Dermatologic Emergencies and Conditions in the Hospital Setting

Definition and Epidemiology

Dermatologic emergencies and conditions in hospital medicine are severe or urgent skin disorders requiring prompt management to prevent systemic complications, organ dysfunction, or death

This guide covers necrotizing fasciitis, erythroderma, purpura fulminans, drug reaction with eosinophilia and systemic symptoms (DRESS), and acute generalized exanthematous pustulosis (AGEP), detailing causes, presentation, diagnostics, treatment, and complications

- Prevalence
  - These conditions are rare, affecting ~0.1-0.5% of inpatients, with necrotizing fasciitis incidence ~0.4-1 per 100,000, erythroderma ~1-2 per 100,000, DRESS ~1 per 10,000, and AGEP ~1-5 per million
- Risk Factors
  - Infections (e.g., Streptococcus), immunosuppression, medications (e.g., antibiotics), autoimmune diseases, malignancy
- Rare Demographics
  - Pediatric purpura fulminans, post-transplant DRESS, bariatric patients with fasciitis

Pathophysiology

- Mechanisms
  - These conditions arise from infectious (necrotizing fasciitis), inflammatory (erythroderma, AGEP), immune-mediated (DRESS), or coagulopathic (purpura fulminans) processes
  - Necrotizing fasciitis involves bacterial toxins causing tissue necrosis
  - Erythroderma results from widespread cutaneous inflammation
  - DRESS is a delayed hypersensitivity reaction with systemic involvement
  - Purpura fulminans stems from microvascular thrombosis and DIC
- Effects
  - Skin barrier loss leads to infection, fluid/electrolyte imbalance, and sepsis
  - Systemic inflammation (IL-6, TNF-α) drives multi-organ failure

- Molecular Pathways
  - **Necrotizing fasciitis:** Streptococcal exotoxins trigger cytokine storm
  - **DRESS:** T-cell activation, HHV-6 reactivation
  - **Erythroderma:** IL-2, IL-17 promote skin inflammation
  - **Purpura fulminans:** Protein C deficiency causes thrombosis
- Key Pathway Trigger (infection, immune, coagulopathy) → Skin/systemic damage → Sepsis, organ dysfunction

Causes and Differential Diagnosis

- Causes:
  - Necrotizing Fasciitis: Group A Streptococcus, MRSA, polymicrobial (diabetes, trauma, surgery)
  - Erythroderma: Psoriasis, eczema, drug reactions, cutaneous T-cell lymphoma
  - Purpura Fulminans: Meningococcemia, protein C/S deficiency, sepsis-induced DIC
  - DRESS: Medications (carbamazepine, allopurinol), HHV-6 reactivation
  - AGEP: Antibiotics (penicillins, macrolides), rapid drug exposure
  - Rare:
    - Vibrio vulnificus fasciitis,
    - paraneoplastic erythroderma,
    - post-vaccination DRESS,
    - congenital protein C deficiency
- Conditions to Rule Out
  - Infection (sepsis, endocarditis)
  - Blood cultures, WBC, CRP
  - Malignancy (lymphoma)
  - PET-CT, lymph node biopsy
  - Viral Reactivation (HHV-6, DRESS)
  - Viral PCR
  - Coagulopathy (DIC)
  - D-dimer, fibrinogen, INR

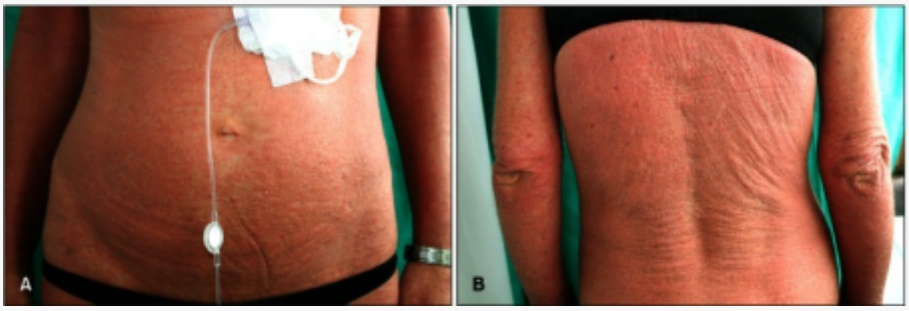
Clinical Presentation

- Symptoms
  - Necrotizing Fasciitis: Severe pain out of proportion, fever, swelling, skin necrosis
  - Erythroderma: Intense pruritus, diffuse redness, scaling, chills
  - Purpura Fulminans: Purpuric rash, fever, shock, limb ischemia

    - DRESS: Fever, rash, facial edema, lymphadenopathy
    - AGEP: Fever, pustules, burning sensation
- Exam:

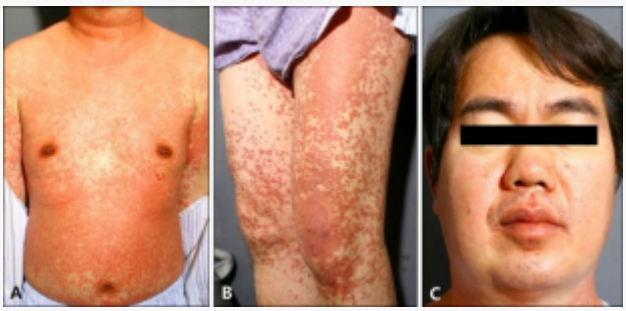
Necrotizing Fasciitis: Erythema, bullae, crepitus, necrosis, LRINEC score >6 (see below)

Erythroderma: Diffuse erythema (>90% BSA), scaling, lymphadenopathy (see below)

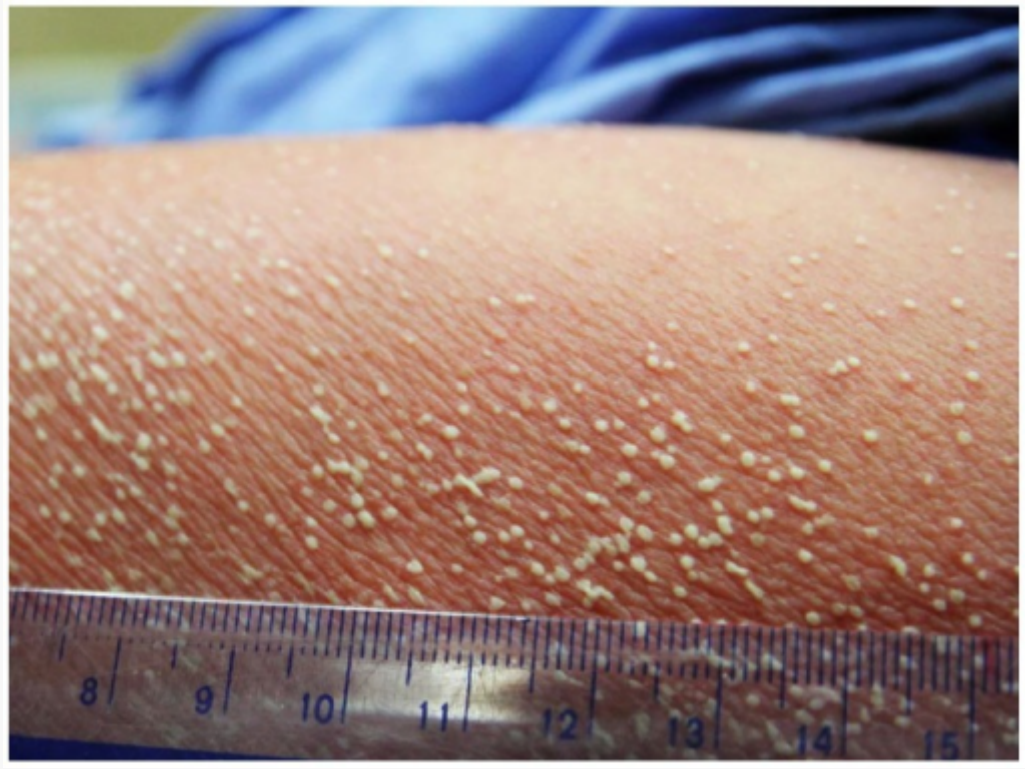


Purpura Fulminans: Purpuric patches, ecchymosis, acral necrosis (see below)

DRESS: Maculopapular rash, facial edema, hepatosplenomegaly (see below)



Acute generalized exanthematous pustulosis (AGEP) Non-follicular pustules, erythema, mucosal sparing



Red Flags: HR >120 bpm, SBP <90 mmHg, Cr >2 mg/dL, DIC, LRINEC >6

Labs and Studies

- Labs
  - CBC:Leukocytosis (fasciitis), eosinophilia (DRESS), anemia (DIC)
  - CMP: Cr (AKI), LFTs (DRESS hepatitis), hypoalbuminemia (erythroderma)
  - Coagulation: D-dimer, fibrinogen (DIC, purpura), INR
  - CRP, ESR:Elevated in infection, inflammation
- Advanced
  - HHV-6 PCR (DRESS), blood cultures (fasciitis), protein C/S (purpura)

- Imaging
  - CT: Soft Tissue Gas, fascial enhancement (fasciitis)
  - CXR: ARDS, pneumonia (fasciitis, DRESS)
  - PET-CT: Lymphoma (erythroderma)
  - Advanced: MRI (fasciitis extent), CT abdomen (DRESS organ involvement)
- Other
  - Skin Biopsy: Fasciitis (necrosis), erythroderma (psoriasis), DRESS (eosinophils), AGEP (pustules)
- LRINEC Score: Fasciitis risk (>6 high suspicion)
- Wound Cultures: Fasciitis (Streptococcus, MRSA)
- Advanced: Immunofluorescence (rule out pemphigus), ANA (autoimmune)

Diagnosis

- Criteria
  - Necrotizing Fasciitis: Severe pain, crepitus, CT/biopsy necrosis, LRINEC >6
  - Erythroderma: >90% BSA erythema, scaling, biopsy-confirmed etiology
  - Purpura Fulminans: Purpuric rash, DIC, infection/coagulopathy trigger
  - DRESS: Fever, rash, eosinophilia, organ involvement, biopsy/PCR
  - AGEP: Pustules, fever, drug history, biopsy subcorneal pustules
- Differential
  - Necrotizing fasciitis (cellulitis), erythroderma (seborrheic dermatitis), purpura (TTP), DRESS (viral exanthem), AGEP (psoriasis)

- Flowsheet:
  - **Step 1** History/Exam: Fever, rash, assess BSA, systemic signs
  - **Step 2** Labs:CBC, CMP, CRP, cultures, HHV-6 PCR
  - **Step 3** Imaging: CT (fasciitis), PET-CT (lymphoma), CXR (ARDS)
  - **Step 4** Biopsy: Confirm diagnosis, rule out mimics
  - **Step 5** Risk Stratify: LRINEC, initiate urgent treatment

Management Strategies

- General Principles
  - Stop offending drugs, provide supportive care, involve specialists (dermatology, ID, surgery)
- Supportive Care
  - IV Fluids: NS 1-2 L bolus, adjust for fluid loss
- Wound Care: Non-adherent dressings, sterile environment

- Monitoring: Vitals q2h, CMP q12h, skin assessment q8h
- Specific Therapies
    - Necrotizing Fasciitis:
        - Surgical debridement, vancomycin 15 mg/kg IV q12h, clindamycin 900 mg IV q8h
        - ID consult, repeat debridement q24-48h
    - Erythroderma
        - Topical steroids (triamcinolone 0.1%), cyclosporine 3-5 mg/kg/day
        - Treat underlying cause (e.g., lymphoma chemotherapy)
    - Purpura Fulminans
        - Heparin 80 units/kg IV bolus, FFP 10-15 mL/kg, ceftriaxone 2 g IV
        - Protein C concentrate if deficient
    - DRESS
        - Stop culprit drug, prednisone 1 mg/kg/day PO, taper over 6-8 weeks
        - Antihistamines for pruritus, monitor LFTs
    - AGEP
        - Stop culprit drug, topical steroids (betamethasone 0.05%), supportive care
        - Antihistamines, short-course prednisone if severe
- Complication Management
    - Sepsis: Antibiotics, vasopressors (norepinephrine)
    - AKI: Hydrate, hold nephrotoxins, dialysis if needed
    - DIC: FFP, platelets, heparin if thrombotic
- Monitoring
- CMP, CBC q12h (organ function, cytopenias)
- Cultures q48h (infection)
- LRINEC (fasciitis), LFTs (DRESS)

Complications

- Acute
    - Sepsis: 20-30% (fasciitis, purpura), leading mortality cause
    - AKI: 10-15% (fasciitis, DRESS), Cr rise >0.5 mg/dL
    - DIC: 10% (purpura, fasciitis), bleeding/thrombosis
- Long-Term
    - Scarring
    - Hypopigmentation (erythroderma, AGEP)
    - Amputation 5-10% (fasciitis, purpura)
    - Organ Damage
    - Hepatitis, CKD (DRESS)

- Rare
  - Lymphoma progression (erythroderma), chronic pain (fasciitis)

Clinical Scenarios

Case 1 Necrotizing Fasciitis

- Presentation: 55 y/o M with diabetes, thigh pain, fever, crepitus
- Vitals: BP 85/55, HR 130, SpO2 90%, RR 24
- Exam: Bullae, necrosis, LRINEC 9
- Labs/Studies: WBC 22K, Cr 2.2 mg/dL, CT gas in tissues
- Interpretation: Necrotizing fasciitis, surgical emergency
- Management: Vancomycin 15 mg/kg IV, clindamycin 900 mg IV, urgent debridement, ID consult, ICU monitoring. Stable by week 2

Case 2 DRESS

- Presentation: 40 y/o F with fever, rash, facial edema post-carbamazepine
- Vitals: BP 120/80, HR 110, SpO2 96%, RR 18
- Exam: Maculopapular rash, lymphadenopathy
- Labs/Studies: Eosinophils 15%, ALT 200 U/L, HHV-6 PCR positive
- Biopsy: Eosinophilic infiltrate
- Interpretation: DRESS, drug-induced
- Management: Stop carbamazepine, prednisone 1 mg/kg/day PO, Antihistamines, monitor LFTs, Rash resolves by week 3

Case 3 Purpura Fulminans

- Presentation: 25 y/o F with meningococcemia, purpuric rash, shock
- Vitals: BP 80/50, HR 140, SpO2 88%, RR 26
- Exam: Ecchymosis, acral necrosis
- Labs/Studies: D-dimer 6000 ng/mL, fibrinogen 90 mg/dL, Neisseria culture
- Interpretation: Purpura fulminans, meningococcal sepsis
- Management: Ceftriaxone 2 g IV, heparin infusion, FFP 15 mL/kg, Protein C concentrate, ICU care, Limb spared, recovery by week 3

Expert Tips

- Calculate LRINEC score for fasciitis; >6 prompts urgent surgery
- Biopsy early for DRESS, erythroderma, AGEP; guides therapy
- Stop culprit drugs in DRESS, AGEP; check HHV-6 PCR for DRESS
- Monitor coagulation in purpura fulminans; heparin, FFP critical

- Assess for lymphoma in erythroderma; PET-CT if persistent
- Pitfall
    - Misdiagnosing DRESS as viral exanthem; eosinophilia, LFTs key
- Advanced
    - Cytokine inhibitors (tofacitinib) for DRESS; genetic testing for purpura

Key Pearls

Necrotizing Fasciitis: Debridement, antibiotics, LRINEC >6

Erythroderma: Topical steroids, treat underlying cause (psoriasis, lymphoma)

Purpura Fulminans: Heparin, FFP, antibiotics for infection

DRESS: Stop drugs, prednisone, monitor HHV-6, LFTs

AGEP: Stop drugs, topical steroids, self-resolving in 2 weeks

References

UpToDate “Dermatologic Emergencies: Diagnosis and Management” (2025)

JAAD “DRESS: Advances in Diagnosis” (2024)

Crit Care Med “Necrotizing Fasciitis: Surgical Management” (2024)

Lancet “Erythroderma: A Review” (2023)

Blood “Purpura Fulminans: Coagulopathy Management” (2024)