I. INTRODUCTION

Cutaneous disorders comprise a small portion (1%–2%) of the board examination content. Pictorial identification is important because many of these questions contain an image. The lesions and rashes you are likely to be asked to identify and manage are described in this chapter. You may also wish to consult a color dermatology atlas for additional examples.

The cutaneous disorders with higher acuity that require immediate recognition and action are presented first. Those with lower acuity are presented later in the chapter.

II. GENERAL APPROACH TO THE PATIENT PRESENTING WITH A RASH

A. Inquire about prodromal symptoms, time course, and antecedent events (eg, new medications).

B. Note patient's age, immune status, past medical history, sexual history, medications, allergies, and presence/absence of toxicity.

C. Examine the rash and determine its characteristics.

1. Appearance
   a. Macular → flat and ≤1 cm
   b. Patchy → flat and >1 cm
   c. Papular → raised and ≤1 cm
   d. Plaque → raised and >1 cm
   e. Maculopapular, nodular → dermal or subcutaneous solid lesion 1–2 cm
   f. Tumor → dermal or subcutaneous solid lesion >2 cm
   g. Vesicular → blister ≤1 cm
   h. Bullous → blister >1 cm
   i. Pustules → small blister containing purulent material
   j. Scales or keratoses → built up epidermis
   k. Crusts, erosions → loss of part or all of epidermis
   l. Ulceration → loss of dermis or deeper
2. Evolution: determine where it started and how it has spread.
3. Distribution: note location of the rash, including involvement of mucous membranes, palms, and soles.
4. Symptoms: determine if pruritic or painful; note any systemic symptoms (fever, odynophagia, malaise).
5. Treatments: determine what, if anything, the patient has done to treat the rash (eg, applied topical steroids, zinc, or a neomycin-containing antibacterial ointment), because this might have changed the appearance of the rash or caused a secondary contact dermatitis.

III. STEVEN-JOHNSONS SYNDROME AND TOXIC EPIDERMAL NECROLYSIS

A. Overview

1. A drug reaction that causes sloughing of skin and mucous membranes
2. Spectrum of disease: Stevens-Johnson syndrome (SJS) → SJS/toxic epidermal necrolysis (SJS/TEN) overlap → toxic epidermal necrolysis
3. Worse outcome with increased body service area involvement, advanced age, malignancy, tachycardia, BUN >10 mEq/L, acidosis, and hyperglycemia.
4. Common inciting agents are NSAIDs, antibiotics (sulfonamides, penicillins, cephalosporins), anticonvulsants, and allopurinol.
5. Erythema multiforme is *no longer* considered to be part of this disease spectrum.

B. Clinical presentation

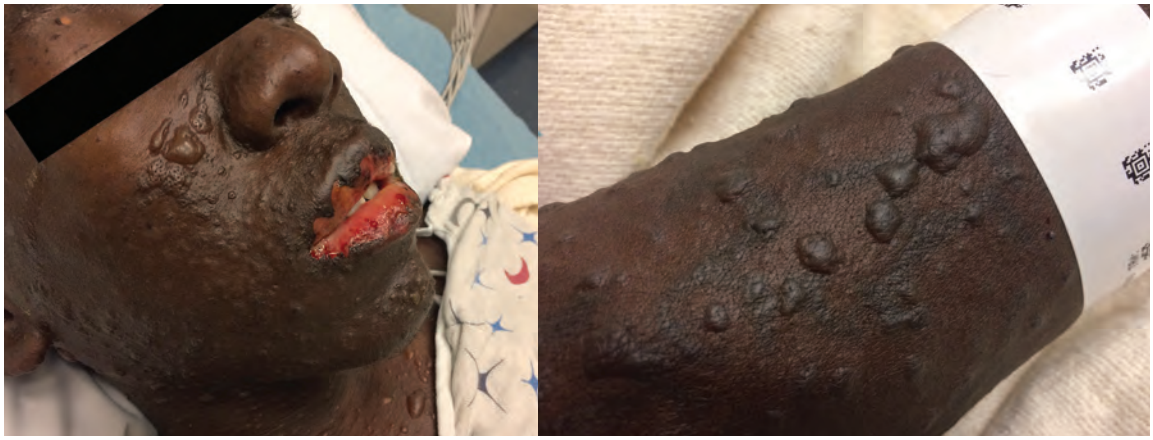
1. **Classic clinical scenario:** Very ill appearing patient with a history of fever, malaise, myalgias, and arthralgias, followed by the abrupt onset of bullous mucocutaneous lesions that subsequently eroded. Multiple mucosal surfaces (eyes, mouth, lips, urogenital area, and anus) are involved. Patients often cannot eat because of painful stomatitis. Conjunctivitis is common, and vesicles on the conjunctiva are sometimes seen; the eyelids may be red, swollen, and crusted. An overall skin examination is likely to reveal widespread erythematous (or purpuric) macules or flat atypical "targets."
2. Disease spectrum

	Body Surface Area Affected (%)	Mortality (%)
SJS	<10	1–5
SJS/TEN	10–30	6
TEN	>30	25–35

3. SJS, SJS/TEN, and TEN are all characterized by widespread bullous lesions, severe mucous membrane involvement, palm and sole involvement, and multisystem pathology.
4. Nikolsky sign is positive in all.
5. TEN is characterized by extensive cutaneous and mucosal blistering as well as desquamation; epidermis becomes necrotic and sloughs in large sheets, leaving behind exposed dermis.
6. Denuded skin and mucous membranes result in fluid loss and susceptibility to secondary bacterial infection.
7. Significant ocular sequelae (corneal ulceration, blindness) are possible.
8. Kidney involvement (hematuria, renal tubular necrosis, kidney failure) can occur but is rare.
9. A flu-like prodrome often precedes development of the mucocutaneous lesions by 1–14 days.
10. Death, when it occurs, is most often due to fulminant sepsis.

C. Management

1. Consult with a dermatologist.
2. Identify the precipitant cause (if possible) and treat accordingly. **Discontinue any drug thought to be the precipitant.**
3. Hospitalization, IV fluid resuscitation to correct hypovolemia, and correction of electrolyte abnormalities.
4. Systemic steroids are controversial. There is no hard evidence that they are of benefit, and there is some concern that they might increase complications.
5. IVIG for TEN
6. Obtain ophthalmology consult for patients with ocular lesions because of the risk of long-term morbidity from scarring.
7. Close monitoring for infections (avoid prophylactic antibiotics).
8. Consider burn unit admission.



Toxic epidermal necrolysis due to allopurinol use
Courtesy of Laura Bontempo, MD, MEd, FACEP

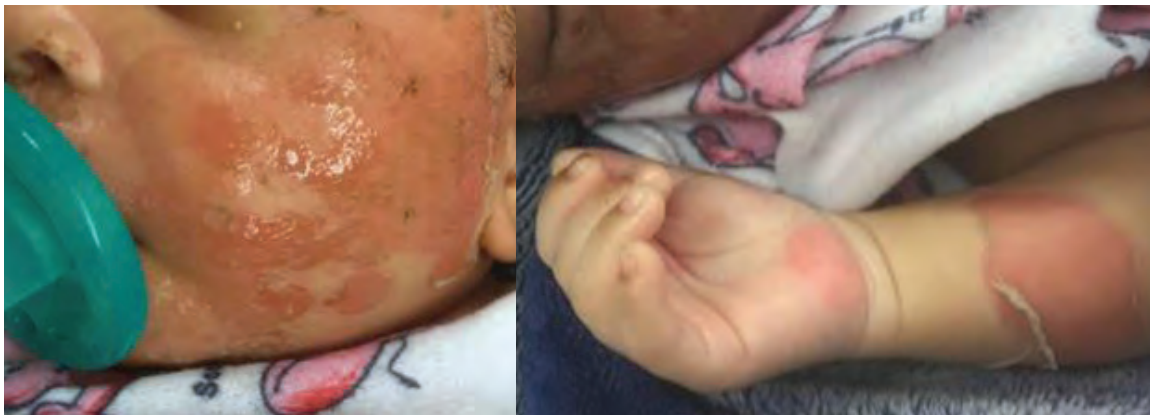
IV. STAPHYLOCOCCAL SCALDED SKIN SYNDROME

A. Overview

1. A skin infection caused by exotoxin-producing *S aureus* of phage group 2
2. Mainly affects children <6 years old
3. The mortality rate in affected adults is >60% and potentially higher if immunocompromised.

B. Clinical presentation

1. **Classic clinical scenario: 5-year-old with recent upper respiratory infection now presents with erythematous sloughing rash that started on the face and became generalized over 2 days.**
2. Often follows an upper respiratory infection or purulent conjunctivitis
3. Tender erythema of the face (perioral area is classic), neck, or axillae that generalizes over the body within 48 hours. Flaccid bullae develop and, within 48 hours, skin sloughs. Desquamation duration is 5 days.
4. Crusting around the mouth and eyes and lip fissuring are also frequently present.
5. In newborns, the entire skin surface may be involved (Ritter disease).



Staphylococcal scalded skin syndrome
Courtesy of Carmen Avendano, MD

6. Mucous membranes are *not* involved, which helps differentiate this syndrome from toxic epidermal necrolysis.
7. Nikolsky sign is positive (extension of bullae with gentle pressure).
8. Lesions usually resolve in 2 weeks without scarring.

C. Management

1. Penicillinase-resistant antibiotics to treat *S aureus*: clindamycin or oxacillin/nafcillin. Vancomycin may be considered if MRSA is a concern.
2. Steroids are contraindicated (they may exacerbate the illness).
3. Hospitalization for hydration and skin care is indicated for most patients (especially infants).
4. Treatment is similar to that for thermal burn patients; therefore, consider burn unit admission.

V. TOXIC SHOCK SYNDROME

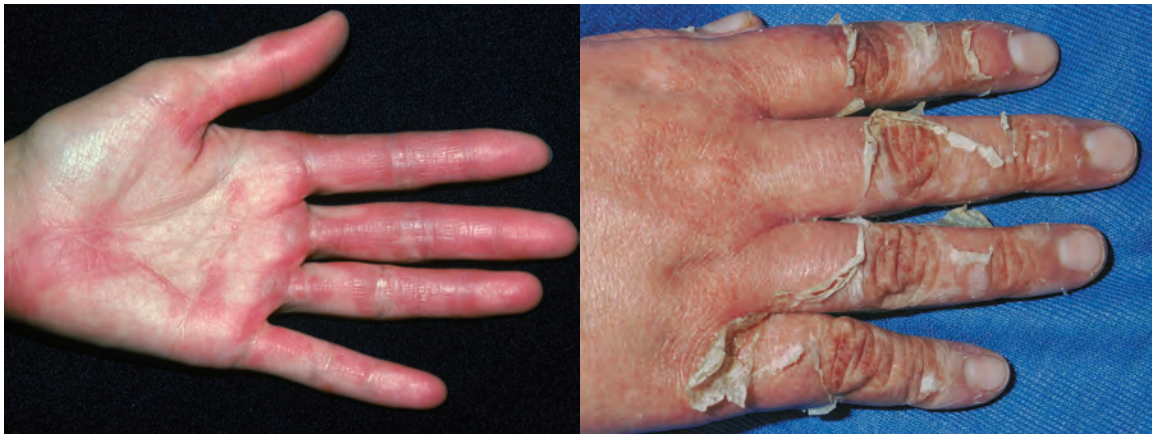
A. Overview

1. A skin infection caused by exotoxin-producing *S aureus* or group A streptococci
2. Mainly affects adults
3. Thought to be related to retained foreign bodies (high absorbency tampons, nasal packing, surgical packing)

B. Clinical presentation

1. Classic clinical scenario: A systemically ill, 23-year-old woman has mucous membrane hyperemia and a diffuse, blanching, macular erythroderma. The rash has the appearance of a first-degree sunburn, and the patient is febrile and hypotensive.
2. Diffuse erythroderma with mucous membranes and involvement of palms and sole.
3. Nikolsky sign is present.

4. Erythroderma will fade within 72 hours of appearance, then desquamate in 1–2 weeks.
5. There is no skin sloughing, which helps to differentiate from staphylococcal scalded skin syndrome.
6. More than 50% of severely ill patients experience hair and nail loss 2–3 months after initial infection.



Toxic shock syndrome showing erythema and (later) desquamation
Courtesy of Visual Diagnosis

C. Management

1. Remove foreign body, if present.
2. Resuscitate (IV fluids, vasopressors as needed).
3. Administer anti-group A β-hemolytic streptococci and MRSA antibiotics (vancomycin or clindamycin).

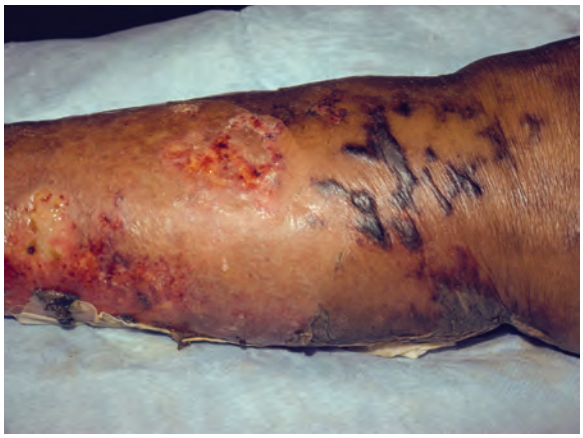
VI. NECROTIZING FASCIITIS

A. Overview

1. A bacterial infection that tracks along the fascial planes; surface erythema often underrepresents the extent of infection.
2. **Type 1 necrotizing fasciitis is polymicrobial.**
3. **Type 2 necrotizing fasciitis is monomicrobial, mostly due to group A β-hemolytic streptococci, *Vibrio vulnificus, Clostridium,* or MRSA.**
4. **Polymicrobial infections may be caused by *S aureus, streptococci, Clostridium,* Enterobacteriaceae, and *Bacteroides*.**
5. ***V vulnificus* infections are related to exposure to seawater.**
6. ***Aeromonas hydrophila* infections are associated with exposure to brackish water, soil, wood, and ditches.**
7. **Risk factors include IV drug use, skin or soft-tissue trauma, diabetes mellitus, malignancy, cirrhosis, recent surgery, peripheral artery disease, and wounds with water exposure.**
8. **Can be rapidly fatal; the mortality rate is high.**
9. **Fournier gangrene is necrotizing fasciitis of the groin, scrotum, and lower abdomen.**

B. Clinical presentation

1. Patients are systemically ill; symptoms include fever, tachycardia, lethargy, and possibly hypotension.
2. Pain and tenderness extend beyond the area of erythema and can be severe.
3. Skin may have erythema resembling cellulitis, edema, hemorrhagic bullae, and crepitus and/or necrosis, depending on the stage of disease.
4. Skin findings progress quickly.
5. Significant leukocytosis, hyponatremia, increased creatinine, and lactic acidosis may be present.



Necrotizing fasciitis
Courtesy of Visual Diagnosis

C. Diagnostic evaluation
1. Contrast-enhanced CT or MRI may show edema along fascial planes or gas in the tissue; however, a negative imaging study does *not* exclude the diagnosis.
2. Blood cultures can be helpful for long-term management.
3. Definitive diagnosis is made through wound exploration and direct tissue sampling.

D. Management
1. Early surgical consultation is essential. Surgical debridement of the necrotic tissue is the mainstay of treatment.
2. Broad-spectrum antibiotics are needed.
   a. Piperacillin-tazobactam plus vancomycin plus clindamycin (clindamycin may reduce exotoxin production by group A streptococci)
   b. Doxycycline plus ceftriaxone if *V fulnificus* or *A hydrophila* infection is suspected
3. Sepsis resuscitation with IV fluids and vasopressors as needed.
4. Admit to ICU.

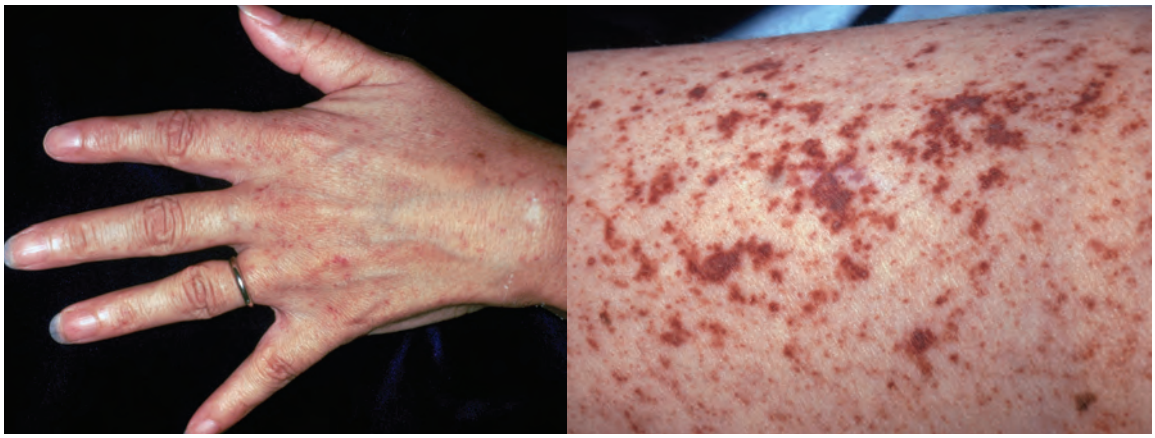
VII. ROCKY MOUNTAIN SPOTTED FEVER

A. Overview
1. A vasculitis due to *Rickettsia rickettsii;* transmitted by the *Dermacentor* tick
2. Seen throughout the US (all states)

B. Clinical presentation
1. Classic triad is fever, headache, and rash.
   a. Present the minority of the time
2. Symptoms begin days to 2 weeks after the tick bite.
3. Flu-like prodrome to the rash
4. Red papules and macules start on the distal extremities and move inward.
   a. Palms and soles are involved.
   b. The face is spared from the rash, but iritis and uveitis can develop.
5. Lesions evolve into purpuric petechia.
6. Complications include CNS (seizure, meningitis) and cardiac (myocarditis) involvement.

C. Diagnostic evaluation
1. Antibody testing (positive >1 week after infection), blood PCR or ELISA
2. Patients may have associated hyponatremia, thrombocytopenia, and increased AST

D. Management
1. Doxycycline × 5–7 days, for all ages



Rocky Mountain spotted fever. The image on the left shows petechia. The image on the right shows the evolution to purpura.
Courtesy of Visual Diagnosis

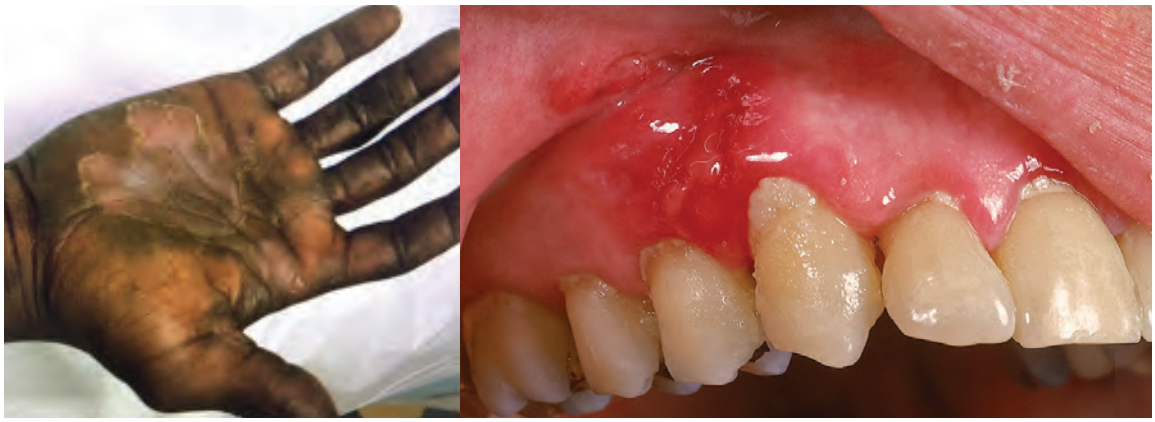
VIII. PEMPHIGUS VULGARIS

A. Overview

1. An autoimmune bullous disease of skin and mucous membranes
2. Most commonly affects patients 40–60 years old with equal incidence in both genders
3. Associated with the presence of other autoimmune diseases
4. Mortality rate is currently 5%; before the use of corticosteroid therapy, this disease was almost always fatal.

B. Clinical presentation

1. Classic clinical scenario: A 55-year-old patient with a past medical history of myasthenia gravis presents with multiple fluid-filled bullae and painful, crusted ulcers. She has a recent history of several months of oral lesions of unknown etiology. Physical examination is significant for a positive Nikolsky sign.
2. Oral lesion(s) typically precede development of cutaneous lesions by several months.
3. The cutaneous lesions are flaccid vesicles and bullae that rupture easily, leaving behind superficial erosions and crusted ulcerations. They are painful and can be seen anywhere.
4. Nikolsky sign is positive.
5. Large areas of denuded skin increase risk of secondary infection.



Pemphigus vulgaris
Courtesy of Laura Bontempo, MD, MEd (left image) and Visual Diagnosis (right image)

C. Diagnostic evaluation

1. Skin biopsy with Tzanck smear: a positive smear is suggestive but not specific.

D. Management

1. Dermatology consult
2. Supportive care

3. High-dose oral steroids (prednisone 1 mg/kg/day)
4. Concomitant immunosuppressive therapy (eg, azathioprine, mycophenolate)
5. Hospitalization for patients with extensive bullae and erosions
6. Other treatments include dapsone, rituximab, IV immunoglobulin, and plasmapheresis.

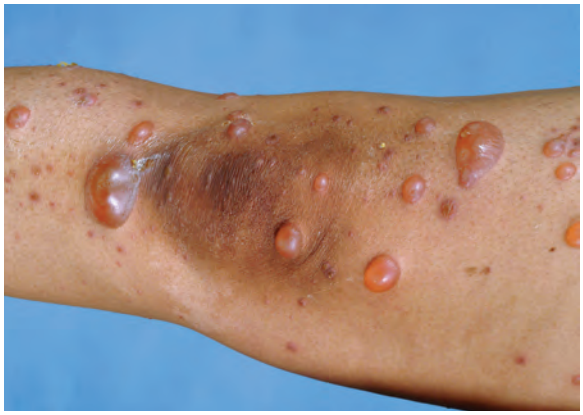
IX. BULLOUS PEMPHIGOID

A. Overview

1. An IgG autoimmune, chronic disease of the elderly
2. Associated with other autoimmune diseases
3. Duration may be months to years

B. Clinical presentation

1. Classic clinical scenario: A well-appearing 80-year-old man with history of rheumatoid arthritis presents with large bullae on the flexor surfaces of his forearms.
2. Large, tense bullae on normal skin or an erythematous base
3. Bullae are filled with clear or blood-tinged fluid.
4. Most often seen on lower abdomen and flexure surfaces of the thighs and forearms
5. Typically, there is no mucosal involvement.
6. No Nikolsky sign (lesions do not erode or spread with light pressure)



Bullous pemphigoid
Courtesy of Visual Diagnosis

C. Management

1. Localized disease: high-potency topical corticosteroids
2. Extensive disease: prednisone, 1 mg/kg/day
3. Dermatology might consider IVIG for resistant cases.

X. EXFOLIATIVE DERMATITIS/ERYTHRODERMA

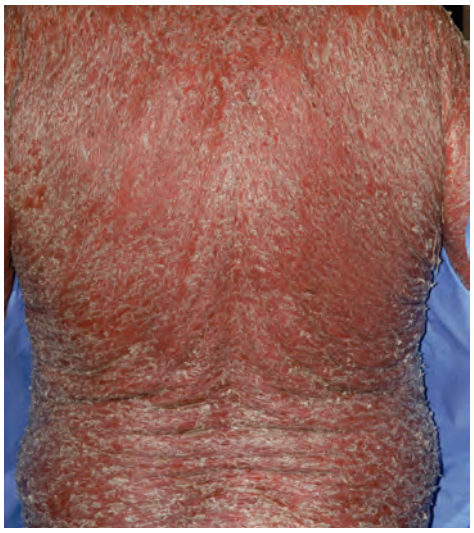
A. Overview

1. Widespread erythematous, pruritic dermatitis covering >90% of the body surface area
2. Etiology
   a. Idiopathic is most common form.
   b. Exacerbation of preexisting dermatoses (eg, psoriasis, eczema, seborrhea)
   c. Drug-induced (>50 drugs have been implicated)
   d. Underlying malignancy (cutaneous lymphoma, leukemia, or other lymphoreticular malignancy) or immunosuppression (HIV)
   e. Allergic contact dermatitis

3. Affects both genders
4. Adults (40–60 years old) are predominantly affected, but children can be affected also.
5. Mortality rate is as high as 30%.

B. Clinical presentation

1. Classic clinical scenario: A 57-year-old man who has a past medical history of psoriasis complains of itching, chills, and "tightness" of the skin. He has a low-grade fever and is hypotensive with tachycardia. On examination, a scaly, warm, erythematous rash is found to be covering nearly all his body surface area. The rash is not tender to the touch. There is no oral involvement, and the Nikolsky sign is negative.
2. Rash is shiny, erythematous, and pruritic with scaling. It begins localized then spreads and generalizes.
3. Palms and soles are spared.



Erythroderma
Courtesy of Visual Diagnosis

4. Other findings can include fever or hypothermia, dehydration, lymphadenopathy, hepatosplenomegaly, lower extremity edema, or gynecomastia.
5. Scratching can result in lichenification and erosions.
6. Because of increased blood flow to the skin, the patient might have high output heart failure.

C. Diagnostic evaluation

1. Consider CBC, serum chemistries, liver function tests, erythrocyte sedimentation rate, urinalysis, and HIV testing in the search for systemic causes.
2. Obtain skin biopsy (lymph node biopsy if significant lymphadenopathy is present).

D. Differential diagnosis

1 Erythema multiforme
2. Toxic epidermal necrolysis
3. Toxic shock syndrome
4. Staphylococcal scalded skin syndrome
5. Kawasaki disease (children)

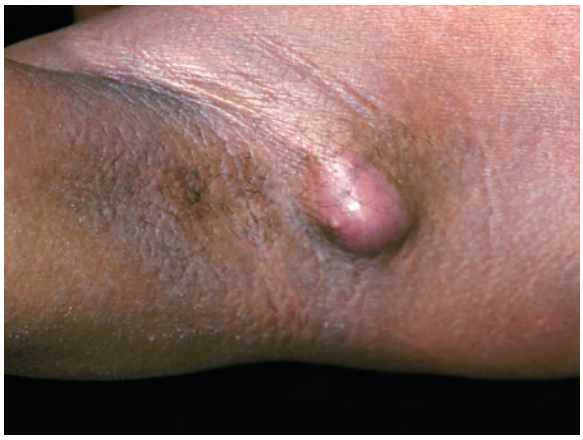
E. Management

1. Goal is to correct/eliminate the underlying cause while providing symptomatic relief and maintaining skin moisture.
2. Stop new medications, if at all possible.
3. Antihistamines
4. Topical steroids covered with an occlusive dressing and continued for weeks or months.
5. Warm water baths with bath oils and skin emollients are also helpful.
6 Patients with severe or resistant disease are treated with systemic corticosteroids.
7. Because this disorder is usually the result of an underlying cutaneous disease, a systemic disease, or a drug or chemical reaction, patients should be admitted for further investigation into the etiology.

XI. CUTANEOUS ABSCESS

A. Overview

1. A cutaneous abscess is a localized collection of pus with associated pain, a fluctuant mass, and erythema.
2. Most abscesses contain *S aureus*: MRSA most commonly followed by coagulase-negative staphylococcus.
3. IV drug abuse abscesses ("shooters abscess") may be polymicrobial.
4. The presence of fever or other systemic symptoms suggests possible bacteremia.
5. A furuncle is a localized abscess associated with a hair follicle. A carbuncle results if several furuncles coalesce and interconnect by sinus tracts. It may develop in areas of thick skin such as the back of the neck.



Courtesy of Visual Diagnosis

B. Clinical presentation

1. Usually a singular, fluctuant mass with tenderness to palpation. There may be associated erythema, thinning of the overlying skin, and purulent drainage.
2. The development of cutaneous abscess depends on location. On the extremities, the cause is usually minor trauma that damages the integrity of the epithelium. In intertriginous regions, abscesses are associated with obstructed apocrine sweat glands.
3. The abscess is often surrounded by a rim of erythema and induration.
4. Erythema extending beyond the rim of the abscess indicates purulent cellulitis.

C. Management

1. Abscess size and depth can be evaluated by ultrasound.
2. Incision and drainage are the mainstay of therapy: culture of the drained fluid is rarely indicated unless the abscess is recurrent.
3. Irrigation and packing of the abscess are not necessary, but using a loop drain may be considered.
4. Although controversial, antibiotics appear to be beneficial.
   a. Trimethoprim-sulfamethoxazole (first-line), doxycycline, or clindamycin
   b. Duration of therapy is 7–10 days.
   c. The Infectious Diseases Society of America recommends antibiotics for patients who are febrile, immunocompromised, or have an abscess larger >5 cm or associated cellulitis.

XII. URTICARIA

A. Overview

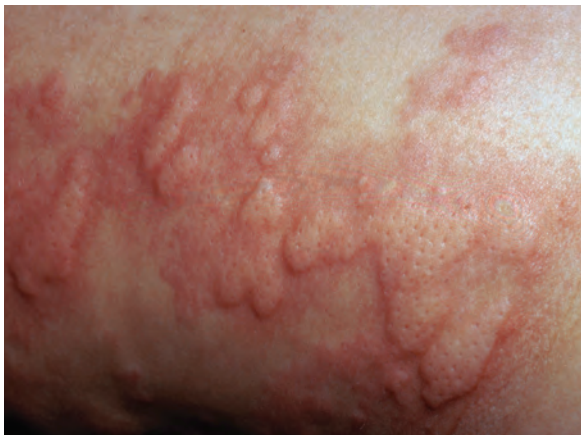
1. Also known as hives
2. Usually due to an IgE-mediated allergic reaction

B. Clinical presentation

1. Raised, very pruritic plaques
2. Lesions vary in shape and size and are transient (minutes to hours).

C. Management

1. Symptomatic treatment with an antihistamine (eg, diphenhydramine)
2. Steroids for diffuse disease
3. Attempt to identify the trigger through detailed history.
4. Epinephrine IM for severe reactions or symptoms consistent with anaphylaxis (multisystem symptoms, hemodynamic instability)
5. Outpatient follow-up with primary care and/or an allergist



Urticaria
Courtesy of Visual Diagnosis

XIII. CELLULITIS/ERYSIPELAS

A. Overview

1. Cellulitis is a local soft-tissue inflammatory reaction secondary to bacterial invasion of the skin resulting from compromised skin integrity, often from occult trauma.
2. Cellulitis is classically associated with comorbidities such as diabetes, obesity, lymphedema/edema, lower extremity ulcers, peripheral vascular disease, and dermatitis.
3. Most infections involve the extremities.
4. In immunocompetent patients, cellulitis is most commonly caused by β-hemolytic streptococci (groups A, B and C) or *S aureus* (methicillin-sensitive).
5. In immunocompromised patients, the cause is most commonly *S pneumoniae* or *E coli*.
6. **Cellulitis secondary to specific causes has different pathogens:**
   a. **Dog or cat bite: *Pasteurella multocida***
   b. **Human bites: *Eikenella corrodens***
   c. **Fresh or saltwater exposure: *Vibrio vulnificus, Aeromonas hydrophila***
   d. **IV drug use: MRSA**
7. Erysipelas is a nonpurulent cellulitis involving only the superficial dermis and lymphatics. It is caused by β-hemolytic streptococci.

B. Clinical presentation

1. Classic presentation: Otherwise healthy patient presents with lower extremity pain, edema, redness, and warmth with tenderness to palpation.
2. The classic symptoms are the result of a localized inflammatory reaction.
3. Most commonly occurs on the extremities, with circumferential erythema.
4. There may be associated regional lymphadenopathy and/or lymphangitis.