates for the present with pustules with no oral thrush in candida and in herpes they present with pustules.

Mucormycosis:

- Is one of the fatal infections in **diabetic patients** when they have diabetic ketoacidosis around the sinuses and has a picture somewhat similar to Necrotizing fasciitis.

Staphylococcal Scalded Skin Syndrome:

- Is common in children **(<6 yrs)**.
- SSSS is caused by the release of two exotoxins (Epidermolysin toxins A and B) from toxigenic coagulase positive strains 71 of the bacteria Staph. aureus.
- They cause very superficial sloughing of the subcorneal sloughing.
- All the nurses and doctors should be screened for staph carriers if a hospitalized patient was diagnosed with it.
- It may resemble TEN theoretically but clinically its more superficial than TEN.
- The initiating infection is usually in the form of crusting impetigo in the umbilical stump or nasopharynx.

Others:

Include erythroderma , pustular psoriasis (pt mistaken to have sepsis), Inflammatory disorders, in children (histiocytosis, astocytosis, hemangioma around orifices i.e oropharynx, eye), ulcerating diseases like pyoderma gangrenosum.

Definition:

Emergency is Acute, Unexpected, Dangerous unforeseen combination of circumstance which requires quick action.

Alarming Morphological patterns:

1. **Acute** Urticaria/ Angioedema
2. Purpura/ Ecchymosis
3. Bullae/ Sloughing
4. Necrosis / Gangrene
5. Exfoliative Erythroderma Syndrome (It's a fancy name of erythroderma don't be confused)
6. Generalized/ widespread rashes in the acutely ill febrile patients.
7. Also Severely painful area + target lesions + detachment of the skin + systemic manifestations very high fever.

Urticaria and Angioedema (الودمة الوعائية)

Essentials of Diagnosis:

- ➔ Wheals or hives.
- ➔ Evanescent: (disappears from a place and reappears on another) individual wheals disappear **within 24 hours** and often within minutes.
- ➔ Changing of configuration.

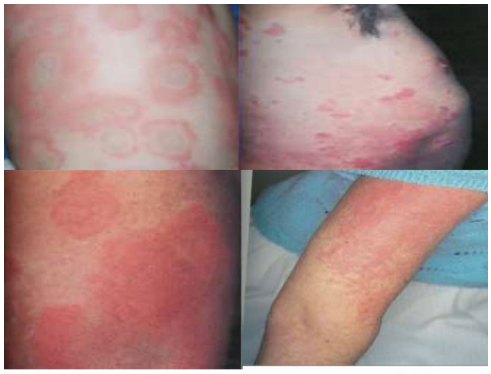
What's the difference between an urticarial wheal and urticated lesion?

- ➔ **Urticated**: looks like urticarial wheals but it leaves marks (pigmentation for an example) unlike urticarial wheal. They may be painful , lasting for > 24 hours
- ➔ **Urticarial wheal:** you know it by **history** + leaves no marks.

Pathogenic mechanism:

What is the major mediator of Urticaria?

- ➢ **In urticaria the pathology is in the dermis but in angioedema it's more deep (in subcutaneous tissue) which makes it more dangerous than urticaria.**
- ➢ Immunologic Type I (IgE mediated) or Type III. Its immediate or quick- Major mediator IgE and major effector cell is the Mast Cell.
- ➢ Non immunologic (activation of the alternative complement pathway and direct release of histamine) from basophils or tissue mast cells by drugs or chemicals.
- ➢ Transient swellings and erythema due to vasodilation and fluid exudation.
- ➢ Can be life threatening especially when associated with angioedema of the larynx. It can happen in eyelid, lip, larynx. Angioedema is more dangerous because it can affect the URT (obstruction ➛ stridor).
- ➢ May take years to resolve.



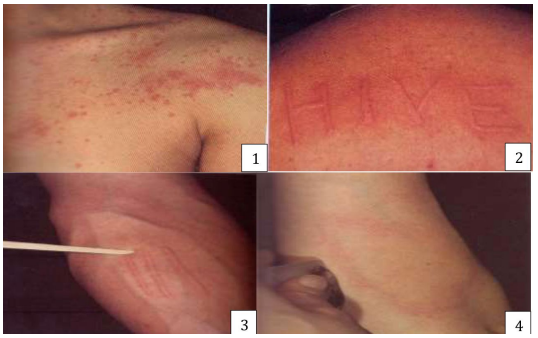
Urticarial wheel , edematous and erythematous , you don't need a biopsy unless you are worried about urticarial vasculitis

Classification of Hypersensitivity Reactions:

Hypersensitivity is a state in which the immune responses frequently take place in such a way that cell damage occurs and harmful pathological lesions may occur.

5 types of hypersensitivity are recognized	**"Immediate (Antibody mediated)":** ➢ **Type I: Anaphylaxis:** • It is IgE-mediated. An antigen (allergen) reacting with specifically sensitized IgE that is fixed to mast cells through its FC portion. • Degranulation of mast cells release of their mediators e.g. histamine, leukotrienes, ECF and NCF. • The offending antigen is identified by intradermal prick tests giving immediate wheal and erythema reactions or by provocation testing. • There is a strong familial predisposition and a tendency to produce high levels of IgE. e.g. anaphylaxis, urticaria, atopy. ➢ **Type III: Immune-Complex Reactions (arthus phenomenon):** • The antigen reacts with specific circulating antibodies antigen-antibody complexes that act through: (a) complement activation and PMNL attraction inflammatory response. (b) platelet aggregation microthrombi and vasoactive amine release. ➔ ***For Example:*** ★ **Serum sickness (Classical example, its was seen at the days of diphtheria Toxins)**. ★ Nephritis in SLE. ★ Allergic **vasculitis, urticarial vasculitis**. "Can present like urtications, but may last for > 24 , leaves pigments , presents with fever + very important to rule out internal organ deposition so start with UA (for cast , RBC, Proteins) to detect kidney involvement + rule out other diseases)". N.B. remember that vasculitis can present with any of the following purpura , petechiae , ecchymosis. ★ Bullous pemphigoid. "Immune Complexes "antigen-antibody in excess" which then will deposit on other major organs like CNS , GIT, Liver, and most commonly Kidney and Joints. **Urticaria may accompany systemic Anaphylaxis or serum sickness reaction.** **Serum sickness:** ➢ 5d 3 weeks after initial exposure (type III) immune complex mediated reaction. From the excess antigen antibody deposit in the vessels and organs. Exposure toxins i.e. anti-snake , anti-scorpion , drugs (classically) ➢ Fever, Urticaria, Angioedema, Joint pain and swelling, lymphadenopathy, occasionally : nephritis or endocarditis with eosinophilia. ➢ Minor form of Serum sickness: fever, Urticaria, **transitory** joint tenderness. "Without inflammation or other organ involvement". ➢ N.B in serum sickness like reaction you won't see major organ involvement only fever, skin and maybe arthralgia. **Acute Urticaria:** less than 6 wks | **Chronic Urticaria:** more than 6 wks. Recurrent: 2 times a year.

Angioedema:	➢ Oedema Involving the deep dermis or subcutaneous and submucosal areas. Why is it important? It may involve the airway like in oropharyngeal edema which is an emergency.
Causes of Urticaria and angioedema:	➢ Drugs, Animal sera, vaccines containing egg protein, desensitizing agents, Antibiotics, ACEI, radiocontrast media, cylogenase inhibitors. ➢ Foods, Inhalants, Infections. ➢ Autoimmune diseases. (SLE, autoimmune thyroiditis it's just an association, correcting the cause doesn't mean the urticaria will go).

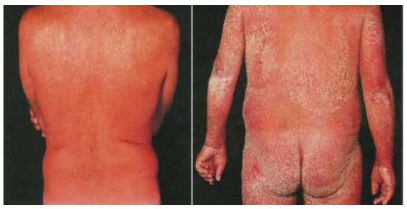
	➢ Insect bites. ➢ Physical causes. (like in red dermatographism (axon or flare reflex)that happens in 5% of people upon scratching the skin they develop a wheal and flare)and it doesn't have to be investigated). ➢ Hereditary causes, hereditary Angioedema. (There are three types quantitative its due to C1 esterase deficiency , functional , and gene mutation (rare) .. C1 usually presents in the form of angioedema and gastric involvement without urticaria, however if such a presentation occurs in elderly adding the symptom of pitting and pain then its acquired C1 deficiency and thus you must R/O things such as lymphoma, solid malignancy, sometimes autoimmune (SLE)hereditary isan emergency which doesn't respond to steroids usually and they need anti-androgens such as danazol to stimulate the C1 esterase secretion. ➢ Vasculitis (most important things to rule out in purpura: Drug, infection, and autoimmune disease. Malignancy (ovarian and silent ones is a very rare cause that you have to rule out if the pt was clinically free before the purpura onset). ➢ Contactants. ➢ Idiopathic. ➢ SLE (can cause chronic urticaria), thyroid disease. ➢ Neoplasm (lymphoma, ca of lung and colon). **Notes:** ➢ Acute most commonly is due to infection and drugs. ➢ In chronic urticarial we usually fail to find the cause (Idiopathic is the most common cause, 50% disappear after 6 months In acute (food, drugs and pollens are important causes) ➢ **Physical causes** (more information mentioned by the doctor): • **Red dermatographism** (Commonest) : wheal after scratching the skin, it's associated with urticaria. • **Cholinergic dermographism:** highly specific any high temperature getting sweaty, climbing the stairs he develops a pinpoint like (pen eraser size) urtication on the skin that disappears after few minutes. This is due to the release of Ach from the body heatno need to investigate, its picked up purely by the pts description • **White dermatographism** is associated with atopy. • **Cold urticaria:** can cause drowning. May be associated with cryoglobulinemia. • **Vibratory urticaria:** with vibration. Like people who work on mechanics. • **Solar:** many types related to waves of light in the sun. **Pic:** 1. Eraser size (cholinergic urticaria) 2. Dermatographism. 3. Dermatographism. 4. Cold urticaria: (after applying an ice cube for 5-20 minutes) happens after rewarming not at the time of cold exposure. 
Approach to the patient with chronic Urticaria:	➢ Start with history (drugs, food, other symptoms i.e. headache, abdominal pain, nasal, chest). ➢ Simple investigations like CBC (looking for eosinophilia, could be by drug, parasite, lymphoma(hodgekins), very high ESR (rule out malignancy by imaging according to thepatients' gender and age "its optional"), rule out infections (ex: stool for parasites) , UA (vasclitis) skin prick test (food and inhalants (IgE)) , CXR, TFT, HCV, metabolic panel.
Treatment:	• Elimination or avoidance of the causative agent. i.e. Drug • **Nonsedating H_1 antagonists.** (The mainstay of treatment for urticaria and histamine • mediated disorders) e.g. loratidine , claritine and cetirizine (can partially cross the BBB).

	• Sedating H1 antagonists (we give sedatives in dermatology in case of **itching** and non • histamine mediated disorders to sedate the brain) i.e. diaphenhydramine. • Addition of H2 antagonists: ranitidine , cimetidine (lots of drug interactions). • Corticosteroids (only with angioedema and acute not in chronic urticaria). • **Epinephrine**: lifesaving in angioedema and anaphylaxis , give the patient a shot and send him to hospital for further management)in the form of epipen then shift pt to hospital. • Montelukast (it's an anti leukotriene). • Thyroxine. • Colchicine. • Sulfasalazine. • Immunosuppressive therapy. • Antimalarial can be given to treat the urticarial vasculitis. • Omalizumab (humanized anti igE monoclonal antibody) • C1-esterase inhibitor deficiency: (we can't use systemic steroids and or antihistamines • epinephrine here) **Oral danazole, Oral tranexamic acid.** "Given in excessive menorrhagia too", For emergency: C1 inhibitor concentrate, fresh frozen plasma. Estrogen & OCP should be avoided. **In acute emergency episodes:** ➔ Secure the airway. ➔ I.V. line. ➔ Adrenaline subcutaneous or I.M., repeated every 10 min. ➔ Diphenhydramine I.M. or I.V. ➔ Hydrocortisone I.V. ➔ Patients with severe angioedema should be admitted for at least 24 hours observation particularly where laryngeal edema has occurred.

Anaphylaxis

Essentials of Diagnosis:	➢ Laryngeal edema or bronchospasm or both. ➢ Erythema, puritus, urtiaria or angioedma (any or all)o Vomitting, cramps, diarrhea. ➢ Hypotension, cardiac arrythmia or shocks.
General considerations:	➔ Within minutes to hours, severe may be fatal. ➔ **IgE mediated.** ➔ Chemical mediators are released. ➔ Anaphylactoid reactions: clinically similar reactions that involve the non-immunologic direct release of mast cells (non antigen-antibody) release of similar mediators e.g. (reactions to radiographic contrast media, aspirin, local anesthetics). **How is aspirin related to urticaria?** It's unlikely to be a direct cause of urticaria, but can increase it, that's why pts with chronic attacks stop aspirin during the attack an resume in between.
Causative agents:	➢ Drugs. ➢ Foods. ➢ Vaccines and Antisera (anti snake, anti scorpion) ➢ Insects – bees, wasps. ➢ Immunotherapy of allergic rhinitis, asthma or stinging insect sensitivity. (when they induce small quantities of allergen, it should be in a place that is prepared for resuscitation) ➢ Other causes: A. Iodinated contrast media, aspirin, local anesthetics and anaphylactoid reactions. B. Treatment of anaphylaxis.

Erythroderma and Exfoliative Dermatitis

Clinical features:	➢ Is defined as Erythema with or without scales affecting >90% of the body. ➢ It's common in extreme of ages (neonates (immunity related, BSA in relation to weight) and old age (drug pharmacodynamics , immunity) and they are vulnerable for complications.  Generalized Erythema Generalized Erythema with scales , erythrodermic psoriasis
Complications:	➢ Hypothermia ➢ Fluid and electrolyte loss ➢ Infections ➢ HF (increase hemodynamic state) ➢ Stress induced GIT ulcer ➢ Malabsorption ➢ Venous thrombosis ➢ Hypoalbuminemia due to loss from the Skin. • They have a poikilothermic animals they adjust their body temperature to the outer one which can put them into the serious complication of hypothermia which is more serious than hyper. • The best outer temperature that suits our body is 23 degrees.
Drug Etiology:	➢ Sulphonamides, antimalarials, penicillin, phenytoin, Carbamazepine (tegretol), phenobarbitone, allopurinol for gout, Steroid and NSAIDs.
Other causes:	• Psoriasis "it can present for the first time as severe erythroderma". • Drugs. • Cutaneous T-cell lymphoma. • Solid malignancies. • Severe atopic dermatitis, seborrheic dermatitis PRP. • Sarcoidosis. - First 3 are the most common. - You have to ask about previous skin diseases and to take a >3 biopsies from different places (unless you're sure of the cause) and if you didn't know the cause repeat the biopsy every 4 months.

Erythema multiforme Major, SJS, Toxic Epidermal Necrolysis:

Steven Johnson Syndrome SJS, TEN May represent variants of the same disease process.

Definition of EM:

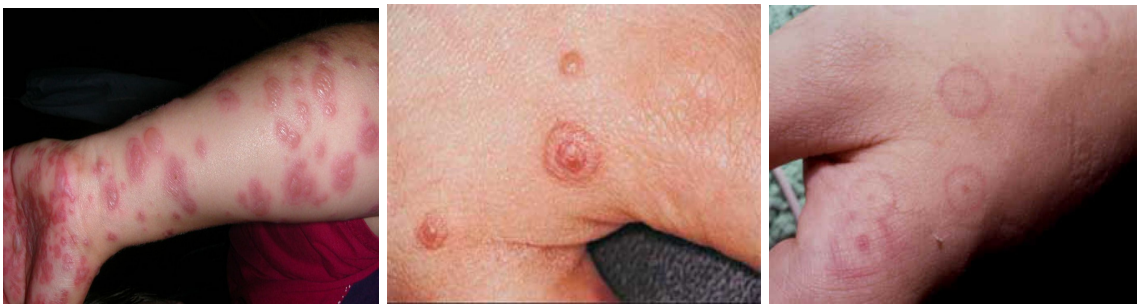
- EM is a cutaneous reaction pattern to several provoking stimuli including **herpes simplex,** bacterial infection and drugs. May be idiopathic.
- **The target (iris-like) lesions** involve the hands and feet and less frequently the elbows and knees.
- There is now consensus that SJS and TEN are different from EM.

- **Clinically:** Mucous membrane erosions (you can't diagnose SJS without it, usually the oral and eye mucosa are involved), target lesions.
- Epidermal necrosis with skin detachment.
- SJS < 10% BSA of epidermal detachment, SJS –TEN Overlap 10-30%, TEN > 30%
- EM minor little or no mucosal involvement, Preceding herpes labialis in 50% of EM.

- Systemic symptoms (Fever, arthralgia, sore throat) present in EM major and you will 2 or 3 mucosal membrane involvement & absent or limited in EM minor.
- In the erythema multiforme , look for the target lesion . if you can't find them it doesn't exclude the dx . usually we don't see it SJS its atypical and crusted.
- The main difference between TEN and stevens johnsons is the involvement of mucus membrane and the extent of the skin involvement which is more in TEN and higher mortality in TEN.

Suspected etiologic factors:	• Infections: Suspect an infection with SJS, erythema multiforme minor and major (in SJS the most common causative infection is **Mycoplasma pneumonia** especially in children and inEM minor it's recurrent herpes), also always look for an infection in children. Other infections include histoplasmosis , parvovirus (ORF virus of sheep nodules). ➢ **Drugs > 95% in TEN:** (but in SJS is it's around 50%) anticonvulsants, sulphonamides, allopurinol, NSAI. ➢ Neoplasia ➢ Collagen disease (less likely with TEN). ➢ Immunizations (some types of vaccines but not very common). ➢ The more severe the reaction it's a drug, and the less severe is with infections.
Risk factors for TEN:	➢ Slow acetylators (problem in getting rid of the metabolites, they usually have a family history of drug reactions (genetic background). ➢ Immunosuppression (HIV, Lymphomas) they have a high risk for drug reactions casing TEN because they have acquired **Glutathione deficiency** therefore, they can't scavenger the metabolites. ➢ Radiotherapy & anticonvulsants ➢ Specific HLA ➢ FDA recently recommended genotyping of all asians for the allele **HLAB 1502** prior **starting Carbazepine.**
Prognostic factors in TEN:	➢ (Mortality rate: 30% with TEN, 1-5% with SJS) > BSA, extreme of age, low immunity, diabetes, high urea, low sodium bicarbonates (all are bad prognostic indicators), ?Metabolic base may trigger an immune response. ➢ The problem with erythema multiforme is with systemic involvement and they lose the functions of the skin and this is emergency they must be admitted (shock and die) . ➢ **Major cause of death: Septicemia**, electrolyte imbalance. ➢ **Differential diagnosis:** • **Pemphigus vulgaris** (especially if there was bad oral mucosa involvement). • **SSSS** (but it's more superficial and it doesn't have the raw red appearance of TEN). • Kawasaki in children (they have erythema around the fingers, coronary involvement, conjunctival injection, strawberry tongue but not the cheilitis of SJS. They have to be treated with aspirin and immunoglobulins immediately)
Pathogenesis:	➢ Metabolic base may trigger an immune response ➢ Prognosis: related to extent of skin involvement. ➢ Mortality for TEN: 30% related to sepsis, fluid and electrolyte imbalance.
Management:	➢ **Early diagnosis, withdrawal of suspected drug.** ➢ Patient best cared in a burn (ideal place) or I.C.U. ➢ Replacement of I.V. fluids and electrolytes. ➢ Don't add unnecessary treatment the pt is already getting a reaction from a drug , adding more will only make things worse. ➢ Systemic corticosteroids-controversial → In TEN we don't start with it , yet in SJS if you start it early in the disease for a short period of 3-5 days may help , but not late as the mortality may increase. ➢ Care for mucous membrane, eye involvement.

	➢ Diagnose and treat complicating infections (take cultures if you suspect infection). ➢ Pt will need high caloric and protein diet just like burns. ➢ Prevention → never give the causing drug again it create a re-challenge reaction and a worse reaction. ➢ Others which aren't used commonly include immunosuppressant therapy like acetylcysteine IV (used in panadol toxicity, cystic fibrosis (oral)), cyclosporine , these should be given early not late , infliximab , etanercept.



★ Lt: Neumorus, well-defined, erythematous, blisters & target lesions, involving the arm.
★ Middle pic: Concentric rings (مركز ها موحد).