# CUTANEOUS DISORDERS

## I. INTRODUCTION

Cutaneous disorders comprise a small portion (3%) of the board exam 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, antecedent events (eg, new medications), and the presence or absence of systemic symptoms (eg. fever, myalgias).**

**B. Note patient's age, immune status, past medical history, sexual history, social history (including the use of IV drugs), 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. For blistering lesions, note the presence or absence of the Nikolsky sign. A rash is Nikolsky positive if, when pressure is applied to an intact blister, the blister spreads as opposed to ruptures. A positive Nikolsky sign indicates a dangerous dermatologic condition.
3. Evolution: determine where it started, how it has spread and time course of its evolution.
4. Distribution: note location of the rash, including involvement of mucous membranes, palms, and soles.
5. Symptoms: determine if pruritic or painful; note any systemic symptoms (fever, odynophagia, malaise).
6. 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. STEVENS-JOHNSON 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 (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, sulfonamides (sulfasalazine), antibiotics (penicillins, cephalosporins, quinolones), anticonvulsants, and allopurinol; can be precipitated by acetaminophen use.
5. Incidence is increased in patients with HIV.
6. Erythema multiforme is no longer considered to be part of this disease spectrum.

### B. Clinical presentation

1. Classic clinical scenario: Very ill appearing patient, recently started on a new medication, who presents 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, crusted and painful. An overall skin examination is likely to reveal widespread erythematous (or purpuric) macules or flat atypical "targets."
2. A flu-like prodrome often precedes development of the mucocutaneous lesions by several days.

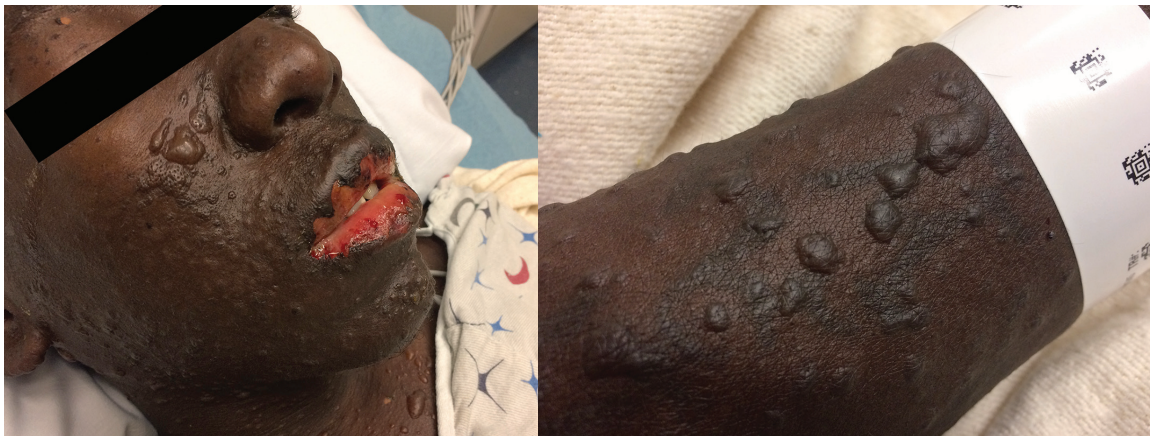
**Table 54:** Disease Spectrum

| | 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. The bullae evolve into painful erosions.
4. Nikolsky sign (extension of bullae with gentle pressure) 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. 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. Check kidney function (BUN/creatinine), heart function (ECG), and lung function (chest radiograph for infiltrates).
8. Close monitoring for infections (avoid prophylactic antibiotics).
9. Consider burn unit admission.



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

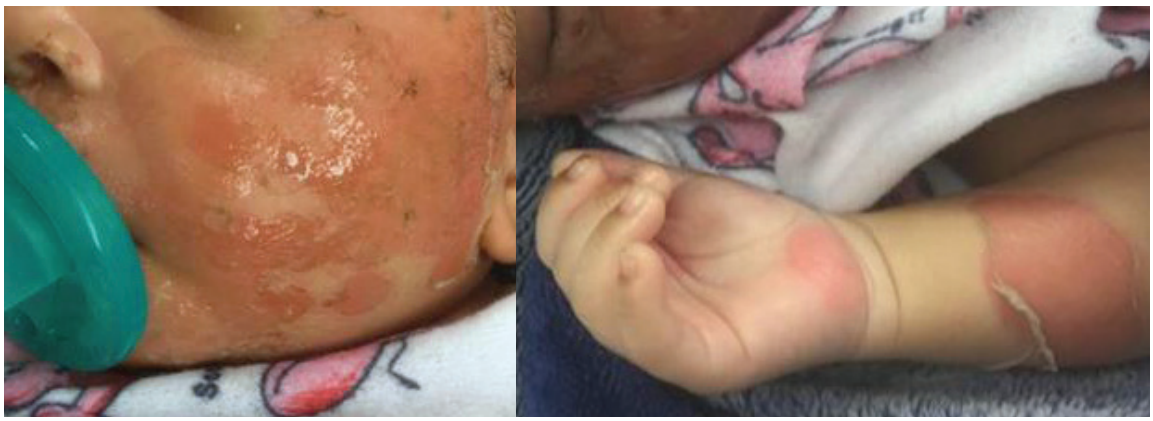
## IV. STAPHYLOCOCCAL SCALDED SKIN SYNDROME

### 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 an erythematous rash that started on the face and then became generalized over 2 days. Rash has now become bullous and is sloughing.
2. Often follows an upper respiratory infection or purulent conjunctivitis.
3. Prodrome includes generalized fussiness, malaise, and fever.
4. 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.
5. Crusting around the mouth and eyes and lip fissuring are also frequently present.
6. In newborns, the entire skin surface may be involved (Ritter disease).



Staphylococcal scalded skin syndrome
*Courtesy of Carmen Avendano, MD*

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

### C. Management

1. Penicillinase-resistant antibiotics to treat S aureus: clindamycin, oxacillin, or 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. There is no benefit to culturing the fluid from the bullae; it is sterile.
5. Treatment is similar to that for thermal burn patients; therefore, consider burn unit admission.

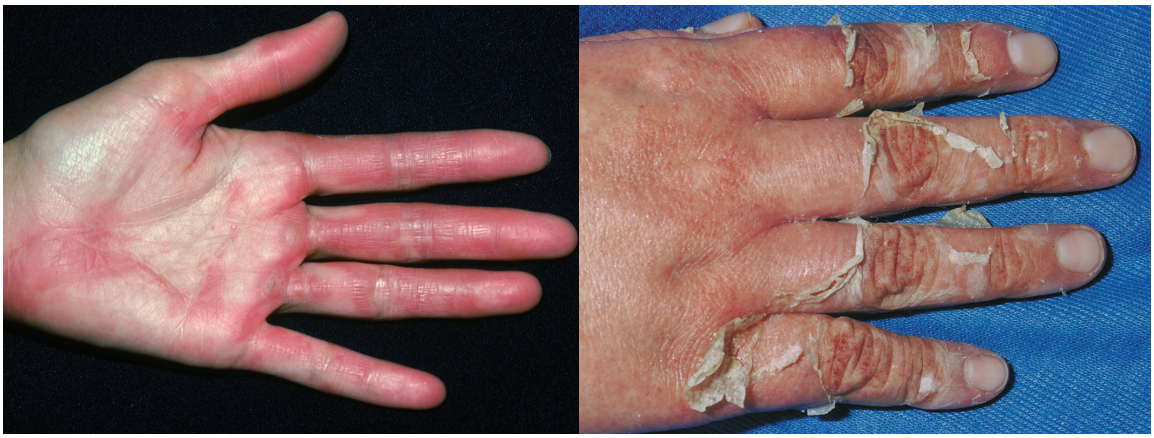
## V. TOXIC SHOCK SYNDROME

### A. Overview

1. A skin infection caused by exotoxin-producing *Staphylococcus aureus* or group A strep (*Streptococcus pyogenes*)
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 (including palms and soles) with possible mucous membrane involvement.
3. Nikolsky sign is absent.
4. Flu-like prodrome with fever that rapidly progresses to rash, sepsis, and organ failure.
5. Multisystem organ failure can involve the kidneys (elevated creatinine), coagulation (thrombocytopenia, DIC), liver (transaminitis), and lungs (respiratory distress, infiltrates).
6. Erythroderma will fade within 72 hours of appearance, then desquamate in 1–2 weeks.
7. There is no skin sloughing, which helps to differentiate from staphylococcal scalded skin syndrome.
8. 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

*Image appears with permission from VisualDx (www.visualdx.com)*

### C. Management

1. Remove foreign body, if present.
2. Sepsis resuscitation (IV fluids, vasopressors as needed, blood cultures)
3. Administer anti-group A β-hemolytic streptococci and anti-MRSA antibiotics, such as vancomycin or linezolid. Clindamycin should be added to suppress toxin production.

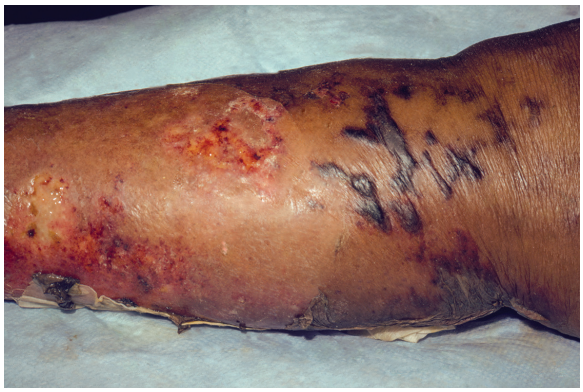
# VI. NECROTIZING FASCIITIS

## 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, MRSA, *Vibrio vulnificus*, or *Clostridium*.
4. Polymicrobial infections may be caused by gram-positive, gram-negative, and anaerobic bacteria, including *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.
10. Approximately 80% of cases are associated with some form of skin trauma, including very minor injuries or procedures.

## 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. Classically, patients have pain out of proportion to their examination.
4. Skin may have erythema resembling cellulitis, edema, hemorrhagic bullae, crepitus, and/or necrosis, depending on the stage of disease. The presence of hemorrhagic bullae is highly suggestive of necrotizing fasciitis.
5. Skin findings progress quickly.
6. Significant leukocytosis, hyponatremia, increased creatinine, lactic acidosis, and increased inflammatory markers may be present.
7. Patients with diabetes mellitus, alcoholism, and/or cirrhosis are at higher risk of this infection.



Necrotizing fasciitis

*Image appears with permission from VisualDx (www.visualdx.com)*

## 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.
4. Patient may have hyponatremia, lactic acidosis.

## 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*

   b. Vancomycin or linezolid *plus*
   c. Clindamycin (clindamycin may reduce exotoxin production by group A streptococci)
   d. Add 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) but most common in the southeastern and south central US.
3. Transmission occurs quickly following the bite of an infected tick. Therefore, transmission can occur without the patient being aware of the exposure.
4. Occurrence is more common in the summer because of the increased risk of tick exposure.

### B. Clinical presentation

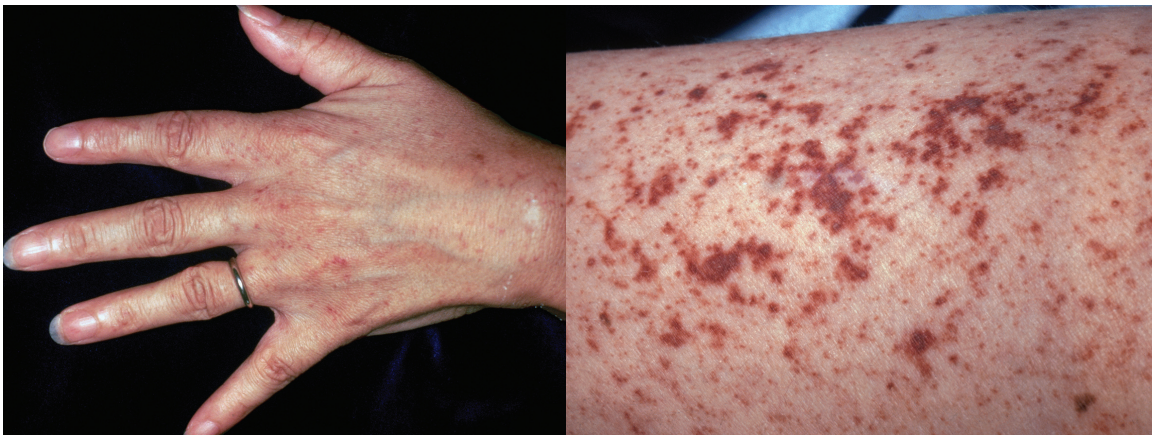
1. Classic triad is fever, headache, and petechial rash.
   a. Present the minority of the time
2. Symptoms begin days to 2 weeks after the tick bite.
3. Flu-like prodrome to the rash.
4. Red papules and macules start on the distal extremities (wrists and ankles) and move inward.
   a. Palms and soles 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:
   a. CNS (headache, seizure, meningitis, mental status change)
   b. Cardiac (myocarditis) involvement.
   c. GI (hepatitis)
   d. Cardiac (dysrhythmias)

### C. Diagnostic evaluation

1. IgM and IgG antibody testing, blood PCR, or ELISA. Antibody tests may be negative during the first week of symptoms.
2. Patients may have associated hyponatremia, thrombocytopenia, and increased AST

### D. Management

1. Doxycycline × 7–10 days, for all ages. Treatment should continue for a minimum of 3 days after fever resolves.
2. Treat based on clinical suspicion, not confirmatory testing.
3. Without antibiotic treatment, mortality is 20%–30%.



Rocky Mountain spotted fever. The image on the left shows petechia. The image on the right shows the evolution to purpura.
*Image appears with permission from VisualDx (www.visualdx.com)*

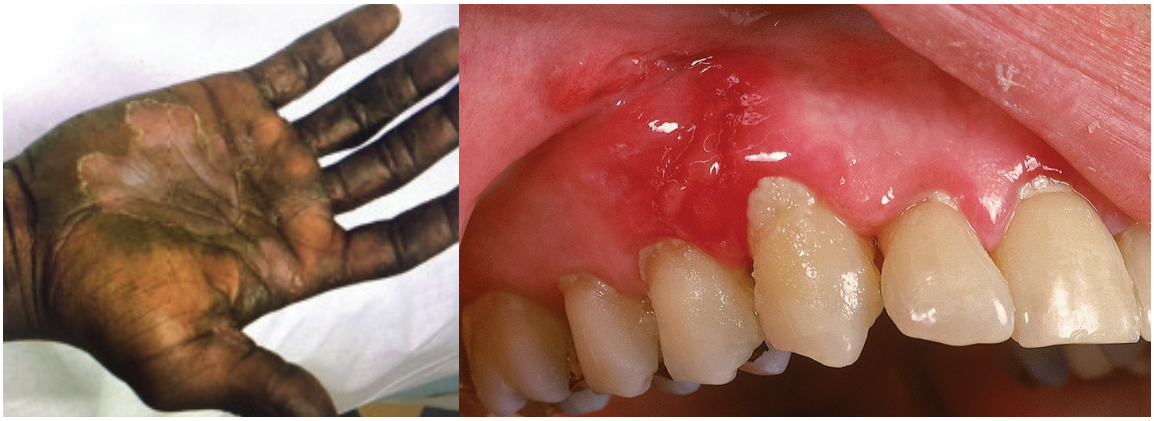
# VIII. PEMPHIGUS VULGARIS

### 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. The patient has a recent history of several months of painful oral blisters 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. Mucosal lesions can affect any mucosal surface, including the conjunctiva, larynx, pharynx, and genitals.
5. Nikolsky sign is positive.
6. Large areas of denuded skin increase risk of secondary infection.



Pemphigus vulgaris

*Image on left courtesy of Laura Bontempo, MD, MEd, FACEP*

*Image on right appears with permission from VisualDx (www.visualdx.com)*

### 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); response to treatment may take weeks.
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.

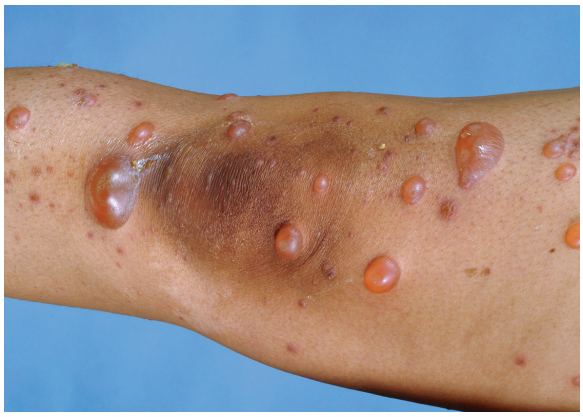
# IX. BULLOUS PEMPHIGOID

### A. Overview

1. An IgG autoimmune, chronic disease of older adults
2. Associated with other autoimmune diseases
3. Duration may be months to years
4. May occur in young patients if it is drug-induced, as opposed to autoimmune.

### 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. The rash was preceded by pruritus.
2. Large (up to 4 cm), 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, flexure surfaces of the forearms, medial thighs, and axillae.
5. Typically, there is no mucosal involvement.
6. No Nikolsky sign (lesions do not erode or spread with light pressure)
7. The patient is systemically well.



Bullous pemphigoid

*Image appears with permission from VisualDx (www.visualdx.com)*

### C. Diagnostic evaluation

1. Diagnosis is clinical.
2. Can be confirmed with direct immunofluorescence studies and ELISA testing

### D. Management

1. Localized disease: high-potency topical corticosteroids (eg. clobetasol)
2. Extensive disease: prednisone, 1 mg/kg/day for ≥6 months
3. Dermatology might consider immunosuppressants or IV immunoglobulin for resistant cases.

## X. EXFOLIATIVE DERMATITIS / ERYTHRODERMA

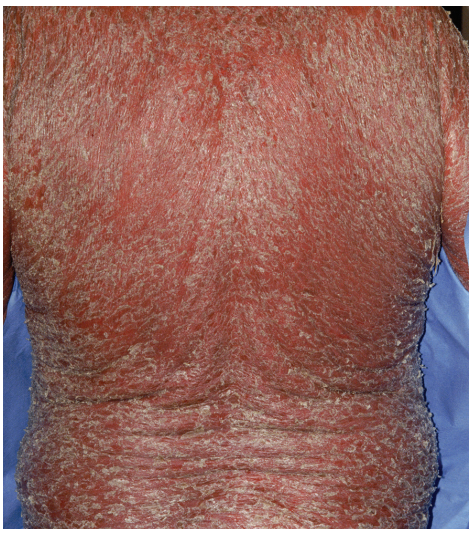
### A. Overview

1. Widespread erythematous, pruritic, scaling dermatitis covering >90% of the body surface area
2. Etiology
   a. Idiopathic is the 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 has symptoms 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 white or yellow scaling. It begins localized then spreads and generalizes.
3. Palms and soles are spared.



Erythroderma

*Image appears with permission from VisualDx (www.visualdx.com)*

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. This is a clinical diagnosis.
2. Consider CBC, serum chemistries, liver function tests, erythrocyte sedimentation rate, urinalysis, and HIV testing in the search for systemic causes.
3. There may be associated anemia, leukocytosis, increased reactive markers, and eosinophilia.
4. Obtain skin biopsy (lymph node biopsy if significant lymphadenopathy is present) for direct immunofluorescence.

### D. Differential diagnosis

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

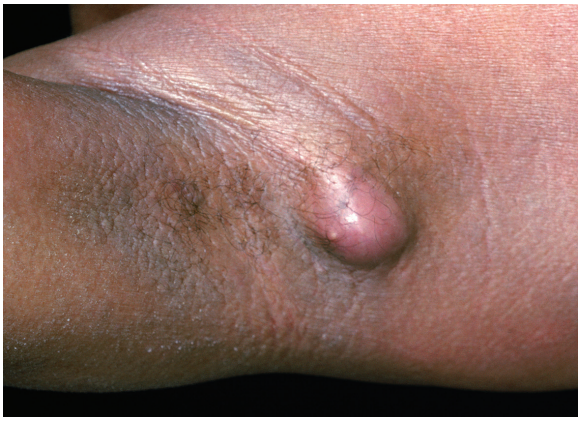
### E. Management

1. Goal is to correct/eliminate the underlying cause, to provide symptomatic relief and supportive care, and to maintain skin moisture.
2. Stop new medications, if at all possible.
3. Antihistamines for pruritus.
4. Topical steroids covered with an occlusive dressing; continue 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 in the dermis and subcutaneous space with associated pain, erythema, and a fluctuant mass..
2. Most abscesses contain *Staphylococcus aureus*: MRSA most commonly followed by coagulase-negative staphylococcus.
3. IV drug use abscesses ("shooter's 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.



Cutaneous abscess of the axilla

*Image appears with permission from VisualDx (www.visualdx.com)*

## 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. Can occur anywhere but is most commonly seen in the axillae, groin, buttocks, and extremities.
3. The development of a 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.
4. The abscess is often surrounded by a rim of erythema and induration.
5. Erythema extending beyond the rim of the abscess indicates purulent cellulitis.

## C. Diagnostic evaluation

1. This is a clinical diagnosis.
2. Ultrasound can be used to confirm the diagnosis and evaluate the size and depth of the abscess.

## D. Management

1. Incision and drainage are the mainstay of therapy: culture of the drained fluid is rarely indicated unless the abscess is recurrent.
2. Irrigation and packing of the abscess are not necessary, but using a loop drain may be considered.
3. 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, have an abscess larger >5 cm, or if there is associated cellulitis.