12-Erythroderma

Background:

- Aka exfoliative dermatitis, pts who have **erythema** and **scaling affecting > 80-90% BSA** regardless of cause
    - At least 50% caused by pre-existing rashes that worsen
    - Children: May also be 2/2 immunodeficiency (e.g. OMENN syndrome)
- **Main Causes (7)**
    - **Papulosquamous**: psoriasis, pityriasis rubra pilaris
        - Psoriasis is the most common cause of erythroderma (~ 20% of cases)
            - Caused by withdrawal of steroids/cyclosporine/methotrexate vs triggers ("SICK LAB"→ Smoking/Stress, Infections, Hypocalcemia, Koebnerization, Lithium, Anti-malarial/ACE-I, BB…also alcohol, obesity, NSAIDs, terbinafine, TNF-alpha inhibitors)
    - **Dermatitis**: atopic, allergic contact, seborrheic, chronic actinic dermatitis, stasis derm
        - *Atopic Derm*: hx of ≥1 features of atopic triad: atopic derm, hay fever, asthma. More severe itching. Look for ↑ IgE + eosinophilia on labs
        - *Allergic Contact*: (e.g. *Parthenian* plant in India.)
        - *Seborrheic*: think of neurologic conditions (Parkinson's) or HIV
        - *Chronic Actinic*: men >50 caused by UVA, UVB, and visible light. ↑CD8:CD4 .
        - *Stasis derm with profound Id reaction*: very rare cause of erythroderma
    - **Drug Rxn:** SJS, TEN, DRESS syndrome, drug rash
        - SJS triggers: Allopurinol, sulfa drugs (Bactrim), phenytoin, HIV pt w/ antiretroviral therapy
    - **CTCL**: erythrodermic MF, Sezary syndrome
        - Erythrodermic MF: pre-existing patches or plaques of MF then progress to erythroderma
        - Sezary Triad: Diffuse lymphadenopathy, malignant T-cells, erythroderma. Also look for alopecia, nail changes, leonine facies.
            - 1000 Sezary cell per microliter or ↑ CD4:CD8 >10, ↑CD4 (CD7/26-)
    - **Infections**: Viral exanthems, Norwegian Scabies, Staph Scalded Skin syndrome
    - **Auto-immune conditions** such as Lupus, GVHD, bullous pemphigoid
    - **Physical causes:** e.g. Burns

- PEARL: Differential for leonine facies? "PALMS" **P**aget's disease of the bone, **A**myloidosis, **L**epromatous Leprosy, **L**ymphoma, **L**eishmaniasis, **M**ycosis fungoides, **S**arcoidosis, **S**cleromyxedema

Clinical Presentation:

- Very scaly rash!
- **Skin vasodilation** = ↓ **peripheral resistance** may lead to **tachycardia**, **high output cardiac failure,** and **edema**
- Extra blood flow in skin can **disrupt thermoregulation** leading to hyper or hypothermia. CC: **Chills**!
- **ROS:**
    - **Severe pruritis**: **atopic dermatitis** or **Sezary**
    - **Joint pain**: **psoriasis**
    - Fever: argues against psoriasis

- **Hints on Physical Exam (suggestive findings):**
    - **Rash** affecting the **face** = argues against **psoriasis**
    - **Facial edema** = **drug rxn** or **DRESS**
    - **Mucosal** Inflammation = **SJS/TEN**
    - **Waxy keratoderma** = **pityriasis rubra pilaris**
    - **Nail changes** (**pitting**, onycholysis, oil spotting) = **psoriasis**
    - **Violaceous** rash = **CTCL**
    - **Salmon** rash = **pityriasis rubra pilaris**
    - **Blisters** = **bullous pemphigoid**
    - **Follicular plugging** w/ **islands** of **sparing** on **dorsal fingers** and **knees** = **pityriasis rubra pilaris**
    - **Larger** areas of **peeling** = **acute drug rxn** (vs **Fine scale** = **atopic dermatitis** or **generalized tinea**)
    - **Lymph nodes** = malignancy (**Sezary**)
        - Note: lymphadenopathy is not uncommon in erythroderma regardless of cause

Diagnosis (suggestive findings):

- CBC: eosinophilia → drug rxn or atopic derm
- CMP: Electrolyte Imbalances & LFT elevations – DRESS syndrome
- Blood Culture & Viral Cultures
- IgE: atopic derm
- ANA: Lupus and dermatomyositis
- Peripheral blood smear & Flow Cytometry: Sezary cells Flow; ↑ CD4:CD8 >10:1
- Multiple Biopsy: may be non-specific in 1/3 of patients; *still a crucial part of workup!
- KOH study if rash is scaly to r/o generalized tinea
- Lymph node biopsy vs PET CT if lymphadenopathy + concern for Sezary syndrome

Treatment:

- **Diffuse scaling:** **Emollients diffusely** w/ **class IV-XI topical steroid**
- If **Secondary infection** present? **Topical antibiotics** like **mupirocin** or bleach baths
- **Pruritis**: **Wet dressings** & **sedating anti-histamines** (**Benadryl**, **hydroxyzine**), consider **prednisone**, cyclosporine, or methotrexate depending on etiology and severity

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