Erythroderma

Basic Dermatology Curriculum

Updated 12/24/2016
The following module contains a number of blue, underlined terms which are hyperlinked to the dermatology glossary, an illustrated interactive guide to clinical dermatology and dermatopathology.

We encourage the learner to read all the hyperlinked information.
The purpose of this module is to help medical students develop a clinical approach to the evaluation and initial management of patients presenting with erythroderma.

By completing this module, the learner will be able to:

- Identify and describe the morphology of erythroderma
- Name common diseases and medications associated with erythroderma
- Explain the potential morbidity and mortality in erythrodermic patients
- Discuss the initial management of an erythrodermic patient
Erythroderma: The Basics

- Also called exfoliative dermatitis
- Defined as generalized redness or scaling of the skin, affecting almost the entire (>80-90%) body surface area (BSA)
  - Vesicles are not typically seen
  - May present with extensive telogen effluvium
- Erythroderma is not a diagnosis, it is a **clinical manifestation** of an underlying disease.
Erythroderma: Clinical Presentation

- Can evolve slowly or develop acutely
  - Common symptoms include: fevers, chills, malaise and pruritus
  - Patients may also experience peripheral edema, lymphadenopathy, secondary skin infection
  - Can often be an exacerbation of a pre-existing dermatologic disorder such as psoriasis, atopic dermatitis, or a drug reaction, among others.
  - Long-standing severe erythroderma is associated with diffuse alopecia (hair loss), keratoderma (hyperkeratosis of the stratum corneum), nail dystrophy (nail plate abnormalities), and ectropion (outward turning of the lower eyelid)
- Significant risk for morbidity and mortality, accounting for 1% of all dermatologic admissions to the hospital
- Complications of erythroderma include sepsis, hypothermia, and high-output cardiac failure
Medications Implicated in Erythroderma

- The most commonly implicated drugs include:
  - Anti-epileptics
  - Allopurinol
  - Antibiotics
    - Penicillin
    - Sulfonamides
    - Vancomycin
  - Calcium channel blockers
  - Cimetidine
  - Dapsone
  - Gold
  - Lithium
  - Quinidine
Erythroderma: Etiology

- Frequently the result of the generalization of an underlying dermatosis
  - Psoriasis
  - Atopic dermatitis
  - Chronic actinic dermatitis

- Drug eruptions

- Idiopathic

- Malignancy
  - Mycosis Fungoides/Sézary Syndrome
  - Paraneoplastic erythroderma

- Drug eruptions
  - Seborrheic dermatitis
  - Pityriasis rubra pilaris
  - Allergic contact dermatitis
Case One

Mr. Robert Ashton
Case One: History

- **HPI**: Mr. Ashton is a 63-year-old man who presents to the dermatology clinic with a rapid progression of skin redness covering most of his body.
- **PMH**: coronary artery disease s/p 3v CABG, hypertension, psoriasis.
- **Medications**: beta-blocker, aspirin, ace-inhibitor, statin, and topical clobetasol. No new medications.
- **Allergies**: none.
- **Family history**: no history of skin disorders.
- **Social history**: lives alone in an apartment.
- **Health-related behaviors**: no tobacco, alcohol or drug use.
- **ROS**: pruritus, fatigue.
Case One: Exam

- Vital signs: T 38.0 (100.4°F), BP 95/68, HR 115, RR 16, O2 Sat 97%
- Gen: moderate distress, patient is shivering
- Skin: diffuse erythema with overlying scale covering > 90% of the BSA
- Mucosal: no mucous membrane involvement
Evaluation of Erythroderma

- Evaluation of erythroderma begins with a thorough history, including a complete medication history.
- Physical exam requires special attention to the vital signs, nails, mucosa, lymph nodes and evaluation for hepatosplenomegaly.
- Baseline blood work, skin biopsy and, at times, cytologic or histologic evaluation of lymph nodes is the next step in evaluation.
  - Multiple (and repeat) skin biopsies may be necessary to make a definitive diagnosis.
Evaluation of Erythroderma

- Underlying malignancy may need to be excluded

- Regardless of the underlying cause, if a patient appears unstable or toxic, admission to the hospital is recommended

- Evaluation of erythrodermic patients *should include* a dermatology consult
Mr. Ashton is a 63-year-old man with a history of psoriasis who presented with generalized erythema. Given his concerning vital signs, Mr. Ashton was admitted to the hospital for evaluation and treatment.
Case One, Question 1

What is the most likely diagnosis in this case?

a. Atopic dermatitis flare
b. Cutaneous T-cell lymphoma
c. Drug Eruption
d. Psoriatic erythroderma
e. S. aureus scalded skin syndrome
Answer: d

What is the most likely diagnosis in this case?

- a. Atopic dermatitis flare (no history of atopic dermatitis. AD erythroderma tends to present more with weeping and crusting)
- b. Cutaneous T-cell lymphoma (hard to tell the difference, but CTCL erythroderma may present with symmetric islands of uninvolved skin. Also may spare areas of skin that are frequently folded, such as the abdomen)
- c. Drug eruption (none of his medications are new and there are no changes in dosages)
- d. **Psoriatic erythroderma** (patient has known psoriasis)
- e. *S. aureus* scalded skin syndrome (usually presents with cutaneous tenderness and widespread superficial blistering and denudation)
Psoriatic Erythroderma

- Erythrodermic psoriasis is a severe form of psoriasis that can arise acutely or follow a more chronic course.
- Can arise in patients with long-standing psoriasis or can occur de novo as the initial presentation of psoriasis.
- Triggers for erythrodermic psoriasis include:
  - Discontinuation potent topical or oral treatment
  - Medications used for other conditions
  - Infections (including HIV)
  - Pregnancy
  - Emotional stress
Case Two

Mrs. Grace Barringer
Case Two: History

- **HPI:** Mrs. Barringer is a 54-year-old woman with progressive redness. It started on her scalp and progressed to the trunk and extremities over the last three weeks.
- **PMH:** asthma, chronic dry, itchy skin, and hay fever.
- **Medications:** daily multivitamin, albuterol inhaler as needed, moisturizers, occasional antihistamines.
- **Allergies:** none.
- **Family history:** noncontributory.
- **Social history:** lives with her husband, has three grown children.
- **Health-related behaviors:** no tobacco, alcohol or drug use.
- **ROS:** itching, chills, emotional distress over skin changes.
Case Two: Exam

- VS: T 98.6, HR 105, BP 110/60, RR 14, O2 sat 100%
- Skin: large erythematous plaques with overlying scale and crust
Case Two, Question 1

What is the most likely diagnosis?

a. Atopic dermatitis flare  
b. Sézary Syndrome  
c. Drug Reaction  
d. Staph Scalded Skin Syndrome  
e. Psoriatic erythroderma
Case Two, Question 1

**Answer: a**

What is the most likely diagnosis?

- **Atopic dermatitis flare** (History of asthma, hay fever and chronic, dry itchy skin suggestive of atopic dermatitis)
- **Sézary Syndrome** (may present similarly, but often spares flexures and skin folds)
- **Drug Reaction** (Possible, but atopic dermatitis more likely given history of atopic disease)
- **SSSS** (typically presents in persons <18yo, and evolves quickly)
- **Psoriatic erythroderma** (No history of psoriasis)
Case Two, Question 2

Which of the following treatments should take priority in any patient with erythroderma?

a. Leg elevation  
b. Oral antibiotics  
c. Discontinue potential offending and unnecessary medications  
d. Topical corticosteroids
Case Two, Question 2

**Answer: c**
Which of the following treatments should take priority in any patient with erythroderma?

- a. Leg elevation
- b. Oral antibiotics
- c. **Discontinue potential offending and unnecessary medications**
- d. Topical corticosteroids
Case Three

Mr. Fred Rasck
Case Three

• Mr. Rasck is a 72yo man who was healthy until one year ago when he started to have itching on his skin. His skin gradually became pink and scaly all over, now his nails are affected and his neck has mildly palpable lymph nodes.

  - PMH: hypertension.
  - Medications: Lisinopril/HCTZ, recently started oral antihistamines
  - Allergies: none
  - Family history: noncontributory
  - Social history: lives with his wife, has two adult children
  - Health-related behaviors: previous marathon runner, no tobacco, alcohol or drug use
  - ROS: itching, chills, visibly frustrated
Case Three- Physical Exam
Case Three- Physical Exam
Case Three- Physical Exam

• You notice widespread pink erythematous skin with fine overlying scale.
• The face, neck, trunk, extremities, palms and soles are affected.
Case Three, additional information

• **Baseline Biopsy #1** shows some epidermal acanthosis, there is no significant epidermotropism, atypical lymphocytes are not observed, and some eosinophils are present.

• **Repeat Biopsy #2** six months later… shows some atypical lymphocytes, acanthosis (epidermal thickening) and mild **spongiosis**. (epidermal edema)
Case Three, Question 1

What is the most likely diagnosis?

a. Atopic dermatitis flare
b. Sézary Syndrome
c. Drug Reaction
d. Staph Scalded Skin Syndrome
e. Psoriatic erythroderma
Case Three, Question 1

Answer: b

What is the most likely diagnosis?

a. Atopic Dermatitis (late adult-onset atopy is rare without history of asthma/allergies or prior dry skin suggestive of eczema)

b. Sézary Syndrome (many patients experience a delay in diagnosis because eczema/psoriasis are more common and skin biopsies in SS are often non-specific, lymphadenopathy suggests this diagnosis but is not necessary to make it, and can be reaction just due to erythrodermic state)

c. Drug Reaction (it is possible that he has an evolving dermatosis to his Lisinopril/HCTZ, and this drug should be discontinued for at least 6mos, and alternate hypertensive medication of a different class such as a beta- or calcium-channel blocker)

d. SSSS (typically presents in persons <18yo, and evolves quickly, patients often mildly febrile)

e. Psoriatic erythroderma (No history of psoriasis)
Case Three, Question 2

• What laboratory/pathology test is necessary to confirm your diagnosis?
  a. Lactate dehydrogenase
  b. Flow cytometry & T cell clonality studies
  c. Complete blood count
  d. PET-CT
  e. Repeat skin biopsies
Case Three, Question 2

Answer: b.

What laboratory/pathology test is needed to confirm your diagnosis?

a. Lactate dehydrogenase (may be elevated but is not diagnostic)
b. Flow cytometry & T cell clonality studies (the malignant cell in Sézary Syndrome is a T-cell that is predominantly leukemic but can also home to the skin before re-entering circulation)
c. Complete blood count (may be completely normal, or may have anemia, leukocytosis, eosinophilia but none are consistently found or confirmatory)
d. PET-CT (may demonstrate lymphadenopathy, but also can be completely normal)
e. Repeat skin biopsies (20-30% of skin biopsies of patients with Sézary Syndrome are non-diagnostic or show inflammation that mimic other inflammatory dermatoses that cause erythroderma: eczema, drug eruptions, pityriasis rubra pilaris, etc.)
Initial Management of Erythroderma

Regardless of the underlying cause, the initial management of erythroderma remains the same:

- Discontinue potential offending and unnecessary medications
- Address nutrition, fluid and electrolyte balance
- Evaluate for signs and systems of cardiac or respiratory compromise
- Generalized skin care with soaks and wrapping weeping or crusted sites, ointment based emollients and coat skin surface with mid-potency topical corticosteroids (face mild-potency)
Initial Management Continued

- Oral antihistamines for relief of pruritus (and anxiety)
- Warm, humidified environment to prevent hypothermia and improve hydration of the skin
- Treat secondary infections with systemic antibiotics
- Treat peripheral edema with leg elevation
Erythroderma: Prognosis

- Prognosis depends on the underlying cause
- Determining the underlying etiology and removing any contributing external factors (especially medications) remain the most important factors in treatment
Take Home Points

- Erythroderma is a clinical manifestation of a variety of underlying diseases
- Defined as generalized redness or scaling of the skin, affecting a significant amount of the BSA
- Potential risk for morbidity and mortality and hospitalization is often required
- Initial management of erythroderma includes removing any potential offending and unnecessary medications
- When in doubt, consider T-cell clonality studies and flow cytometry for Sézary Syndrome, which can mimic other dermatoses on pathology and clinical exam.
Acknowledgements

- This module was developed by the American Academy of Dermatology Medical Student Core Curriculum Workgroup from 2008-2016.
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End of the Module