Answers and Critiques

Item 1  Answer:  A

Educational Objective:  Diagnose basal cell carcinoma.

Basal cell carcinoma (BCC) typically presents as a pearly, pink papule or nodule with telangiectatic vessels. As BCC grows, the central area often ulcerates, resulting in its characteristic rolled edge. Flecks of melanin pigment are commonly present. A biopsy is necessary, as amelanotic melanoma may have a similar appearance. Common biopsy techniques include shave or punch. Most nodular BCCs are treated with excision, whereas ill-defined lesions, high-risk histologic types, and tumors on the face and hands are treated with Mohs micrographic surgery. Selected superficial lesions can be treated with curettage, imiquimod, cryotherapy, or excision.

Pyogenic granulomas are typically bright red and friable, are commonly crusted, and develop over a few days to weeks. Removal is necessary only if the lesion is cosmetically unacceptable, painful, causes unwanted bleeding, or is otherwise bothersome.

Seborrheic keratoses is a painless, nonmalignant growth appearing as a waxy, brownish patch or plaque. Seborrheic keratoses lack a pearly appearance and typically exhibit horn cysts (epidermal cysts filled with keratin) on the surface that can best be visualized with a magnifying lens. Treatment is necessary only if lesions are symptomatic or interfere with function.

Squamous cell carcinomas are rapidly growing, hyperkeratotic, ulcerated macules, papules, or nodules that commonly appear on the scalp, neck, and pinna. A shave or punch biopsy is used to confirm the diagnosis of suspicious lesions.

**KEY POINT**

- Basal cell carcinomas present as pink, pearly nodules with telangiectases and, commonly, flecks of melanin pigment.

Bibliography

Item 2  Answer:  B

Educational Objective:  Understand the skin complications associated with the use of topical corticosteroids.

Clobetasol propionate 0.05% is an ultra-high-potency corticosteroid. Potential cutaneous complications associated with the use of topical corticosteroids include thinning of the skin, development of striae (stretch marks), development of purpura, pigmented changes (hypo- or hyperpigmentation), aeniiform eruptions, and increased risk of infections. Striae formation has been documented in 1% or more of patients using a mid-potency corticosteroid; the incidence may be higher with the use of more potent agents. The risk increases when corticosteroids are used for prolonged periods, are applied under occlusion, or are applied in skin folds where there is natural occlusion, as in this patient.

Calcipotriene, a vitamin D analog, inhibits proliferation of keratinocytes, normalizes keratinization, and inhibits accumulation of inflammatory cells (neutrophils and T-lymphocytes). Calcipotriene's efficacy is comparable to that of medium-strength topical corticosteroids, but the drug is not associated with the cutaneous side effects seen in this patient.

Phototherapy induces T-lymphocyte apoptosis and therefore decreases proinflammatory cytokines. The most commonly reported side effects include photoaging, cataracts, and skin cancer. Severe cutaneous atrophy with striae formation is not a side effect of phototherapy.

Topical tar compounds are frequently used as corticosteroid-sparing drugs for patients with refractory psoriasis and are associated with excellent results when combined with ultraviolet B phototherapy. Coal tar products do not result in thinning of the skin.

**KEY POINT**

- Potential side effects of topical corticosteroids include development of striae and atrophy of the skin.

Bibliography

Item 3  Answer:  B

Educational Objective:  Diagnose a neuropathic ulcer.

This patient has a neuropathic ulcer. Neuropathic ulcers are common in patients with diabetes mellitus who have severe peripheral neuropathy. Neuropathic ulcers occur at pressure points such as the plantar aspect of the foot in the region of the metatarsal heads. The ulcers characteristically have a thick surrounding zone of hyperkeratosis, and accompanying deformities of the foot, including hammer toes and a flattened foot arch, are common.
A comprehensive foot examination should be performed annually in all patients with diabetes to identify high-risk foot problems. Early detection of sensory deficits in the foot alerts the patient to take extra care in compulsive visual inspection to detect early changes. Meticulous self-care, including daily washing, rehydration with emollient creams, and the use of comfortable, protective, and well-fitting shoes and plain cotton socks, is recommended. Patients should also avoid walking barefoot. Particular attention should be paid to the feet after purchasing new footwear so that evidence of new pressure points may be ascertained. Callus formation, evidence of irritation from improperly fitting shoes, and unrecognized foreign bodies on the foot are of particular importance.

Ischemic ulcers are typically distal and are not associated with surrounding hyperkeratosis. The skin is thin and atrophic. Pulses and capillary refill are poor.

Vasculitic ulcers are irregular in shape and have a punched-out appearance. They lack surrounding hyperkeratosis and rarely occur over pressure points.

Venous stasis ulcers are associated with chronic venous insufficiency and typically occur on the medial aspect of the lower leg, especially over the medial malleolus. Hemorrhagic staining is typically prominent.

**KEY POINT**
- Neuropathic ulcers occur over pressure points, usually on the plantar surface of the foot, and commonly have surrounding hyperkeratosis.

Bibliography

**Item 4 Answer: A**

**Educational Objective:** Diagnose drug reaction with eosinophilia and systemic symptoms (DRESS).

This patient has a drug reaction characterized by a generalized poplar eruption, eosinophilia, and systemic symptoms and is consistent with drug reaction with eosinophilia and systemic symptoms (DRESS). With cessation of the causative drug, most likely sulfasalazine, the skin reaction rapidly subsides along with lymphadenopathy, fever, elevated aminotransferase levels, and eosinophilia. Patients with DRESS may develop severe hepatitis, and fulminant hepatic necrosis may occur if the condition is unrecognized.

Erythema multiforme (EM) is an acute, often recurrent mucocutaneous eruption that usually follows an acute infection, most frequently recurrent herpes simplex virus infection, but it may also be drug related or idiopathic. Lesions range in size from several millimeters to several centimeters and consist of erythematous plaques with concentric rings of color. Patients may have low-grade fever during an EM outbreak. However, this patient’s skin lesions are not consistent with EM, and EM does not cause lymphadenopathy, aminotransferase elevations, or eosinophilia.

The reactions that are generally classified within the spectrum of severe cutaneous adverse reactions include acute generalized exanthematous pustulosis, Stevens-Johnson syndrome/toxic epidermal necrolysis, DRESS, and vasculitis. Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) are characterized by fever, skin pain, and mucocutaneous lesions resulting in epidermal death and sloughing. The clinical difference between SJS and TEN is the severity and percentage of body surface involved. SJS involves less than 10% of body surface area, SJS/TEN overlap involves 10% to 30%, and TEN involves greater than 30%. This patient’s skin lesions are not compatible with SJS or TEN, and these conditions cannot explain the patient’s other systemic findings.

**KEY POINT**
- Drug reaction with eosinophilia and systemic symptoms (DRESS) is a serious cutaneous adverse reaction characterized by a generalized papular eruption, facial edema, fever, arthralgia, and generalized lymphadenopathy and is commonly associated with elevated aminotransferase levels, eosinophilia, and lymphocytosis.

Bibliography

**Item 5 Answer: D**

**Educational Objective:** Diagnose squamous cell carcinoma in a renal transplant recipient.

Long-term immunosuppression to prevent allograft rejection increases the risk of malignancy about 100 times that of the general population. The most common posttransplant cancers involve the skin, including the lips. Skin cancers in transplant recipients differ from those in the general population. Squamous cell carcinoma is more common than basal cell carcinoma in transplant recipients; transplant recipients develop lesions at an earlier age and at multiple sites; squamous cell carcinomas are usually associated with multiple warts and premalignant keratoses, such that their appearance may be misleading; and tumors in transplant recipients are more aggressive and are more likely to recur after resection. Superficial tumors can be managed with excision, and, in select instances, destructive methods including cryotherapy or electrodesiccation and curettage. Invasive cancers require excision with margin evaluation.

Kaposi sarcoma usually manifests as a purple nodule or plaque predominantly affecting the legs in persons of Mediterranean, Jewish, Arabic, Caribbean, or African descent, possibly related to the geographic distribution of human herpesvirus 8.
Melanoma may account for up to 6% of posttransplant skin cancers in adults, but the clinical features in this patient are not consistent with melanoma.

Psoriasis may occur in transplant recipients, but this patient’s lesion is not typical of psoriasis, which typically presents as a thick, silvery scale on an erythematous plaque.

**KEY POINT**
- The most common cancers in transplant recipients involve the skin, particularly squamous cell carcinoma; these cancers are more aggressive than those in the general population.

**BIBLIOGRAPHY**

**Item 6**
**Answer:** C

**Educational Objective:** Diagnose chigger bites.

The correct diagnosis is chigger bites. The history of outdoor activity and the finding of erythematous excoriated papules along clothing lines, occasionally with a central punctum, are consistent with the diagnosis of chigger bites. Appropriate therapy consists of a mid-strength topical corticosteroid. Camphor and menthol lotion is helpful for the symptomatic relief of pruritus associated with insect bites.

If a patient asked the pharmacist for a camphor and menthol lotion, it is likely he or she would receive Sarna® lotion, as it is the most common brand of camphor and menthol lotion available. “Sarna” is also a Spanish word for scabies. In fact, the product is widely used in Latin countries for the symptomatic relief of pruritus associated with scabies. For Spanish-speaking patients, forewarning the patient about the possibility of receiving a product labeled “Sarna” can prevent concerned revisits or office telephone calls in this situation.

Bedbug bites occur most frequently on exposed sites such as the face, neck, arms, and hands and present as pruritic, urticaria-like papules arranged in a row with a central hemorrhagic punctum. The patient’s outdoor activities and the distribution of the bites along the sock line make this diagnosis unlikely.

Brown recluse spider bites are rare and are most likely to present as rapidly progressive, necrotic skin lesions. Furthermore, brown recluse spiders are not found in Florida, the Southeast coast, the mid-Atlantic or northern states, or the far West. In the United States, brown recluse spiders are located in the central and south central regions.

Scabies infestation causes intense itching, a papular or vesicular rash, and subcutaneous burrows that are most often found in the interdigital webs, flexure surface of the wrists, penis, axillae, nipples, umbilicus, scrotum, and buttocks. This patient’s lesions do not have these characteristics.

**KEY POINT**
- A history of outdoor activity and a finding of erythematous, excoriated papules along clothing lines, occasionally with a central punctum, are consistent with the diagnosis of chigger bites.

**BIBLIOGRAPHY**

**Item 7**
**Answer:** C

**Educational Objective:** Diagnose herpes simplex infection (herpetic folliculitis).

Folliculitis is a superficial or deep infection or inflammation limited to the hair follicles. This patient has herpetic folliculitis. The figure reveals multiple, clear, fluid-filled vesicles, which are the characteristic primary lesions of herpetic skin infections. This patient most likely shaved through the active lesion of his recurrent herpes labialis, spreading the infection across his beard area. These vesicular lesions will erode and crust over. Treatment options include a course of antiviral drugs such acyclovir, valacyclovir, or famciclovir.

There are no comedones present, as one would expect in acne vulgaris. Furthermore, acne is not generally tender, and an extensive eruption appearing in 1 day would be unusual for acne vulgaris.

*Pseudomonas* folliculitis primarily affects the trunk. The papules and pustules characteristically have a small, pink flare and are pruritic, and there is usually a history of hot-tub, whirlpool, or swimming-pool exposure. The distribution, appearance, and lack of water exposure make *Pseudomonas* folliculitis unlikely.

Staphylococcal folliculitis may occur as a beard folliculitis, but the lesions are typically raised, pruritic, erythematous, and less than 5 mm in diameter, with apical pustules. Lesions of staphylococcal folliculitis are not vesicular.

Eosinophilic folliculitis is an intensely pruritic, non- vesicular eruption of 3- to 5-mm papules or pustules found mainly on the scalp, face, neck, and upper chest. In the United States, eosinophilic folliculitis is found primarily in adults with advanced HIV infection but also has been reported in infants and patients with hematologic diseases and as a side effect of medication. Excoriation of the lesions is quite common and alters the primary morphology. This patient has no history of HIV infection, hematologic disease, or medication use, which makes eosinophilic folliculitis unlikely.

**KEY POINT**
- The vesicle is the primary lesion of herpes simplex infection and may cause folliculitis in the beard area.
Bibliography

Item 8  Answer: C
Educational Objective: Manage cutaneous lupus erythematosus.

Smoking is known to adversely affect the efficacy of therapy with antimalarial agents in patients with cutaneous lesions of lupus erythematosus. The mechanism of this phenomenon is not understood, but the products of cigarette smoking may interfere with antimalarial agents, lupus may be worsened by these chemicals, or both. Smoking cessation is therefore the most appropriate choice prior to initiating more aggressive and potentially toxic therapy.

Both methotrexate and thalidomide are treatments for cutaneous lupus erythematosus that is resistant to antimalarial agents, but smoking-cessation efforts should occur prior to the initiation of either of these more toxic alternatives.

Both amiodarone and hydrochlorothiazide are known to cause or exacerbate subacute cutaneous lupus erythematosus, but they have not been implicated as a cause of chronic cutaneous lupus erythematosus.

KEY POINT
- Smoking interferes with therapy of cutaneous lupus erythematosus.

Bibliography

Item 9  Answer: A
Educational Objective: Manage lentigo maligna.

This patient’s lesion is large and irregular with uneven pigmentation. The most likely diagnosis is malignant melanoma in situ (lentigo maligna type). Lesions may have a black as well as light-tan pigmentation. Lentigo maligna grows slowly and is commonly present for many years before the diagnosis is established. In early stages, the lesion is confluent to the epidermis; however, once it invades the dermis, it is just as lethal as any other melanoma. Lentigo maligna is the most important exception to the general rule that pigmented lesions should never receive shave biopsy: The atypical melanocytes grow at the dermal-epidermal junction, and a broad, shallow shave biopsy allows accurate diagnosis without disfiguring the patient. This sampling technique is different from the complete excision recommended in other forms of melanoma.

Punch biopsies should never include normal skin as this only increases the risk of sampling error. In the setting of lentigo maligna, even a punch biopsy from the center of the lesion has a false-negative rate of up to 80% and is not preferred to shave biopsy.

Even though this skin lesion is asymptomatic and has been present for many years, reassurance is inappropriate in this setting because lentigo maligna melanoma is slow growing, may be present for many years before the diagnosis is made, and can be lethal if not diagnosed and treated properly.

KEY POINT
- A broad, shallow shave biopsy is the preferred method for diagnosis of lentigo maligna.

Bibliography

Item 10  Answer: C
Educational Objective: Diagnose urticarial vasculitis.

Urticarial plaques that are fixed in location for more than 24 hours should be biopsied to rule out urticarial vasculitis. Patients often report that the lesions burn rather than itch. Lesions commonly heal with bruising. Most cases of urticarial vasculitis are idiopathic, but it can also be associated with autoimmune diseases (most commonly systemic lupus erythematosus), drug reactions, infections, or cancer. If urticarial vasculitis is diagnosed on skin biopsy, the next diagnostic step is measurement of serum complement levels. The presence of hypocomplementemia predicts the presence of systemic vasculitis.

Laboratory testing and imaging studies are appropriate for patients with urticaria when prompted by additional signs or symptoms. A chest radiograph is not appropriate in the absence of respiratory symptoms. Latex radioallergosorbent testing (RAST) would be appropriate for a patient with a history of hives that develop at the site of contact with latex. A stool sample for ova and parasites would be appropriate for a patient with peripheral eosinophilia, diarrhea, or other gastrointestinal symptoms.

KEY POINT
- Urticarial plaques that remain fixed in position for longer than 24 hours should be biopsied to rule out urticarial vasculitis.

Bibliography

Item 11  Answer: D
Educational Objective: Manage toxic epidermal necrolysis.

This patient has toxic epidermal necrolysis (TEN), which is characterized by mucous membrane involvement (eyes,
oral mucosa, and genitalia in this patient), epidermal detachment affecting greater than 30% of the body surface area, and a positive Nikolsky sign (lateral pressure on nonblistered skin leads to demodulation). TEN is almost always caused by a medication. The medications most closely associated with TEN are allopurinol, aromatic anticonvulsants (carbamazepine, phenytoin, phenobarbital), lamotrigine, sulfasalazine, sulfonamide antibiotics, NSAIDs, and nevirapine. In patients with TEN, survival and severity of disease are improved when the suspected causative medication and all unnecessary medications are immediately stopped.

The two most important determinants of outcome in patients with TEN are stopping all unnecessary medications and management in a burn unit.

Empiric treatment with acyclovir is not indicated because there is no evidence of an active herpes simplex infection; TEN is rarely caused by infection (including viral infections), and starting acyclovir would add an unnecessary medication.

Empiric antibiotics are not indicated in the initial management of TEN because they expose the patient to unnecessary drugs when the goal is to minimize medication exposures. However, because TEN typically results in loss of epidermis over a significant portion of the body surface area, patients are at risk for life-threatening infections during the course of the disease.

Systemic corticosteroids may be useful in the management of severe allergic drug reactions, particularly early in the course; however, clinical trials do not support the use of corticosteroids once the diagnosis of TEN is established, as their use predisposes the patient to serious side effects, including superinfection.

**KEY POINT**

- The most important initial step in managing patients with toxic epidermal necrolysis is stopping the suspected causative medication, as well as stopping all unnecessary medications.

**Bibliography**


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**Item 12**  **Answer:** D

**Educational Objective:** Treat pemphigus vulgaris.

Painful oral erosions are a common presenting manifestation of pemphigus vulgaris, which then may spread to the skin, usually the scalp, chest, face, axillae, and groin. Referral to a dermatologist who is skilled in treating pemphigus is recommended. Oral corticosteroids (1 to 2 mg/kg/d of prednisone) have the most rapid onset and are therefore indicated for initial therapy of pemphigus vulgaris. Once patients are on prednisone, immunosuppressive agents such as azathioprine should be added to limit the corticosteroid toxicity.

Cyclophosphamide, mycophenolate mofetil, rituximab, and intravenous immune globulin are viable corticosteroid-sparing agents in the treatment of pemphigus, and one of these might be initiated early in the course of the disease; however, they are not first-line treatments.

Cyclophosphamide is an excellent immunosuppressive/cytotoxic agent; however, it is a third choice for pemphigus therapy due to long-term consequences associated with its use.

A few studies involving a small number of patients suggest that intravenous immune globulin may improve disease in patients with pemphigus. Treatment is generally recommended for disease that is recalcitrant to corticosteroid therapy.

Mycophenolate mofetil has been used in patients with pemphigus vulgaris, but it is usually reserved for the patient who is either intolerant to azathioprine or in whom azathioprine is contraindicated.

Rituximab is approved for use in patients with B-cell lymphomas and rheumatoid arthritis, and its usefulness in treating pemphigus has been documented in open label trials.

**KEY POINT**

- The initial therapy for pemphigus vulgaris is prednisone with the later addition of an immunosuppressive agent to limit corticosteroid toxicity.

**Bibliography**


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**Item 13**  **Answer:** C

**Educational Objective:** Diagnose rosacea.

This patient has rosacea, which is an inflammatory dermatitis characterized by erythema, telangiectasias, papules, pustules, and sebaceous hyperplasia that develops on the central face, including the nasolabial folds, Rhinophyma, or the presence of a bulbous, red nose, is a variant of this condition.

Dermatofibrosis may also be associated with various skin manifestations. Periungual erythema and malar erythema, consisting of a light purple (heliotrope) edematous discoloration of the upper eyelids and periorbital tissues, are the most common presentations. Dermatofibrosis also may cause an erythematous, papular eruption that develops in a V-shaped pattern along the neck and upper torso; in a shawl-shaped pattern along the upper arms; and on the elbows, knees, ankles, and other sun-exposed areas. Involvement of the hands may include scaly, slightly raised, purplish papules and plaques that develop in periangular areas of the metacarpal and interphalangeal joints and other bony prominences (Gottron sign) and dry, rough, cracked, horizontal lines on the palmar and lateral aspects of the fingers (mechanic's hands).
Psoriasis usually involves the scalp, elbows, or other areas but does not typically manifest as an isolated facial rash. Characteristic findings of psoriasis include an erythematous plaque with an adherent, variably thick, silvery scale. Scleroderma dermatitis causes white, scaling macules and papules that are sharply demarcated on yellowish-red skin and may be greasy or dry. Sticky crusts and fissures often develop behind the ears, and significant dandruff or scaling of the scalp frequently occurs. Scleroderma dermatitis may develop in a "butterfly"-shaped pattern but also may involve the nasolabial folds, eyebrows, and forehead. This condition usually improves during the summer and worsens in the fall and winter.

Distinguishing rosacea from systemic lupus erythematosus can be difficult and is frequently a reason that patients are referred to a dermatologist. Systemic lupus erythematosus is unlikely in this patient because the malar rash associated with this condition is usually photosensitive and often spares the nasolabial folds and the areas below the nares and lower lip (areas relatively protected from the sun).

**KEY POINT**
- Rosacea is an inflammatory dermatitis characterized by erythema, telangiectasias, papules, pustules, and sebaceous hyperplasia that affects the central face, including the nasolabial folds.

### Bibliography


### Item 14  
**Answer: C**

**Educational Objective:** Diagnose vasculitic ulcers.

The patient has atypical-appearing ulcers. The presence of irregular, punched-out–appearing ulcers with fibrinous bases in a patient with known rheumatoid arthritis suggests the possibility of vasculitis. The appropriate diagnostic test is a punch biopsy. Biopsy should also be considered for nonhealing ulcers.

Ulcers on the medial leg are most commonly due to venous insufficiency and are often associated with the hyperpigmentation of venous stasis. This patient’s ulcers are located on the lateral aspect of the leg and are not associated with hyperpigmentation. Neuropathic ulcers are often associated with peripheral neuropathy; appear over pressure points such as the metacarpal joints, and may have a hyperkeratotic border. Arterial ulcers are typically found at the end of digits or over pressure points and are found in association with other chronic findings of ischemic disease, such as a history of claudication; atrophic, shiny, and hairless skin; and thickened toenails. Pulses are often absent, the skin is cool, and capillary refill is slow. Arterial ulcers are not commonly located on the leg unless this happens to be a point of pressure (as might occur with a cast, brace, or prosthesis).

Measurement of the ankle-brachial index is useful when arterial ulcers are suspected, but this patient’s physical findings do not support arterial disease.

Testing with a monofilament is useful in assessing risk for neuropathic ulcers but is not useful in diagnosing the etiology of ulcers.

Venous duplex Doppler ultrasonography is useful in assessing the presence of venous incompetence and planning surgery for deep venous disease; however, it is unlikely to be helpful in a patient who lacks physical examination findings typical of venous insufficiency.

### Bibliography


### Item 15  
**Answer: C**

**Educational Objective:** Diagnose pustular psoriasis.

An erythematous eruption that involves greater than 90% of the body surface area is indicative of erythroderma. The most common causes of erythroderma are drug eruptions, psoriasis, atopic dermatitis, and cutaneous T-cell lymphoma; however, the erythroderma may also be idiopathic. Patients such as this, who have a history of psoriasis and are treated with systemic corticosteroids, are particularly prone to developing an acute pustular erythrodermic flare days to weeks after discontinuation of the corticosteroids. The appropriate management is to treat the underlying disease (psoriasis in this patient), provide general supportive care for the erythrodermic skin, and treat complications such as temperature dysregulation, fluid and electrolyte shifts, and superinfections.

Drug hypersensitivity syndromes are classically associated with anticonvulsants, allopurinol, dapsone, and NSAIDs. They begin 3 to 6 weeks after the initiation of therapy. Angiotensin-converting enzyme inhibitors (lisinopril) are not commonly associated with hypersensitivity drug eruptions. In addition, patients with hypersensitivity drug eruptions typically present with widespread erythema that evolves over weeks rather than days and skin that is itchy more than painful, as well as facial swelling, lymphadenopathy, eosinophilia, atypical lymphocytosis, and elevated aminotransferases.

Because of this patient’s history of prostate cancer, paraneoplastic erythroderma may be a consideration. However, paraneoplastic erythroderma evolves slowly over months to years rather than in a few days.

Sézary syndrome is a leukemic form of cutaneous T-cell lymphoma that evolves slowly and is intensely pruritic. A severe, fissured keratoderma and lymphadenopathy are
often present. This patient’s symptoms are not consistent with the diagnosis of Sézary syndrome.

Staphylococcal scalded skin syndrome (SSSS) may be difficult to differentiate from pustular psoriasis, as it also presents with widespread erythroderma and skin pain. It is most common in children less than 6 years of age, but adults with underlying immunosuppression or renal failure may be affected. This patient is an adult with normal renal function who is without significant immunosuppression. Clinical features that are characteristic of SSSS include peripheral edema and early involvement of the intertriginous areas. The diagnosis is made clinically, but it can be confirmed by isolation of *Staphylococcus aureus* with bacterial culture of any suspected source of infection, blood, and mucous membranes.

**KEY POINT**
- Patients with a history of psoriasis who are treated with systemic corticosteroids may develop an acute pustular erythrodermic flare days to weeks after the systemic corticosteroids are discontinued.

**Bibliography**

**Item 16 Answer: A**
**Educational Objective:** Manage a patient with neurogenic pruritus.

This patient has subacute, severe, and generalized pruritus. In addition, only secondary erosions are present, with no identifiable primary dermatologic lesions (for example, papule, vesicle, pustule). These characteristics suggest neurogenic pruritus. Neurogenic pruritus is itch caused by a systemic disease or circulating pruritogens and requires an investigation into the underlying etiology. Important underlying causes of neurogenic pruritus include cholestasis, end-stage renal disease, thyroid disease (hypo- or hyperthyroidism), iron deficiency anemia, malignancy (usually hematologic or lymphoma), medications (opiates), and HIV infection. Laboratory studies in patients who have pruritus without obvious cause might include a complete blood count, erythrocyte sedimentation rate, serum creatinine level, serum thyroid-stimulating hormone level, and liver chemistry tests. Patients should also be evaluated with age- and sex-appropriate cancer screening tests, and additional tests should be guided by the presence of symptoms.

Because this patient has no identifiable primary skin lesions, a skin biopsy is unlikely to reveal the etiology of her pruritus. Patch testing is useful in the diagnosis of contact dermatitis, but this diagnosis is unlikely in the absence of an eczematous dermatitis. The use of systemic corticosteroids is not indicated to treat pruritus prior to establishing a diagnosis, and it exposes the patient to unnecessary side effects.

**KEY POINT**
- Patients with atopic dermatitis are more susceptible to disseminated cutaneous herpesvirus infection (eczema herpeticum).

**Bibliography**

**Item 17 Answer: C**
**Educational Objective:** Diagnose locally disseminated herpes simplex virus infection as a complication of atopic dermatitis.

The skin around the left eye has multiple coalescing, crusted vesicles on an erythematous base and is red and edematous. The presence of discrete and coalescing vesicles should immediately suggest the diagnosis of herpesvirus infection. There are also widespread, symmetrical, eczematous patches on the trunk and extremities, supporting the diagnosis of atopic dermatitis. Herpesvirus infection can locally disseminate in abnormal skin, such as that of patients with atopic dermatitis, in which the normal barrier function is lost.

Patients with eczema herpeticum may feel ill, be febrile, and have regional lymphadenopathy. The diagnosis can be confirmed by direct fluorescent antibody testing or herpes viral culture. Emergent opthalmologic consultation should be obtained in this patient to evaluate for herpes keratitis. Primary herpes keratoconjunctivitis is treated with topical trifluridine, vidarabine ointment, or acyclovir gel. Systemic antiviral therapy is typically prescribed for primary herpesvirus infection of the skin, but evidence for its efficacy is unclear. Because eczema herpeticum is considered to be a locally disseminated disease, treatment with systemic antiviral agents is appropriate. A more prolonged course may be needed for treatment of immunosuppressed patients. In some cases, hospitalization is needed for management of acutely ill patients.

Allergic contact dermatitis can cause edema and small papulovesicles in affected skin, but it does not cause the discrete vesicles that are characteristic of herpes simplex virus infection. Vesicles are not seen in atopic dermatitis. The presence of serum and crusting may suggest staphylococcal infection, and, indeed, a secondary infection is possible; however, the finding of vesicles and, later in the course of infection, punched-out-appearing ulcers or erosions is characteristic of herpes simplex virus infection.

**KEY POINT**
- Neurogenic pruritus should prompt a thorough evaluation for systemic causes.

**Bibliography**
Item 18  Answer: C

**Educational Objective:** Diagnose keratoacanthoma.

Keratoacanthoma is an epithelial neoplasm that is characterized by rapid growth over 2 to 6 weeks and by a crater-like configuration. Early lesions are frequently misdiagnosed as skin infections. The typical early lesion is a hard, erythematous nodule with a keratotic (horny) center. Keratoacanthomas typically occur on heavily sun-damaged skin, usually in older persons, with a peak age of 60 years. As the lesion enlarges, the center of the crater becomes more prominent. Unlike typical squamous cell carcinomas, keratoacanthomas are capable of spontaneous resolution by terminal differentiation, in which the tumor “keratinizes itself to death.” The clinical presentation and characteristic histologic features establish the diagnosis.

Keratoacanthomas occurring together with sebaceous adenomas suggest a diagnosis of Muir-Torre syndrome, an autosomal dominant syndrome associated with colon cancer. Muir-Torre syndrome is allelic to the hereditary non-polyposis colorectal cancer syndrome.

Because keratoacanthomas may cause significant local tissue destruction, simple observation is generally not recommended despite the tendency for spontaneous involution. Prompt surgical excision is recommended for solitary lesions on the trunk or extremities. Intraligamental 5-fluorouracil or methotrexate, topical imiquimod, and radiation therapy have also been used to treat large lesions or those in areas where surgical excision would be anatomically difficult.

Abscesses are warm, red, and tender and may be fluctuant if the lesion is palpated.

Keloids present as slow-growing, hard nodules, often with a dumbbell shape. They occur at sites of prior trauma.

Nodular basal cell carcinoma is often found on the face and is characterized by slow growth, the presence of a skin-colored papule, pearly, translucent papule with telangiectasia, rolled borders, and central depression, often with ulceration.

**KEY POINT**

- Keratoacanthomas are rapidly growing, non-tender, firm nodules with depressed keratotic centers that are often misdiagnosed as cutaneous infections.

**Bibliography**


Item 19  Answer: A

**Educational Objective:** Manage scarring ocular cicatricial pemphigoid.

This patient probably has cicatricial pemphigoid based upon the history of dryness of the eyes and evidence of conjunctival scarring (trichiasis and symblepharon). Ocular cicatricial pemphigoid can result from several immunologic phenomena, including linear IgA deposition, linear IgG deposition resembling bullous pemphigoid, or linear IgG deposition resembling epidermolysis bullosa acquisita. This disorder can be sight-threatening and, therefore, warrants accurate diagnosis with biopsy and appropriate histopathologic studies. Biopsy of the conjunctiva will reveal subepithelial separation below the basement membrane, and direct immunofluorescence will reveal linear deposition of IgG and C3 at the basement membrane zone. Once the diagnosis is confirmed, aggressive management with corticosteroids and cyclophosphamide is indicated. However, treatment with prednisone and cyclophosphamide should wait until confirmation of the diagnosis.

Herpes zoster ophthalmicus is a complication of varicella-zoster virus infection involving the ophthalmic division of the fifth cranial nerve. Most patients with herpes zoster ophthalmicus will experience headache and fever associated with pain or hypesthesia in the affected eye and forehead. With outbreak of the characteristic cutaneous vesicles, patients typically develop hyperemic conjunctivitis. Severely ill patients are often treated with intravenous acyclovir, but less ill patients may be successfully treated with oral valacyclovir or famciclovir. In the absence of the typical vesicular eruption of herpes zoster, there is no indication for intravenous acyclovir.

Bacterial conjunctivitis is caused by a range of gram-positive and gram-negative organisms and is characterized by presentation in one eye, but this condition often spreads to involve the other eye and is associated with purulent discharge. Empiric treatment with broad-spectrum topical antibiotics is indicated in patients with bacterial conjunctivitis. The patient’s 6-month history of ocular symptoms is not compatible with an acute bacterial conjunctivitis, and treatment with a topical antibiotic should not take precedence over a conjunctival biopsy.

**KEY POINT**

- Ocular cicatricial pemphigoid is sight-threatening and warrants accurate diagnosis with biopsy and appropriate histopathologic studies, as well as aggressive management with corticosteroids and cyclophosphamide.

**Bibliography**


Item 20  Answer: D

**Educational Objective:** Treat pyoderma gangrenosum.

This patient has pyoderma gangrenosum (PG). PG typically begins as tender papules, papulopustules, or vesicles that spontaneously ulcerate and progress to painful ulcers with a
purulent base and undermined, ragged, violaceous borders. Active lesions often show a gummatous-gray border surrounded by an erythematous halo. Pathergy, a phenomenon characterized by exacerbation of disease after trauma, is observed in 20% to 30% of patients and can initiate or aggravate PG. PG is associated with an underlying systemic disease in 50% to 78% of patients, and it can present before, concurrently with, or after the development of the associated underlying condition. Diseases most commonly associated with PG are inflammatory bowel disease (either ulcerative colitis or Crohn disease), rheumatoid arthritis, seronegative spondyloarthropathy, and hematologic disease or malignancy, specifically acute myeloid leukemia. PG reflects the activity of the bowel disease in about 50% of patients with inflammatory bowel disease. Treatment of PG is immune suppression, most often with systemic corticosteroids.

The diagnosis of PG requires ruling out other entities that can mimic PG, including infection. This patient’s normal leukocyte count and lack of fever argue against infection and, therefore, do not support empiric treatment with antibiotics. Wet-to-dry dressings and surgical debridement are contraindicated, because both are traumatic to the skin and may worsen the PG. In patients with extensive, poorly controlled colitis, proctocolectomy has been suggested as a treatment option for those with severe, refractory PG. However, there are case reports of PG first appearing after proctocolectomy. Nevertheless, proctocolectomy is an inappropriate treatment for a patient with well-controlled colitis prior to initiation of first-line treatment with systemic corticosteroids.

**KEY POINT**

- Systemic corticosteroids are the initial treatment of choice for pyoderma gangrenosum.

Bibliography


**Item 21 Answer: B**

**Educational Objective:** Treat gram-negative folliculitis as a complication of acne vulgaris.

Gram-negative folliculitis is a complication of long-term oral antibiotic therapy for acne vulgaris. The typical history is an acute pustular exacerbation of preexisting common acne or nodular acne. Physical examination reveals many inflamed pustules, most often on the central face. The diagnosis is confirmed by a culture positive for gram-negative bacteria, usually *Escherichia coli*, in the nares and/or from a pustule. The broad-spectrum antibiotic allows overgrowth of gram-negative organisms in the anterior nares. Nearly 85% of patients treated with an oral antibiotic for longer than 6 months have a dominant gram-negative organism present in the nares. This organism can then secondarily infect acne lesions, causing the flare of pustules. Isotretinoin is not an antibiotic, but it is the treatment of choice for gram-negative folliculitis. Isotretinoin modulates epidermal proliferation; induces orthokeratosis; and inhibits comedo formation and comedolysis via disruption of desmosomes, inhibition of inflammation, shrinkage of sebaceous glands, and inhibition of sebum secretion. Common side effects include dry skin, chapped lips, dry eyes, nosebleeds, and hair shedding. It also dries the anterior nares, ridding the mucosa of the colonized organisms, and it treats the underlying acne effectively. Isotretinoin is a pregnancy risk category X drug. All prescribers, patients, wholesalers, and dispensing pharmacies must be registered in the U.S. Food and Drug Administration-approved iPLEDGE program (1-866-495-0654).

The antibiotics ciprofloxacin, minocycline, and trimethoprim-sulfamethoxazole would likely result in a temporary improvement of the pustules, but they would not eliminate the gram-negative overgrowth from the nares. Recurrence would rapidly follow discontinuation of the antibiotic.

Spirotetractone may improve this patient’s acne but would not address the underlying gram-negative overgrowth.

**KEY POINT**

- Gram-negative folliculitis is a complication of long-term oral antibiotic therapy for acne vulgaris; isotretinoin is the treatment of choice.

Bibliography


**Item 22 Answer: D**

**Educational Objective:** Diagnose lentigo maligna.

This patient’s lesion is large and irregular with mottled pigmentation. The most likely diagnosis is malignant melanoma in situ (lentigo maligna type), which is confined to the epidermis. Lentigo maligna grows slowly and is commonly present for many years before the diagnosis is established. Once it invades the dermis (lentigo maligna melanoma), it is just as lethal as any other melanoma. The patient should be sent to an experienced dermatologist for a broad, paper-thin shave biopsy to sample the lesion. This biopsy technique is specific to lentigo maligna and is different from techniques used in other melanomas.

Pigmented actinic keratosis is usually 4 mm or less; has a keratotic, rough surface; and appears on sun-exposed skin. It ranges in clinical presentation from macular erythematous patches to large hyperkeratotic excrescences. The erythema surrounding the base of the lesion and its rough, prickly surface texture help differentiate this lesion from other skin conditions.

Pigmented basal cell carcinoma typically presents as a firm, pearly papule with telangiectasia and flecks of brown
pigment. With time, the center may umbilicate and ulcerate to produce the characteristic rolled borders.

Benign solar lentigo is tan to light brown and evenly pigmented, which helps distinguish it from lentigo maligna. However, like lentigo maligna, it may have irregular borders. Typical lesions range in size from a few millimeters to over 1 cm in diameter. Like lentigo maligna, these lesions are typically found in older individuals on sun-exposed skin.

Seborrheic keratosis is a painless, nonmalignant growth that appears as a waxy, brownish patch or plaque. Seborrheic keratoses typically exhibit horn cysts (epidermal cysts filled with keratin) on the surface that can best be visualized with a magnifying lens. Treatment is necessary only if lesions are symptomatic or interfere with function.

### Key Point

- Lentigo maligna melanoma begins as a tan-brown macule on sun-exposed skin of older individuals and may be present for many years before it invades the dermis.

Bibliography


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**Item 23 Answer: D**

**Educational Objective:** Treat a patient with perioral dermatitis caused by fluorinated corticosteroid use.

This young woman has perioral dermatitis, which resulted from the use of a fluorinated topical corticosteroid to control seborrheic dermatitis. Perioral dermatitis is a papular and pustular eruption that appears around the mouth and is usually caused by the use of topical or inhaled corticosteroids. This patient initially had seborrheic dermatitis, which is itchy, recurrent, red, and scaly and responds well to fluorinated topical corticosteroids. However, fluorinated topical corticosteroids should be avoided on the facial skin. If used chronically, they may produce atrophy, telangiectasia, or a rosacea- or perioral dermatitis-type papular and pustular rash. Facial seborrheic dermatitis should instead be treated with nonsteroidal preparations such as ketoconazole cream. If this patient’s seborrheic dermatitis flares upon discontinuation of the trimcinolone, ketoconazole cream is the treatment of choice.

Topical corticosteroid therapy should be avoided in this patient. Increasing the corticosteroid potency, as with clobetasol, will only worsen the perioral dermatitis.

Benzoyl peroxide is an excellent treatment for acne; however, it is known to be quite irritating and drying. It is likely to exacerbate this patient’s seborrheic dermatitis. Additionally, this patient’s perioral dermatitis will resolve spontaneously after discontinuation of the topical corticosteroid.

Neomycin ointment is not known to improve acne, rosacea, or perioral dermatitis. It is, however, one of the top culprits in causing allergic contact dermatitis reactions. There is no role for this medication in the treatment of perioral dermatitis.

### Key Point

- Perioral dermatitis is a papular and pustular eruption that appears around the mouth and is usually caused by the use of topical or inhaled corticosteroids.

Bibliography


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**Item 24 Answer: E**

**Educational Objective:** Understand the association between recurrent herpes zoster and HIV infection.

This patient most likely has HIV infection. Herpes zoster infection is the reactivation of the varicella virus in a single cutaneous nerve. Recurrence of herpes zoster infection in the immunocompetent host is uncommon but does occur. When recurrent disease is present, the underlying cause is overwhelming HIV infection. In this patient, there is a band of crusts and blisters on an erythematous base along a dermal distribution on the left thorax. There is evidence of scarring in a dermatome several centimeters above the currently involved site, representing a previous herpes zoster infection. Almost half of all herpes zoster episodes diagnosed in patients with HIV are recurrences. The advent of highly active antiretroviral therapy has not lessened the incidence of recurrent herpes zoster infection in patients with HIV infection. Patients on chemotherapy and patients who have undergone organ transplant may also develop recurrent herpes zoster. All patients with HIV infection and herpes zoster infection are treated with antiviral therapy regardless of the age of the zoster lesions. Most patients with HIV infection can be treated with an oral antiviral drug with good bioavailability, such as valacyclovir, but patients with severe disease, evidence of dissemination, or ophthalmologic involvement may have better outcomes if treated with intravenous acyclovir.

A patient with unexplained weight loss and fatigue may have an underlying metabolic disease like diabetes mellitus, but diabetes is not associated with recurrent herpes zoster. Because of this patient’s injection drug use, he is at risk for hepatitis B and hepatitis C, and screening for these infections is recommended. However, neither of these infections is associated with recurrent herpes zoster. This patient’s alcoholism is a risk factor for cirrhosis but not for recurrent herpes zoster infection.

### Key Point

- Recurrent herpes zoster infection should trigger testing for possible associated HIV infection.
Item 25  Answer: D

Educational Objective: Understand the association between vitiligo and autoimmune diseases.

This patient has vitiligo. Vitiligo is primarily a clinical diagnosis based on characteristic skin findings of depigmented, “chalk”-white, clearly demarcated, round or oval macules that may present in a variety of distributions. Thyroid screening with measurement of the thyroid-stimulating hormone level is the single most reasonable screening test for this patient. Vitiligo has been significantly associated with various autoimmune diseases, which should lower the clinician’s threshold for evaluating such diseases. A survey study of 2624 patients with vitiligo revealed a statistically significant association between vitiligo and several autoimmune diseases when compared with population frequencies of these diseases. The study found that 19.4% of all white patients aged 220 years had autoimmune thyroid disease; 1.78% had pernicious anemia; 0.67% had inflammatory bowel disease; 0.38% had Addison disease; and 0.19% had systemic lupus erythematosus. No statistically significant association with diabetes mellitus was found in this study. Variants of the gene NALP1, a regulator of the innate immune system, have been shown to be associated with the risk of developing vitiligo, as well as other epidemiologically associated autoimmune diseases, including autoimmune thyroid disease, diabetes, rheumatoid arthritis, psoriasis, pernicious anemia, systemic lupus erythematosus, and Addison disease.

Although there is a statistically significant association between vitiligo and pernicious anemia, the absolute prevalence is quite small. Because this patient has a normal complete blood count, pernicious anemia is unlikely, and measurement of the serum vitamin B12 level is not indicated. Similarly, the likelihood of Addison disease in a patient with normal vital signs and normal serum electrolytes is quite small, and a morning cortisol measurement is not needed. This patient has a normal BMI and no symptoms, and the vitiligo probably does not increase her risk of diabetes enough to warrant screening with a fasting glucose measurement.

**KEY POINT**

- The most common systemic condition associated with vitiligo is autoimmune thyroid disease.

Bibliography


**Item 26  Answer: E**

Educational Objective: Manage a patient with erythroderma.

This patient’s signs and symptoms are consistent with a slowly evolving erythroderma. An underlying cause for erythroderma should always be sought in order to guide therapy and determine prognosis. The diagnosis of idiopathic erythroderma is one of exclusion and should only be made after all other potential causes have been ruled out. Skin biopsy with routine hematoxylin and eosin staining should be performed in every patient with erythroderma; however, histopathologic findings diagnostic of the underlying cause are present in only 50% of patients. If the initial biopsy is nondiagnostic, additional biopsies may be useful and are recommended. This patient’s disease, previously diagnosed as eczema, began in adulthood and has not responded to therapy (topical corticosteroids) that is typically effective in the treatment of atopic dermatitis. In addition, he had no personal or family history of atopy (asthma, atopic dermatitis, allergic rhinitis). Atopic dermatitis rarely presents in adulthood in patients without a personal or family history of atopy and is most commonly confused in this setting with cutaneous T-cell lymphoma. Therefore, the most important next step in the management of this patient is a skin biopsy to rule out cutaneous T-cell lymphoma/Sézary syndrome.

Cyclosporine and phototherapy are potential treatments for erythroderma, either idiopathic or related to a particular cause. However, before treating erythroderma with a systemic agent or phototherapy, the cause of the erythroderma should be sought.

Antinuclear antibody and rapid plasma reagin are tests for autoimmune connective tissue disease and syphilis, respectively. Neither autoimmune connective tissue disease nor syphilis commonly causes erythroderma, making these options incorrect.

**KEY POINT**

- A skin biopsy is always required in the evaluation of a patient with erythroderma.

Bibliography


**Item 27  Answer: A**

Educational Objective: Diagnose allergic contact dermatitis.

This patient has chronic hand dermatitis, the differential diagnosis of which includes allergic contact dermatitis (ACD) and irritant contact dermatitis. Several features of this patient’s dermatitis suggest ACD, including the relatively recent onset of her condition (as opposed to lifelong), pruritus, and improvement when she is away from work. Epicutaneous patch testing is the appropriate test to
evaluate for ACD and may help distinguish this from other types of chronic eczematous dermatitis, including irritant dermatitis and atopic dermatitis. This patient has a history of childhood eczema and, with her history of allergic rhinitis and family history of asthma, may also have atopic dermatitis. Hairdressers are exposed to several allergens that not infrequently cause occupational ACD. Accelerants and other chemicals can also cause allergies. Gloves may not protect a sensitized individual from reactions to all allergens, because some chemicals are able to penetrate natural rubber latex and synthetic rubber gloves.

Lymphocyte stimulation or proliferation assays are used primarily in the laboratory setting; their clinical utility in the evaluation of allergy remains uncertain. Prick testing and radioallergosorbent testing (RAST) are useful in diagnosing immediate-type hypersensitivity reactions; however, these tests are less appropriate in the setting of ACD, which is a delayed-type hypersensitivity reaction.

**KEY POINT**

- Epicutaneous patch testing is the gold standard for diagnosis of allergic contact dermatitis in patients with persistent eczematous dermatitis.

**Bibliography**


**Item 28**  **Answer: A**

**Educational Objective:** Diagnose actinic keratoses.

This patient has actinic keratoses, common lesions that occur on sun-exposed skin of older white-skinned persons. Actinic keratoses are believed to be the earliest clinically recognized step in a biologic continuum that may result in invasive squamous cell carcinoma. Actinic keratoses are 1- to 3-mm, elevated, flesh-colored or red papules surrounded by a whitish scale. They are often easier to feel as "rough spots" on the skin than they are to see. Most patients will have, on average, 6 to 8 lesions. Most remain stable and some regress, but others enlarge to become invasive squamous cell carcinomas.

A basal cell carcinoma classically presents as a pink or pearly or translucent, dome-shaped papule with telangiectasias. The papule may have central umbilication.

A melanoma is classically a pigmented macule or plaque that is asymmetric and has irregular, scalloped, notched, or indistinct borders. It is black or dark brown or has variegated (multiple) coloration, including shades of black, red, and blue. Melanomas may also have depigmented or white areas, which represent regression of the lesion. Rarely, melanomas are not pigmented and can resemble basal cell carcinomas.

Seborrheic keratoses can be brown or black, but have discrete borders, are elevated above the surface of the skin, and have a "stuck-on" warty or waxy appearance.

**KEY POINT**

- Patients with asthma and nasal polyps may experience hives due to aspirin sensitivity.

**Bibliography**

Item 30  Answer:  B

Educational Objective: Manage a neuropathic ulcer.

This patient with long-standing diabetes mellitus and evidence of peripheral neuropathy has a neuropathic ulcer. In addition to ensuring adequate vascular supply, removing devitalized tissue, and treating infection, removing pressure from the ulcer aids in healing. This can typically be accomplished by prolonged bed rest or the application of a total-contact cast. Most patients prefer the contact cast with its attendant freedom of mobility.

Neuropathic ulcers are recognized by their location at pressure points such as over the metatarsal heads. The ulcers characteristically have a thick surrounding zone of hyperkeratosis, and hammer toes and a flattened foot arch are commonly found. The presence of foot ulcers, good color and warmth, and a near-normal ankle-brachial index strongly suggests that revascularization is not necessary for this patient. Antibiotics should not be used in the absence of infection. The patient’s ulcer appears clean, and the absence of erythema, warmth, tenderness, and swelling also suggests that infection is absent. The ability to probe to bone is a highly sensitive test for the presence of an underlying osteomyelitis; however, the inability to probe to bone does not rule out infection. This patient should probably have an MRI of the foot to assess for underlying bone infection.

Empiric antibiotics are not indicated. Furthermore, vancomycin would be a poor choice for empiric antibiotic treatment because of the necessity to cover streptococci, methicillin-resistant Staphylococcus aureus, aerobic gram-negative bacilli, and anaerobes.

An Unna boot is appropriate compression therapy for venous ulcers, which are typically found on the medial ankle around the malleolus. An Unna boot is inappropriate therapy for a neuropathic ulcer because the pressure that may be applied over the ulcer may inhibit healing.

Whirlpool hyperthermia is of no proven benefit in the treatment of neuropathic ulcers.

**KEY POINT**
- Offloading, usually with contact casting, can accelerate the healing of a neuropathic ulcer.

**Bibliography**

Item 31  Answer:  B

Educational Objective: Understand the association between erythema multiforme and recurrent herpes simplex virus infection.

Erythema multiforme is a mucocutaneous reaction characterized by targetoid lesions and, in most cases, both skin and mucosal involvement. The majority (up to 90%) of recurrent cases of erythema multiforme have been associated with infections, the most common of which is herpes simplex virus (both HSV-1 and HSV-2). No virus is routinely recovered with culture, and treatment with antiviral agents does not affect the outcome of an acute outbreak. Suppressive antiviral therapy, however, may minimize the number of erythema multiforme recurrences. It is important to recognize that recurrences of erythema multiforme can occur in the absence of apparent clinical reactivation of HSV; patients may not be aware that they are infected with HSV.

Erythema migrans (also called erythema chronicum migrans) is the hallmark cutaneous lesion of early Lyme disease. A centripetally spreading ring of erythema that resembles a bull’s-eye usually develops at the site of infection 3 to 30 days after a tick bite. Erythema migrans lesions are most typically found near the axilla, inguinal region, popliteal fossa, or at the belt line, and palmar involvement is rare, if it occurs at all. Lesions slowly expand over days or weeks, with central clearing producing a target or bull’s-eye appearance, and increase in size to 20 cm or more. Erythema migrans is distinguished from erythema multiforme by the lesion size, its location, and lack of associated mucosal involvement. This patient’s findings are not consistent with erythema migrans, and neither current nor past infection with *Borrelia burgdorferi* predisposes the patient to recurrent episodes of erythema multiforme.

Rocky Mountain spotted fever (RMSF) is a tick-borne disease caused by *Rickettsia rickettsii*. RMSF may present with subtle, fine, pink, blanching macules and papules on the wrists and ankles that then spread centripetally and to the palms and soles. As the rash spreads, the characteristic petechial and purpuric “spots” appear. Most patients have fever, severe headache, and myalgia. This patient’s findings are not consistent with RMSF, and neither current nor past infection with *R. rickettsii* predisposes the patient to recurrent episodes of erythema multiforme.

Streptococcal infections have been associated with erythema nodosum, flares of psoriasis, and several skin infections, including perianal cellulitis and blistering distal dactylitis; however, they are not commonly associated with erythema multiforme.

**KEY POINT**
- Up to 90% of cases of recurrent erythema multiforme are associated with infections, the most common of which is herpes simplex virus.

**Bibliography**

Item 32  Answer:  C

Educational Objective: Manage dermatitis herpetiformis using a gluten-free diet.

Most, if not all, patients with dermatitis herpetiformis have gluten sensitivity, even when they have no evidence of
Enteropathy. Treatment with a gluten-free diet is successful in greater than 70% of patients with dermatitis herpetiformis, but excellent adherence to the diet is required for a minimum of 3 to 12 months. In the interim, initial suppression of symptoms with dapsone is usually necessary for more rapid relief of symptoms. Continued compliance with the gluten-free diet will allow a decrease in the dapsone, and it can often be discontinued. A gluten-free diet treats the cause, rather than the symptoms, of the disease. Dapsone treatment requires careful monitoring. Hemolysis is the most common side effect of treatment and may be severe in patients with glucose-6-phosphate dehydrogenase (G6PD) deficiency. Pretesting for G6PD deficiency prior to initiating therapy with dapsone is generally recommended. Additional adverse reactions include toxic hepatitis, cholestatic jaundice, psychosis, and both motor and sensory neuropathy. Patients with dermatitis herpetiformis and their first-degree relatives are at increased risk for other autoimmune diseases, including thyroid disease, rheumatoid arthritis, and lupus erythematosus.

There is no role for cyclosporine, a lactose-free diet, or intravenous immune globulin in the treatment of dermatitis herpetiformis.

**KEY POINT**
- Treatment with a gluten-free diet is successful in greater than 70% of patients with dermatitis herpetiformis, even in the absence of symptomatic enteropathy.

**Bibliography**

**Item 33** Answer: E

**Educational Objective:** Diagnose solar (senile) purpura in a patient aged and photodamaged skin.

Solar or actinic purpura occurs frequently in individuals who have extensive photodamage of the skin. Minimal trauma, which may or may not have been recognized, causes large, asymptomatic ecchymoses. The surrounding skin is fragile and tears easily; in some cases, it heals with stellate scars. The forearms are most frequently involved, but other extremities and the face can develop similar lesions. Chronic sun damage weakens the blood vessel walls and surrounding stroma, allowing minimal trauma to cause extravasation of blood and a large bruise. Use of topical or systemic corticosteroids can exacerbate fragility.

Ecchymoses in this setting should not suggest an underlying coagulopathy; however, this can be ruled out by performing appropriate coagulation studies. This patient's normal coagulation studies rule out a liver-related coagulopathy.

**KEY POINT**
- Solar or actinic purpura is characterized by large, asymptomatic ecchymoses that occur with minimal trauma; it occurs frequently in individuals who have extensive photodamage of the skin.

**Bibliography**

**Item 34** Answer: D

**Educational Objective:** Diagnose Stevens-Johnson syndrome.

This patient has Stevens-Johnson syndrome (SJS). SJS is characterized by fever followed by the onset of erythematous macules and plaques that progress to epidermal necrosis and sloughing limited to less than 10% of the body surface area. Mucous membranes are affected in most patients, and ocular, oral, and genital surfaces may be involved. A sulfonamide is the most likely causative drug. Treatment in a burn unit is preferable for patients with extensive blistering and erosions.

Acute generalized exanthematous pustulosis (AGEP) is an exanthem that follows an infection or drug ingestion. It is characterized by the acute onset of widespread pustules that may resemble pustular psoriasis, along with fever,
leukocytosis, and possibly eosinophilia. AGEP is usually self-limiting and clears without residual skin changes approximately 2 weeks after drug cessation.

Drug reaction with eosinophilia and systemic symptoms (DRESS) is characterized by a generalized papular eruption, fever, arthralgia, and generalized lymphadenopathy. Associated laboratory findings may include elevatedaminotransferase levels, eosinophilia, and lymphocytosis. This patient’s cutaneous and laboratory findings are not compatible with DRESS.

The “red man syndrome” (RMS) is the most common adverse reaction to vancomycin. This reaction appears not to be antibody related and is characterized by flushing, erythema, and pruritus involving primarily the upper body, neck, and face. In a few individuals, it may be associated with back and chest pain, dyspnea, and hypotension. This patient has none of the clinical characteristics of RMS and was not exposed to vancomycin, making RMS an unlikely diagnosis.

**KEY POINT**

- Stevens-Johnson syndrome is an acute severe cutaneous reaction that is characterized by fever followed by the onset of erythematous macules and plaques that progress to epidermal necrosis and sloughing.

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**Bibliography**


**Item 35**  **Answer:** B

**Educational Objective:** Diagnose localized neuropathic pruritus (natalgia paresthetica).

This patient has natalgia paresthetica, which is characterized by recurrent pruritus, burning, or stinging on the mid-back. On physical examination, signs of chronic scratching or rubbing such as hyperpigmentation or lichenification (thickened skin with increased and exaggerated skin markings due to scratching) may be present, but there are no primary skin abnormalities that cause the itching. The skin may appear entirely normal. Natalgia paresthetica and brachioradial pruritus, a similar type of recurrent itching on the forearms, are examples of neuropathic pruritus—itch caused by an anatomic lesion in the peripheral or central nervous system. Some cases of natalgia paresthetica or brachioradial pruritus are associated with disease in the cervical and/or thoracic spine (frequently degenerative changes, such as osteophytes), and radiographic imaging may be appropriate; however, it is unusual to find a cause that would benefit from surgical management. Most patients have a normal neurologic examination despite their intense pruritus. These localized forms of neuropathic itch are difficult to treat; because they are noninflammatory and are not histamine mediated, corticosteroids are generally ineffective, as are antihistamines. Topical anesthetics (such as pramoxine) and topical capsaicin may provide some relief. Successful use of gabapentin, as well as tricyclic antidepressants including amitriptyline, has also been reported. For some patients, clarifying the diagnosis alone can be beneficial.

Herpes zoster presents as pruritic or painful vesicles in a dermatomal distribution. Itching or burning can precede the onset of vesicles; however, this patient’s symptoms have persisted for years.

Nummular dermatitis is a pruritic eczematous condition that presents as annular, coin-shaped, erythematous plaques with pinpoint vesicles, overlying oozing scale, and honey-colored, scaly crusting. This patient does not have a presentation consistent with nummular dermatitis.

Xerosis is the medical term for dry skin. While dry skin can be itchy, it presents with diffuse flaking of the skin without localized areas of hyperpigmentation. It is most common on the extremities and flanks.

**KEY POINT**

- Neuropathic pruritus should be considered in patients with localized itching without associated skin lesions; it can be associated with disease in the cervical and/or thoracic spine.

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**Bibliography**


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**Item 36**  **Answer:** A

**Educational Objective:** Diagnose eruptive xanthomas.

This patient has eruptive xanthomas, a specific subtype of xanthoma. Xanthomas are yellow, orange, or reddish papules, plaques, or nodules that are associated with underlying primary or secondary hyperlipidemias. Eruptive xanthomas present suddenly as crops of 1- to 4-mm, erythematous, yellow papules on extensor surfaces. They are specifically associated with marked elevations in serum triglyceride levels, often greater than 3000 mg/dL (33.9 mmol/L). Eruptive xanthomas and the associated hypertriglyceridemia may be caused by underlying diseases such as lipoprotein lipase deficiency, dysfunctional apoprotein CII, impaired insulin activity, familial hypercholesterolemia, and medications (isotretinoin, ritonavir). Treatment of hypertriglyceridemia leads to clinical improvement.

Plane xanthomas are yellow-red, slightly elevated plaques located in the skin folds of the neck and upper trunk. They may be associated with homozgyous familial hypercholesterolemia, dysbetalipoproteinemia, or no lipid disorder. Nonlipid disease associations include multiple myeloma, paraproteinemia, leukemia, and lymphoma. This patient’s findings are not consistent with plane xanthomas.

Tendon xanthomas are flesh-colored, smooth, firm, and lobulated subcutaneous nodules that move with the
underlying extensor tendon. Tendon xanthoma is associated with familial hypercholesterolemia caused by a defect in the LDL cholesterol receptors on the cell membrane. This patient’s sudden eruption of multiple small, colored papules is not consistent with tendon xanthoma.

Xanthelasmas are soft, yellow-orange, polygonal papules and plaques that are localized to the eyelids. They are the most common of all xanthomas and may be an isolated finding unrelated to hyperlipidemia or may be associated with familial hypercholesterolemia or familial dyslipoproteinemia.

**KEY POINT**
- Eruptive xanthomas present suddenly as crops of 1- to 4-mm, erythematous, yellow papules on extensor surfaces.

**Bibliography**

**Item 37 Answer: C**

**Educational Objective:** Understand the association between proximal white subungual onychomycosis and HIV infection.

This patient has proximal white subungual onychomycosis (PWSO), a rare form of onychomycosis. Onychomycosis usually presents as distal subungual debris; the infection rarely begins proximally in patients who are not immunocompromised. Studies show PWSO to be a common presentation of onychomycosis in patients with HIV infection. It can also be found in patients with other causes of immunodeficiency. If a patient presents with PWSO, HIV infection should be suspected as a predisposing factor.

In some studies, diabetes mellitus has been shown to be independently associated with an increase in onychomycosis, but the onychomycosis is not specifically of the PWSO type. Cushing disease, leukemia, and metastatic cancer, while often associated with opportunistic infections, have not been reported to be associated with an increased incidence of onychomycosis of any type.

**KEY POINT**
- Proximal white subungual onychomycosis is a common presentation of onychomycosis in patients with HIV infection.

**Bibliography**

**Item 38 Answer: A**

**Educational Objective:** Treat mixed cryoglobulinemia associated with hepatitis C infection.

This patient’s clinical signs and symptoms are most consistent with a diagnosis of mixed cryoglobulinemia associated with hepatitis C virus (HCV) infection. Mixed cryoglobulinemia, which is associated with HCV infection in greater than 90% of cases, is one cause of palpable purpura, the histological correlate of which is leukocytoclastic vasculitis. Although a skin biopsy showing leukocytoclastic vasculitis does not distinguish between the different etiologies for cutaneous small vessel vasculitis, the HCV infection, low complement levels, and presence of circulating cryoglobulins point to mixed cryoglobulinemia as the cause of his signs and symptoms.

Indications for treatment of mixed cryoglobulinemia include evidence of progressive systemic disease affecting the small blood vessels (such as cutaneous vasculitis), kidneys, liver, or peripheral nerves. The first-line therapy for mixed cryoglobulinemia associated with HCV is treatment of the underlying infection with pegylated interferon alfa and ribavirin.

There is no evidence that treatment with systemic corticosteroids is beneficial in patients with mixed cryoglobulinemia, and there is a theoretical concern that immunosuppression may lead to enhanced replication of HCV. Plasma exchange can lower circulating levels of cryoglobulins, and this therapy is typically reserved for patients with very aggressive disease, including advanced renal failure, distal necroses requiring amputation, or advanced neuropathy. These clinical findings are absent, and plasma exchange is therefore not indicated. Topical corticosteroids will have no effect on the systemic manifestations of circulating cryoglobulins and minimal, if any, effect on the cutaneous lesions.

**KEY POINT**
- The therapy of choice for mixed cryoglobulinemia associated with hepatitis C virus is treatment of the underlying infection with pegylated interferon alfa and ribavirin.

**Bibliography**

**Item 39 Answer: C**

**Educational Objective:** Diagnose seborrheic keratoses.

This patient has seborrheic keratosis, a benign skin condition. These lesions are common in adults and increase in number with age. They are characterized by sharply demarcated, tan to dark brown, warty papules, plaques,
and nodules that have a waxy texture and appear to be “stuck on” the skin. While they can arise on any area of the skin, they are frequently located in the scalp and on the back and chest.

Skin cancers tend to occur on the sun-exposed parts of the body. Basal cell carcinoma is a pearly or translucent papule or nodule with associated telangiectasias. Melanomas, like seborrheic keratoses, are pigmented, but do not classically have a waxy, warty surface. Melanomas often have irregular borders, whereas seborrheic keratoses are usually well demarcated. Distinguishing between the two can be difficult, however, and a biopsy may be necessary if the diagnosis is in question. Squamous cell carcinoma presents as a scaly, hyperkeratotic, red or pink papule, patch, or plaque. It is not brown, tan, or black and does not have a warty appearance like seborrheic keratoses.

**KEY POINT**

- Seborrheic keratoses are common, benign neoplasms that present as brown to black, well-demarcated, “stuck-on”-appearing papules with waxy surfaces.

**Bibliography**


**Item 40**

**Answer: C**

**Educational Objective:** Diagnose Paget disease of the breast.

This patient has Paget disease of the breast, defined as a persistent, scaling, eczematous, or ulcerated lesion involving the nipple/areolar complex. This disease is actually an extension of an underlying ductal adenocarcinoma of the breast that may be present even in the absence of abnormal physical examination findings or mammogram. It is often misdiagnosed on the first presentation as either eczema or psoriasis, but when there is a lack of response to appropriate therapy a biopsy should be performed.

The most characteristic lesions of chronic cutaneous lupus erythematosus are discoid lesions (erythematous, infiltrated plaques that are covered with scale and are associated with follicular plugging). These lesions are most often found on the face, neck, and scalp. As they expand, they develop depressed central scars. This patient’s lesion is not clinically compatible with chronic cutaneous lupus.

Lichen simplex chronicus is a localized disorder characterized by intense pruritus, which leads to a localized area of lichenified skin (thickened skin with increased and exaggerated skin markings due to scratching). This patient has no evidence of lichenification.

The most common form of psoriasis is plaque psoriasis. The skin lesions of this disorder are sharply demarcated, erythematous plaques covered by silvery-white scales that affect the scalp and extensor surfaces (elbows and knees) as well as the nails. A single patch of psoriasis located on the nipple would be a very rare presentation.

**KEY POINT**

- Paget disease of the breast presents as a persistent, scaling, eczematous, or ulcerated lesion involving the nipple/areolar complex and may be mistaken for more benign conditions such as eczema.

**Bibliography**