Dermatology

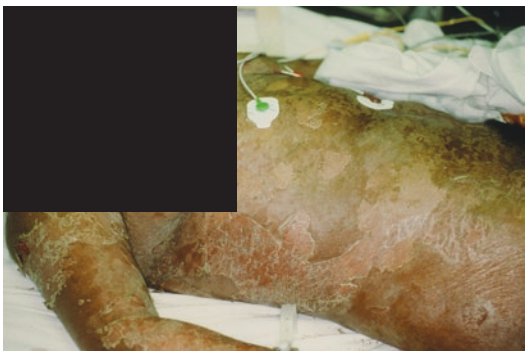
7

Donna M. Simmons, Michael D. Lanigan, and Ashley Keyes Jacobs

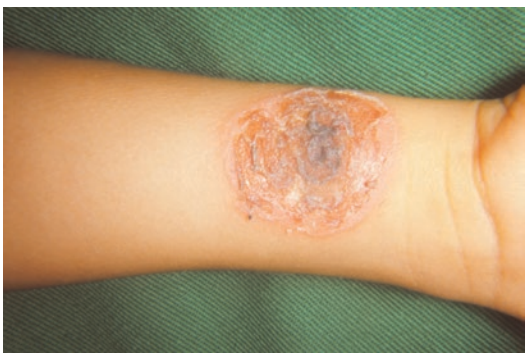
Question 1

A 4-year-old previously healthy boy developed a pruritic, vesicular rash with a fever. After initial improvement, he now has tender, indurated erythema of the right thigh. He is ill appearing and complains of pain; vital signs include a temperature of 103 °F, heart rate of 150 beats/minute, respiratory rate of 32 breaths/minute, and blood pressure of 85/60 mm Hg. What is the most likely cause of his symptoms?

A. Epidermolysis bullosa
B. Bullous impetigo
C. Scarlet fever
D. Toxic shock syndrome
E. Staphylococcal scalded skin syndrome



Toxic shock



Bullous impetigo

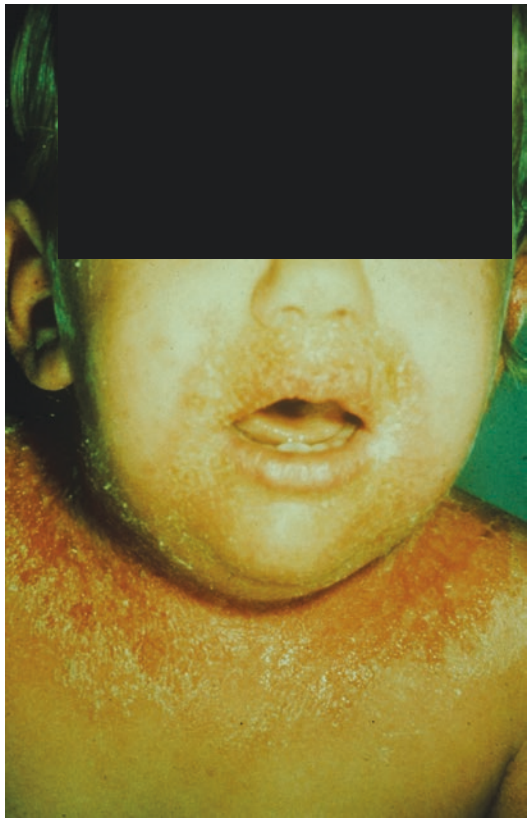
D. M. Simmons (✉)
St. Joseph's Health, Department of Emergency Medicine, St. Joseph's Hospital Medical Center, Paterson, NJ, USA
e-mail: simmonsd@sjhmc.org

M. D. Lanigan
Clinical Associate Professor, Department of Emergency Medicine, SUNY Downstate Medical Center, Brooklyn, NY, USA

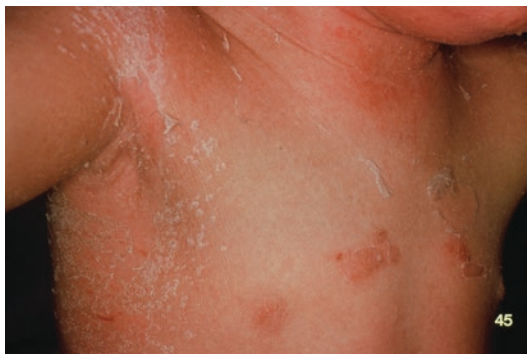
A. K. Jacobs
Interim Chair of Dermatology, Lincoln Medical Center, Bronx, NY, USA

Assistant Professor of Clinical Dermatology, Weill Cornell Medicine, New York, NY, USA
e-mail: Ashley.keyes@nychhc.org

M. Waseem et al. (eds.), *Prepare for the Pediatric Emergency Medicine Board Examination*,
https://doi.org/10.1007/978-3-030-28372-8_7



Staph scalded skin syndrome



Staph scalded skin syndrome

Correct Answer: D

This child has symptoms consistent with toxic shock syndrome (TSS). It may be caused by either *Streptococcus pyogenes*, group A streptococcus (GAS), or *Staphylococcus aureus*. Streptococcal TSS is more serious and arises from invasive, soft tissue infections such as necrotizing fasciitis and myositis. Clinical manifestations are fever (>102 °F [38.9 °C]), hypotension, diffuse erythroderma, and multisystem involvement. Common laboratory abnormalities include elevated creatine kinase, elevated blood urea nitrogen or creatinine, transaminitis, and thrombocytopenia. These patients require ICU-level supportive care. Antibiotic therapy should include clindamycin to suppress toxin production well as an antistaphylococcal penicillin or vancomycin pending wound cultures. Surgical debridement may be necessary if necrotizing fasciitis or myositis develop. Varicella is a rare predisposing factor for toxic shock in children.

Epidermolysis bullosa (EB) refers to a group of inherited disorders characterized by mechanically fragile skin with a propensity to develop blisters and/or erosions. The blisters develop after minimal trauma. Esophageal strictures may also occur. Infection with *Staphylococcus aureus* or GAS is a common complication.

The rash of scarlet fever has punctate erythematous papules which has a "sandpaper" consistency. It is associated with streptococcal pharyngitis. The rash may accentuate in skin folds with petechiae, a finding called "Pastia's lines." Exudative erythema of the tonsils and a red tongue with prominent papillae, "strawberry tongue," are additional cutaneous findings. A positive culture of group A β-hemolytic strep and elevated serum antistreptolysin-O titer support the diagnosis. With early diagnosis and initiation of penicillin, the prognosis is excellent.

Staphylococcal scalded skin syndrome (SSSS) is caused by infection with exfoliative toxin-producing *Staphylococcus aureus*, usually involving the conjunctivae, nares, or perineum. Patients develop fever, malaise, and irritability due to the systemically circulating toxin, along with characteristic erythema and fragile blisters in skin folds. The skin may be tender and have a sandpaper-like texture similar to that present in scarlet fever. Of note, the rash of scarlet fever is not tender. Superficial desquamation leaves a moist, red, glistening surface, and light stroking may result in separation of the upper portion of the epidermis (Nikolsky's sign). SSSS should be differentiated from TEN. Early distinction between SSSS and

TEN is important because the treatment for SSSS involves anti-staphylococcal antibiotics, whereas in TEN discontinuations of the offending agent and initiation of aggressive burn unit intervention are essential.

Bullous impetigo is caused by cutaneous infection with toxin-producing *Staph. aureus* and is considered a localized form of SSSS. Topical antibiotics may be effective for limited disease; however, systemic antibiotics are often needed.

Take-Home Message
Toxic shock syndrome is a rare and life-threatening complication of bacterial infection with *Streptococcus pyogenes* or *Staphylococcus aureus*.

Question 2

An 8-month-old boy is brought to the emergency department for evaluation of a rash and facial swelling. He has a runny nose and cough without fever, vomiting, or diarrhea. He has been active at home and is voiding normally. On examination, he is noted to have facial edema with rosette-shaped purpuric lesions on the face and both upper and lower extremities. The most appropriate treatment for this patient is:

A. IVIG
B. IV ceftriaxone
C. Supportive treatment
D. Skin biopsy to evaluate for IgA deposition
E. Systemic corticosteroids

Correct Answer: C

Fever, edema, and rosette-shaped or targetoid purpura primarily over the face, ears, and extremities in a nontoxic infant is consistent with acute hemorrhagic edema of infancy (AHEI). AHEI usually occurs following an infectious trigger or vaccination. It uncommonly presents with abdominal or joint pain. Leukocytoclastic vasculitis is seen on histopathology; however, the diagnosis is most often made clinically. No treatment is generally needed and resolution occurs over approximately 3 weeks.

AHEI is distinct from Henoch–Schonlein purpura (HSP), which manifests as symmetric palpable purpura on the lower extremities and abdominal pain with or without joint pain. The treatment is supportive; however, systemic corticosteroids may be required for severe symptoms such as abdominal pain, joint pain, or bullous skin lesions. The diagnosis can be made clinically. A skin biopsy is confirmatory showing leukocytoclastic vasculitis and IgA deposition on direct immunofluorescence. Urinalysis should be obtained to evaluate for renal involvement in the form of hematuria or proteinuria.

The child in this vignette has had symptoms for 3 days and is otherwise well appearing. Purpura fulminans is characterized by rapidly progressive hemorrhagic necrosis of the skin with disseminated intravascular coagulation. Treatment is directed at the underlying cause.

IVIG is a treatment option for patients with immune thrombocytopenic purpura.

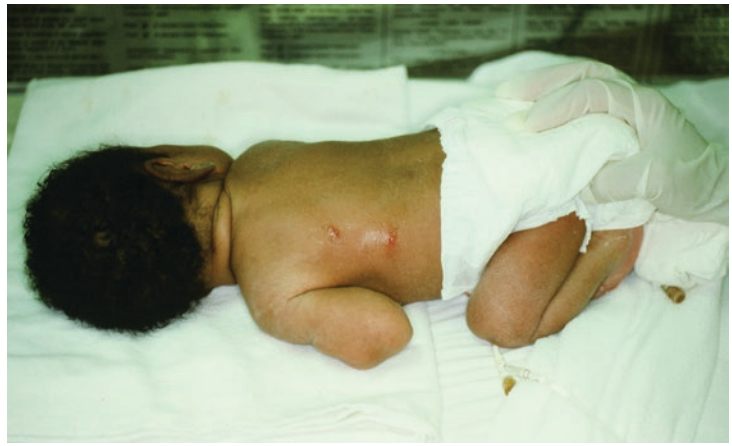
Take-Home Message
Acute hemorrhagic edema of infancy is a benign and self-limiting illness requiring supportive care. Common causes include infection, such as a viral upper respiratory infection, medications, and immunizations.

Question 3

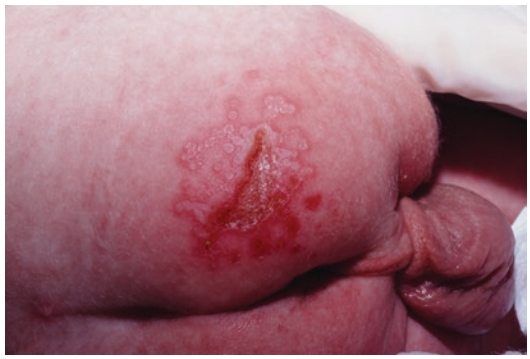
A 10-day-old male infant presents with vesicles on an erythematous base on the face. His mother states he has not been feeding well and has been sleeping more than usual. The most appropriate next step in your management of this child is:

A. Tell mother that a spinal tap is not needed because the child does not have fever
B. Obtain viral cultures of the rash and start oral acyclovir
C. Isolate the baby, evaluate for CNS and disseminated infection, and begin IV acyclovir
D. Reassure the family that all newborns sleep
E. Obtain measles IgG

Correct Answer: C



Herpes simplex



Neonatal herpes simplex

A vesicular rash in a neonate is concerning for herpes simplex virus infection (HSV). Neonatal herpes is classified by pattern: skin, eyes, and mouth, central nervous system, or disseminated. A vesicular rash or suspected HSV in a neonate requires contact isolation; evaluation for systemic involvement, including lumbar puncture; and empiric IV antiviral treatment while awaiting confirmatory diagnostic tests. In this patient, a scraping from the base of the lesion as well as cerebrospinal fluid can be sent for both viral culture and polymerase chain reaction (PCR). The neonate should be treated with high-dose acyclovir while awaiting test results. Expert groups recommend caesarean section if primary genital herpes is present in the third trimester or within 6 weeks of delivery.

Although skin lesions are the most common presentation, neonates may not have cutaneous findings in the initial course of disease; when a mother has a history of genital herpes, fever, irritability, lethargy, seizures, hepatitis, or pneumonitis in a newborn should raise consideration of neonatal herpes.

Measles does not present with vesicles; therefore, measles IgG is not indicated in this child.

Take-Home Message

Herpes or suspected herpes in the neonate is a life-threatening illness requiring empiric high-dose antiviral treatment and evaluation including lumbar puncture. Neonates born to mothers with active primary genital herpes at the time of delivery are at highest risk for infection.

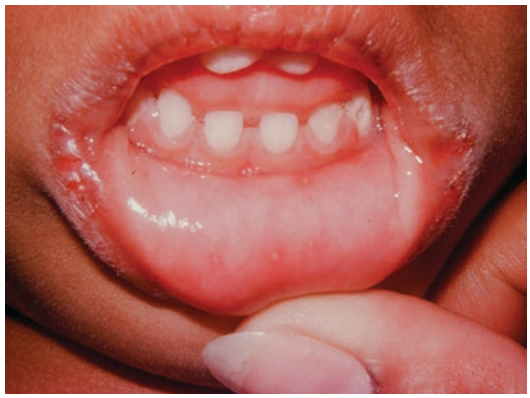
Question 4

A 5-year-old girl with a history of eczema develops a blister in her mouth. Her mother reports decreased oral intake and drooling. On examination, there are erythematous blanching patches with oval vesicles on her hands, feet, mouth, and buttocks. Which of the following statements is true regarding the most likely cause of this child's symptoms?

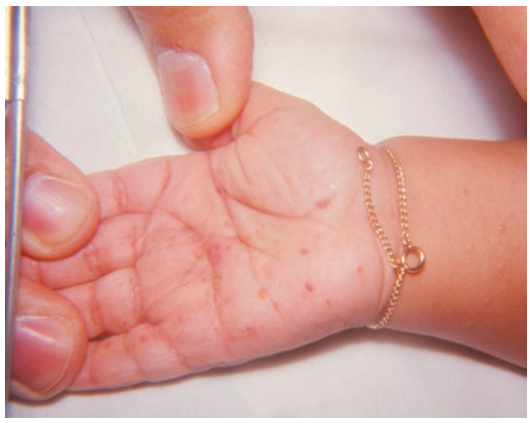
A. Topical acyclovir will decrease viral shedding
B. Topical corticosteroids will prevent recurrence
C. She may develop desquamation of the fingernails after the infection resolves

D. Involvement of the diaper area is atypical
E. The condition is not contagious

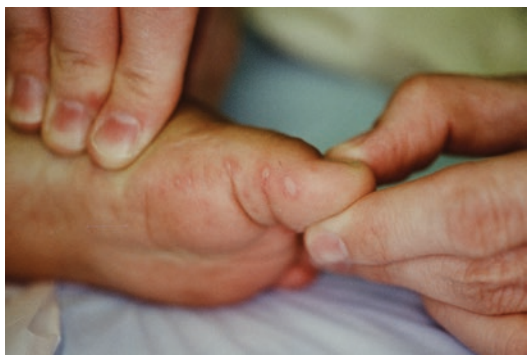
Correct Answer: C



Hand foot mouth disease



Hand foot mouth disease



Hand foot mouth disease

This child has a vesicular eruption in the typical distribution of hand, foot, and mouth disease (HFMD). The most common cause is coxsackie A16. It is highly contagious. There may be a prodrome of low-grade fever, malaise, diarrhea, and irritability which is followed by the appearance of papulovesicles on the palate, tongue, or buccal mucosa.

Vesicles are also found on the hands, feet, and diaper area and may generalize to the upper and lower extremities. Nail changes such as onychomadesis, which is separation of the proximal portion of the nail plate from the nail bed, may occur 2–4 weeks later.

In chicken pox, oral lesions are less common; the lesions are more extensive and centrally distributed; generally palms and soles are not involved. Also, herpangina may resemble HFMD, but it usually involves the posterior oropharynx; typical sites are fauces and soft palate. The vesicles of herpes develop in clusters. Multinucleated giant cells are present in the smear from a vesicle of varicella and herpes but not present in lesions of HFMD. It is usually self-limited and the treatment is supportive.

Take-Home Message
Vesicles on the hands, feet, perioral, and diaper area are characteristics of HFMD.

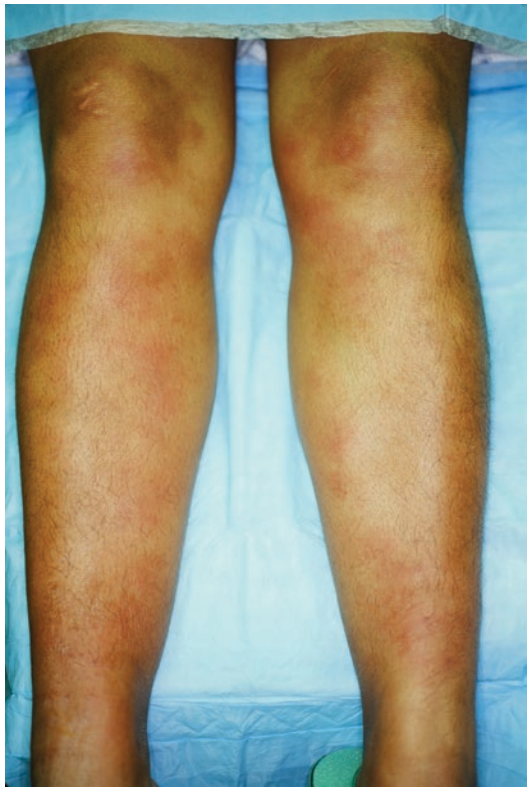
Question 5

A 15-year-old girl presents to the emergency department with painful bumps on her shins. She denies abdominal pain, vomiting, or fever. On physical examination she has tender subcutaneous nodules on her shins bilaterally. She denies recent trauma to her legs and had no improvement after applying ice to the affected areas. Which of the following is true?

A. This rash may be associated with streptococcal pharyngitis
B. She has erythema nodosum associated with irritable bowel syndrome
C. The rash is characteristic of cold panniculitis

D. The rash is due to necrosis of the subcutaneous fat
E. The condition is equally common in women and men

Correct Answer: A



Erythema nodosum

Erythema nodosum (EN) is a panniculitis, or inflammation in the fat, which is characterized by erythematous tender nodules typically localized to the anterior surface of the shins. The thighs and forearms are occasionally involved.

EN is a delayed-type hypersensitivity reaction and is rarely seen in children less than 2 years of age. In most cases the cause is unknown; however, associated conditions include group A streptococcal infection, inflammatory bowel disease, tuberculosis, and sarcoidosis. Medications (such as oral contraceptives), pregnancy, and malignancy have also been implicated. Episodes usually resolve spontaneously within several weeks and may recur. Treatment should be directed towards the underlying disorders.

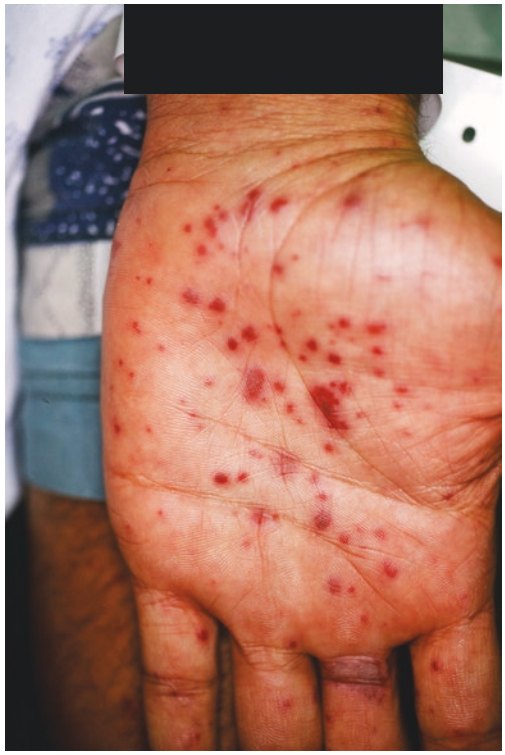
Cold panniculitis occurs within a few hours to days following exposure to cold and is characterized by erythematous plaques or subcutaneous nodules in healthy individuals. Infants and children are particularly susceptible. In this case, the nodules developed prior to the application of ice.

Take-Home Message
Erythema nodosum is a panniculitis requiring evaluation for underlying systemic disease. Common triggers include streptococcal throat infection, inflammatory bowel disease, sarcoidosis, pregnancy, and medications.

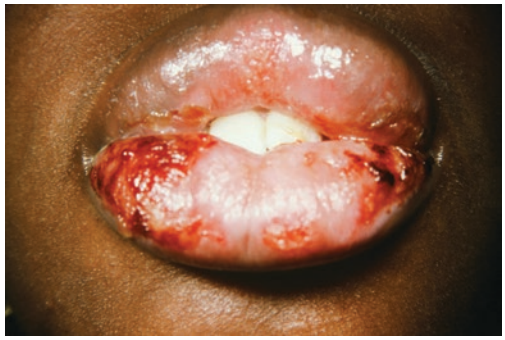
Question 6

An 11-year-old boy presents to the emergency department with an expanding red rash on his left leg. On examination, there is an annular erythematous patch. He complains of a headache and myalgias. He recently returned from a visit to Vermont. All of the following statements are true *except*:

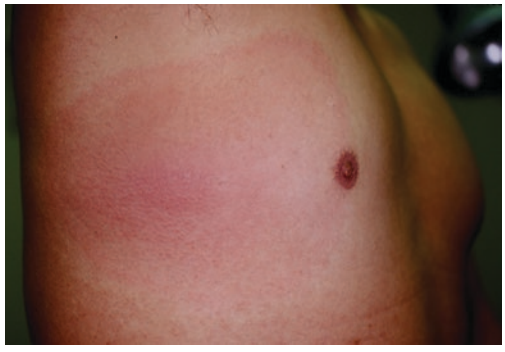
A. This rash is not typical of tinea corporis
B. This rash can be associated with AV nodal conduction blocks
C. He most likely does not have erythema multiforme
D. The rash is caused by *Borrelia burgdorferi*
E. The rash is not consistent with Lyme disease without a history of a tick bite and should not be treated



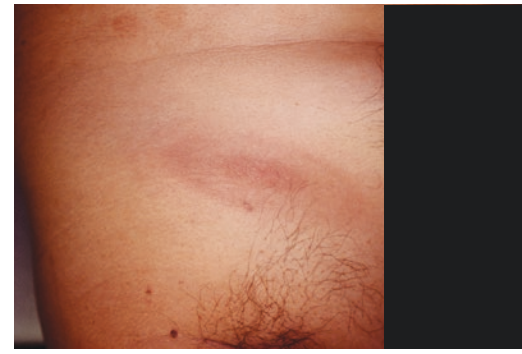
Erythema multiforme



Erythema multiforme



Lyme



Lyme

Correct Answer: E

This child has constitutional symptoms, an annular rash consistent with erythema chronicum migrans (ECM) and possible tick exposure. This suggests a diagnosis of Lyme disease caused by the spirochete *Borrelia burgdorferi* and requires treatment. It is the most common clinical manifestation of Lyme disease. The rash begins at the site of inoculation with spreading erythema expanding several centimeters per week. An annular appearance develops as the edge continues to advance and the center clears. Most cases do not itch.

Untreated Lyme disease can be complicated by heart disease manifested by conduction delays, myocarditis, and pericarditis. Neurologic symptoms can include cranial nerve palsies and meningitis. Arthritis is the most common symptom of disseminated infection.

Amoxicillin is the treatment of choice for children less than 8 years of age and doxycycline is preferred in older children.

Tinea corporis classically presents with annular erythematous patches or plaques with scale. The lesions are pruritic. Partial resolution can occur spontaneously leaving areas of post-inflammatory hyperpigmentation. Topical antifungals can be effective, though widespread involvement may require systemic treatment.

Erythema multiforme is a cytotoxic hypersensitivity reaction characterized by a target lesion. The classic target has an erythematous to dusky center surrounded by a ring of paler erythema and edema and a brightly erythematous border. Due to necrosis, the center may develop a vesicle

or erosion. Extensive mucosal involvement can occur. In children, herpes simplex is the most common triggering infection.

Take-Home Message
An annular rash after potential tick exposure requires empiric treatment for Lyme disease. It is important to note a history of tick bite is only elicited in approximately half of patients with ECM.

Question 7

A 3-year-old child presents to the emergency department with red patches on his cheeks. On examination, he is well appearing and has erythematous reticular patches on the trunk and extremities. Which of the following is true?

A. The child is infectious
B. The condition is associated with rheumatoid arthritis in children
C. In utero infection poses no risk to a fetus
D. This virus is associated with aplastic crisis
E. The child should be given topical corticosteroids for eczematous dermatitis

Correct Answer: D

Erythema infectiosum, also known as fifth disease, is caused by parvovirus B19. The exanthem is characterized by facial erythema with a "slapped-cheek" appearance. The rash then spreads to the trunk and proximal extremities with diffuse macular erythema evolving into a lacy, reticular pattern. Patients are only contagious during the prodromal viremic phase prior to the onset of cutaneous findings. For this reason, isolation and exclusion from school or childcare are unnecessary and ineffective after diagnosis. Exposure in nonimmune pregnant women has potentially severe consequences to the fetus, including anemia, congestive heart failure, hydrops fetalis, and intrauterine demise. This infection is associated with aplastic crisis in predisposed individuals. Joint involvement is more common in adults than children. The rash is self-limited and not responsive to topical corticosteroids.

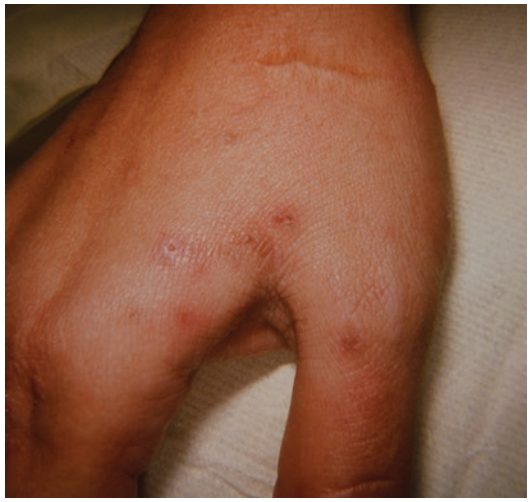
Take-Home Message
Parvovirus is a self-limited viral illness. The presence of rash indicates a patient is no longer contagious. Infection during the first half of pregnancy can have devastating consequences such as fetal loss or hydrops fetalis.

Question 8

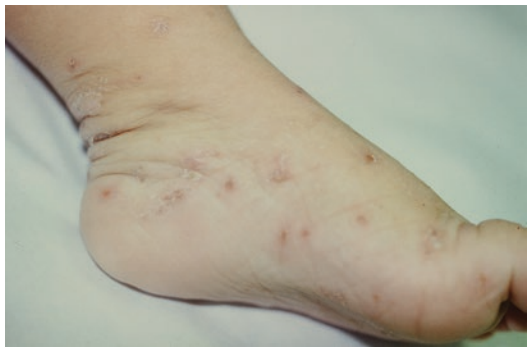
A father brings his 9-month-old son in with a pruritic rash that developed after a month-long visit with his mother. On exam, he has papules and vesiculopustules most densely on the axillae, hands, feet, and penis. The best treatment is:

A. Lindane from head to toe for the entire family
B. Permethrin 5% cream from head to toe overnight
C. Topical corticosteroids for the itching
D. Permethrin for all the adults but not the infant
E. Oral ivermectin 200 mcg/kg

Correct Answer: B



Scabies



Scabies

The presentation is consistent with a scabies infestation, caused by the mite *Sarcoptes scabiei hominis*. The incubation period for scabies is approximately 3–4 weeks. Characteristic findings include burrows, which represent the intraepidermal tunnel created by the moving female mite. Inflammatory papules and nodules are typically found on the ventral wrists, axillae, breasts, umbilicus, and genitalia. Nodules on the male genitalia can be a particularly important clinical finding. Infants commonly have involvement of the palms and soles, and unlike adults, the scalp and face may be affected.

Permethrin 5% cream is FDA approved for infants 2 months and older; it is the treatment of choice. Treatment should be applied to all skin surfaces including the genitalia and beneath the nails. In children, permethrin should be applied to the scalp and face. Lindane is not recommended for infants due to CNS toxicity. Additionally, there is a high rate of resistance. Ivermectin is an oral antihelminthic used for a variety of parasitic infections worldwide. It causes paralysis of parasites by affecting GABA-mediated nerve synapses and is available in topical and oral formulations. The safety of ivermectin has not been established in patients weighing less than 15 kg. All household members and intimate contacts should be treated simultaneously due to the risk of transmission via asymptomatic carriers.

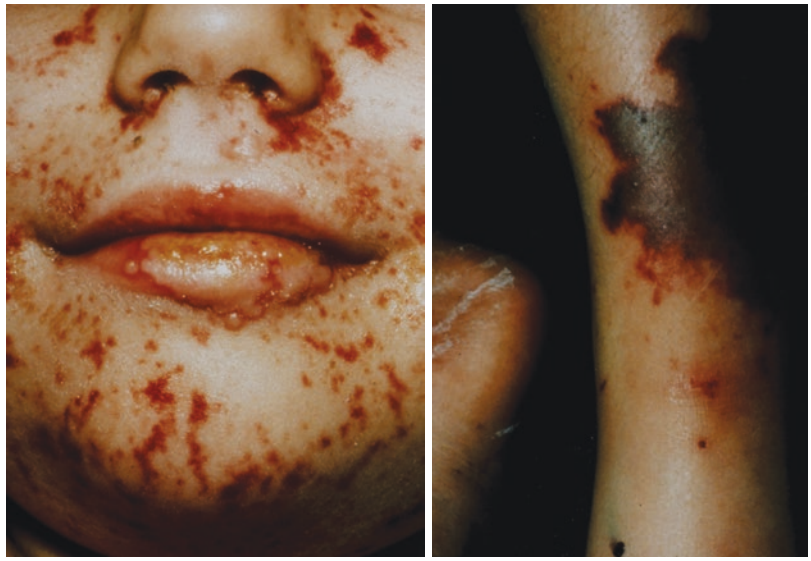
A topical steroid may be given for the associated hypersensitivity reaction and pruritus, after treatment with permethrin.

Take-Home Message
Eradication of scabies requires appropriate application of topical therapy and treatment of close contacts. Unlike adults, involvement of the scalp and face as well as acral pustules are seen in children.

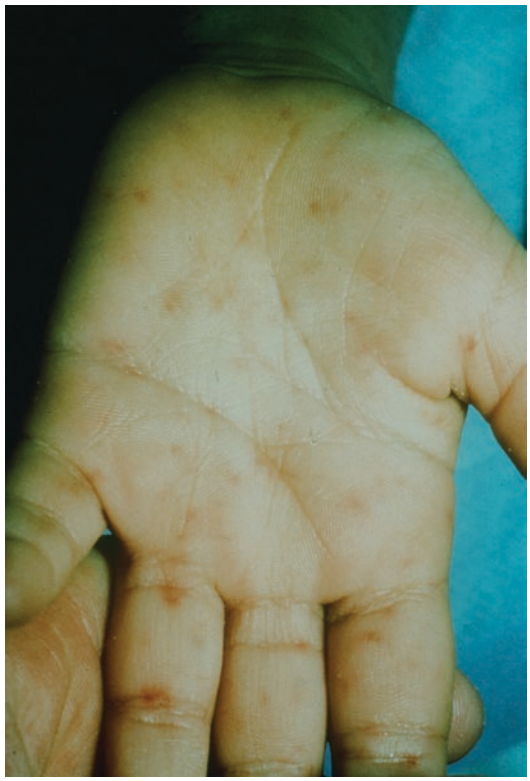
Question 9

An 11-year-old boy recently arrived from Europe after a visit with his family. He has had a few days of cough and rhinorrhea followed by macular erythema of the face which has spread to the body. He also has injected conjunctivae which the family believes is due to the high fever and lack of sleep after his long flight home. His vaccinations are not up to date. He most likely has:

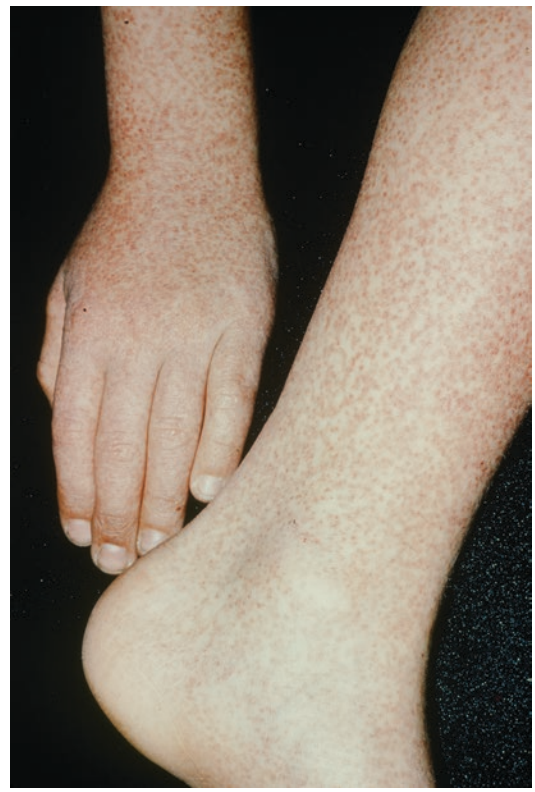
A. Meningococcemia
B. Rocky Mountain spotted fever
C. Measles (rubeola)
D. Erythema infectiosum (fifth disease)
E. Scarlet fever



Meningococcemia



Rocky mountain spotted fever



Atypical measles

Correct Answer: C

The most likely diagnosis in this child is measles, which presents with fever and the 3 Cs: cough, coryza, and conjunctivitis. During this prodrome, some patients will develop Koplik spots, bluish-white macules on the buccal mucosa, which are pathognomonic. Around day four, a rash begins on the face and spreads to the trunk and extremities. Initially macular, the rash evolves to fine papules and may be slightly pruritic. Complications include secondary bacterial infections such as pneumonia and otitis media; encephalitis is a rare but serious complication. Routine vaccination of children has dramatically reduced the incidence of measles worldwide.

Meningococcemia is caused by a gram-negative organism, *Neisseria meningitidis*. Classic skin findings include petechiae or purpuric plaques due to angioinvasion by the organism and subsequent development of disseminated intravascular coagulation. Patients are critically ill.

In Rocky Mountain spotted fever (RMSF), symptoms include headache, gastrointestinal symptoms, malaise, and myalgias followed by fever and rash. The petechial rash begins on the wrists, ankles, palms, and soles and then spreads centrally. The classic triad of RMSF consists of fever, headache, and rash; however, patients may not present with all findings. A history of a tick bite may or may not be present. Doxycycline is the treatment of choice.

The rash of erythema infectiosum presents with "slapped-cheek" facial erythema followed by a lacy, reticulated eruption on the trunk and extremities. It is caused by human parvovirus B19. Conjunctivitis is not a characteristic finding.

Take-Home Message

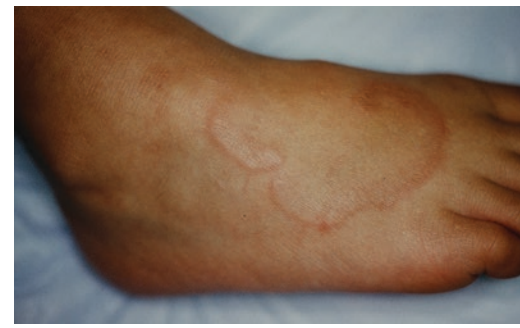
The classic triad of measles includes cough, coryza, and conjunctivitis. Koplik spots are considered pathognomonic and occur before the exanthematous stage.

Question 10

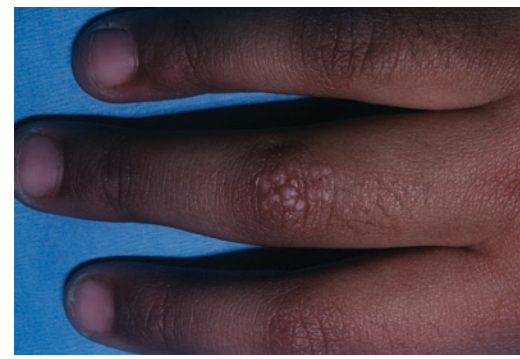
A 4-year-old boy presents with a solitary, asymptomatic lesion on his lower leg. On exam, he has an annular, erythematous plaque with small papules peripherally. There is no scale. His doctor prescribed antifungal cream without improvement. The next best step is:

A. No treatment as the condition is self-limited
B. Systemic antifungals
C. Topical corticosteroids
D. Combination antifungal/corticosteroid cream
E. Systemic antibiotics

Correct Answer: A



Granuloma annulare



Granuloma annulare

This patient most likely has granuloma annulare (GA). The clue is the absence of scale and lack of response to topical antifungals. The cause of GA is unknown, though some studies suggest an association with diabetes mellitus. Potent topical corticosteroids may be helpful; however, given the self-limited and asymptomatic nature and potential side effects of topical corticosteroids, observation is an acceptable approach.

Systemic griseofulvin is the treatment of choice for tinea capitis or extensive tinea corporis. The diagnosis of dermatophyte should be confirmed with microscopic examination of skin scrapings and fungal culture.

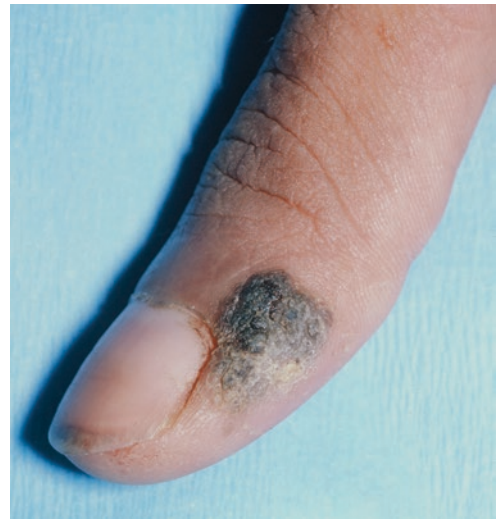
Combination treatments with antifungal and corticosteroids should generally be avoided for their potential for misuse and adverse effects such as atrophy, dyschromia, and telangiectasias.

Take-Home Message
Granuloma annulare is a benign skin condition which presents as annular, dermal papules or plaques.

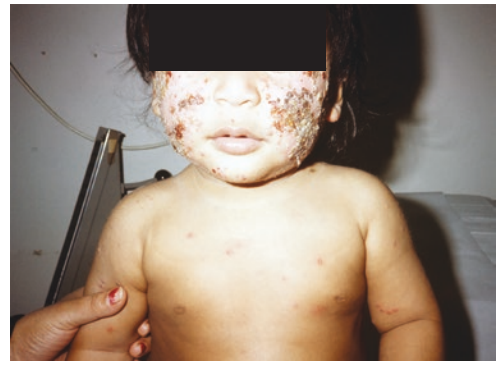
Question 11

A 3-year-old boy is brought to the emergency department with a several-month history of a rash on the left thigh, now with a nodule in the groin. On examination, there is a 1-cm nontender nodule in the left inguinal fold without overlying erythema. On the same thigh there are many skin-colored pink papules with central umbilication, some with heme-colored and honey crust. Your primary diagnosis is:

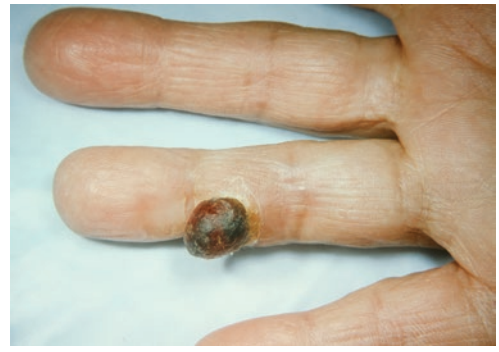
A. Verruca vulgaris
B. Pyogenic granuloma
C. Cold panniculitis
D. Molluscum contagiosum
E. Impetigo



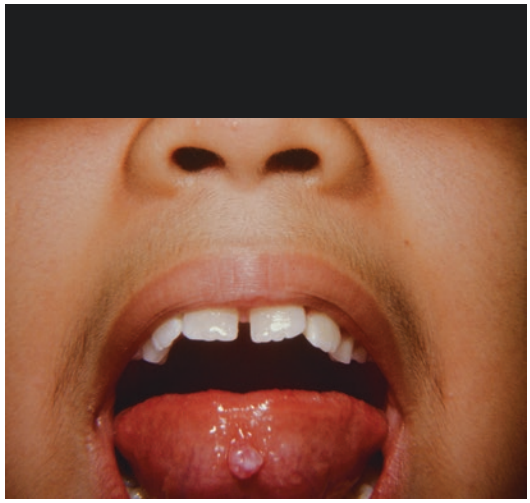
Wart



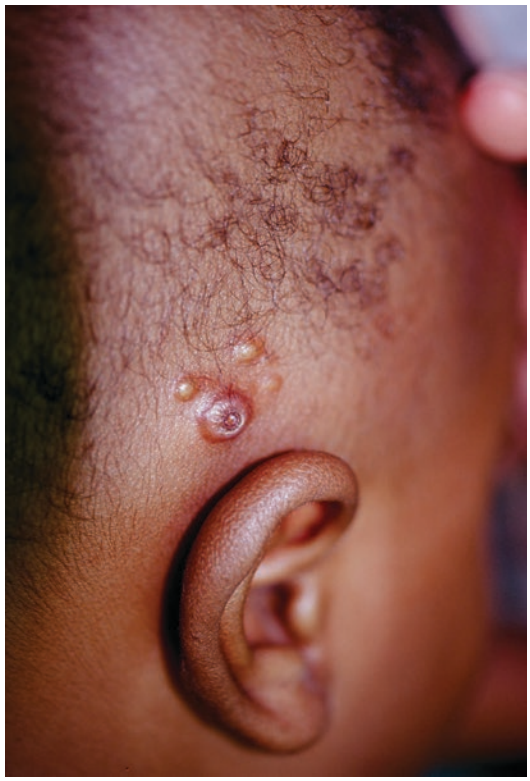
Impetigo



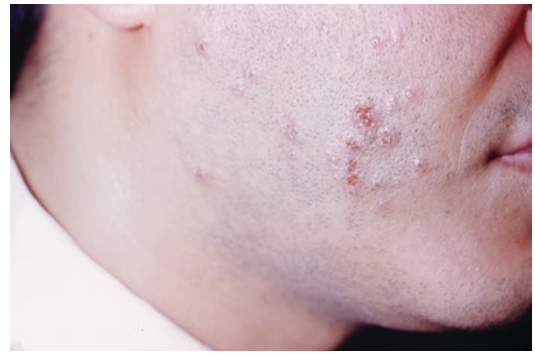
Pyogenic granuloma



Pyogenic granuloma



Molluscum contagiosum



Impetigo

Correct Answer: D

Skin-colored to pink, umbilicated papules are typical of molluscum contagiosum (MC). Caused by the poxvirus, the condition is benign and self-limited. Occasionally, individual lesions may become inflamed, which may indicate impending resolution. Destructive methods may be considered for treatment, if tolerated by the patient; however, watchful waiting is a reasonable choice. MC may become secondarily impetiginized resulting in regional lymphadenopathy. When MC involves an area of atopic dermatitis, topical corticosteroids should be continued for the treatment of eczema to prevent further spread via scratching.

Verruca vulgaris, the common wart, is a benign tumor caused by human papillomavirus (HPV) infection of the skin. These hyperkeratotic papules do not have central umbilication. Like molluscum, warts are benign and resolve spontaneously. A variety of topical and destructive therapies are available if treatment is desired.

Pyogenic granuloma (PG) is a benign, acquired vascular tumor presenting as an exophytic friable papule. A collarette of scale at the base is characteristic. Ulceration and bleeding prompt patients to seek medical attention. PG is treated by shave removal followed by destruction of the base with electrodesiccation to prevent recurrence.

Cold panniculitis is characterized by painful erythematous plaques or subcutaneous nodules that occur in areas exposed to cold.

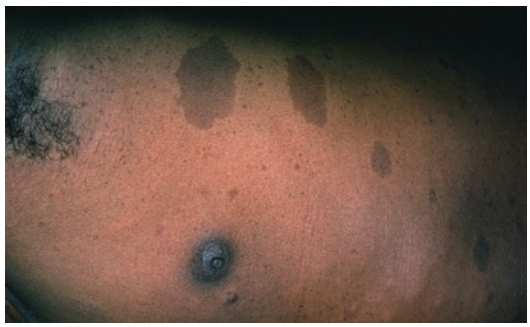
Impetigo can be bullous or non-bullous. The non-bullous type is the most common form of impetigo, easily recognized by its characteristic honey-colored crust. In the bullous form, vesicles enlarge to form flaccid bullae with clear yellow fluid. Often, patients will present with erosions and a collarette of scale. Bullous impetigo is the result of exfoliative toxin, which is also implicated in staph scalded skin syndrome.

Take-Home Message
The primary lesion of molluscum contagiosum is a skin-colored to pink umbilicated papule. Secondary impetiginization with *Staphylococcus aureus* is not uncommon.

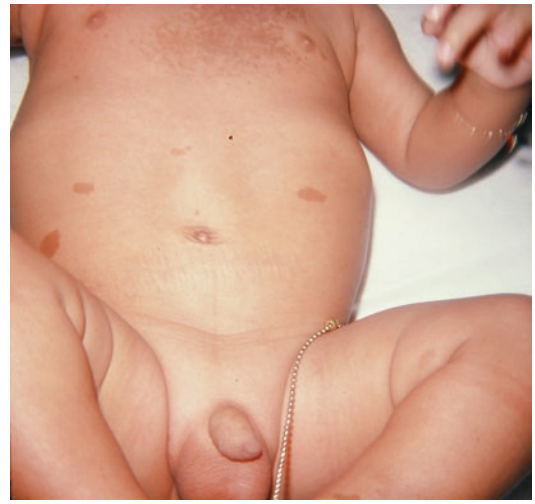
Question 12

A 4-year-old boy is brought to the emergency department due to multiple seizures. On examination there are hypopigmented macules over the stomach and back. He also has firm pink-brown papules on the nasolabial folds and cheeks. After admission to the hospital, the EEG shows a hypsarrhythmic pattern and MRI of the head shows subependymal nodules. This constellation of features is consistent with:

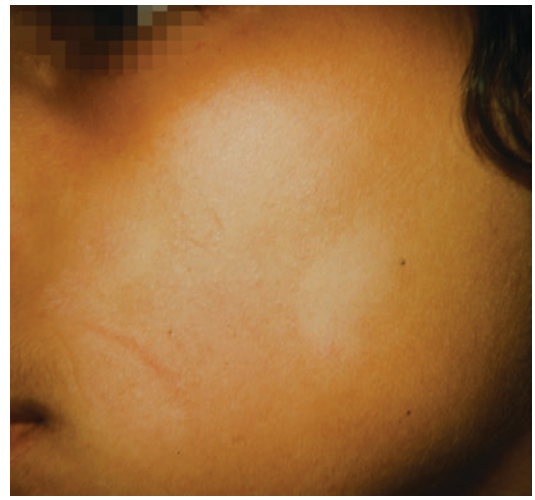
A. West syndrome
B. Tuberous sclerosis complex (TSC)
C. Neurofibromatosis
D. Pityriasis alba
E. Sturge–Weber syndrome



Neurofibromatosis



Neurofibromatosis



Pityriasis alba

Correct Answer: B

This child has the typical lesions of tuberous sclerosis complex (TSC). TSC occurs by autosomal dominant inheritance; however, the majority of cases are thought to be due to new mutations. Patients with TSC may have any of the following cutaneous findings:

- Hypopigmented macules
- Confetti skin lesions
- Angiofibromas (adenoma sebaceum)
- Fibrous forehead plaque
- Shagreen patch (connective tissue nevus)

- Ungual or periungual fibromas
- Gingival fibromas

The majority of patients will have cutaneous findings: angiofibromas, periungual fibromas, the shagreen patch, and hypopigmented ash-leaf-shaped macules, which are considered pathognomonic. The classic features of TSC include skin changes, intellectual impairment, and epilepsy; however, early diagnosis is important to screen for hamartomas of the eye, kidney, or heart. Prognosis is largely dependent on the extent of neurologic involvement.

West syndrome is a severe epilepsy syndrome with infantile spasms, characteristic EEG pattern termed hypsarrhythmia, and mental retardation. There are no cutaneous lesions associated with West syndrome.

Neurofibromatosis is a neuroectodermal genodermatosis. Inheritance is autosomal dominant, though up to 50% of cases occur due to spontaneous mutation. The cutaneous findings in neurofibromatosis type I include café au lait spots, axillary freckling, and neurofibromas. Epilepsy can occur; however, neurologic problems occur in less than half of patients.

Pityriasis alba is a common skin disorder in children characterized by the presence of ill-defined, hypopigmented patches, usually on the face, neck, or upper trunk. There may be fine scale present. The condition is considered a marker for atopy.

Sturge–Weber syndrome (SWS) is a sporadic neurocutaneous disorder characterized by a facial capillary malformation. The port-wine stain follows the distribution of the trigeminal nerve, usually the first branch (V1). Early recognition is important to evaluate for leptomeningeal angiomatosis, which may cause seizures and glaucoma.

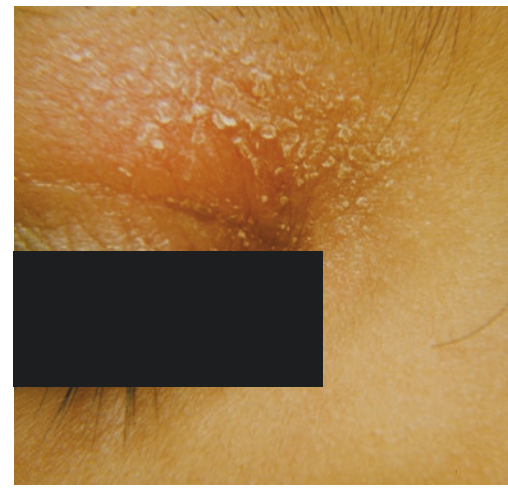
Take-Home Message

Three or more hypopigmented macules present at birth should prompt consideration of tuberous sclerosis.

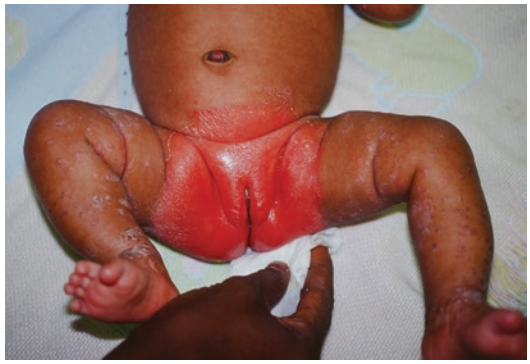
Question 13

An 18-year-old girl was sunbathing and noticed a non-itchy rash on her upper trunk and neck. She denies any recent illnesses or systemic symptoms at the present time. In the emergency department, she appears well, and her skin exam reveals multiple coalescing tan macules with scale. The rest of her physical exam is normal. What is the most likely diagnosis?

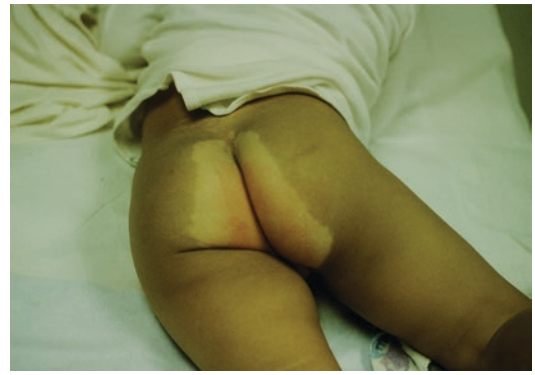
A. Psoriasis
B. Pityriasis versicolor
C. Sarcoidosis
D. Vitiligo
E. Insect bites



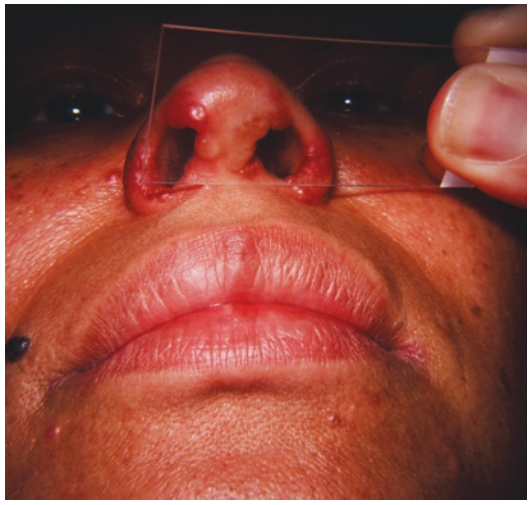
Psoriasis



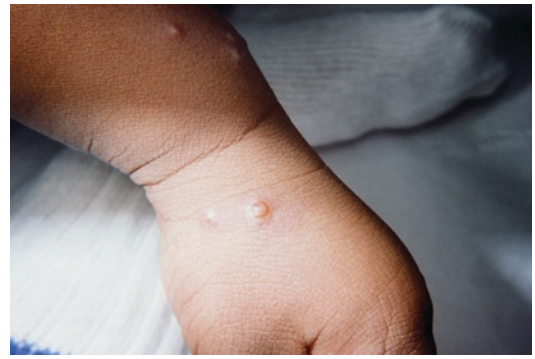
Psoriasis



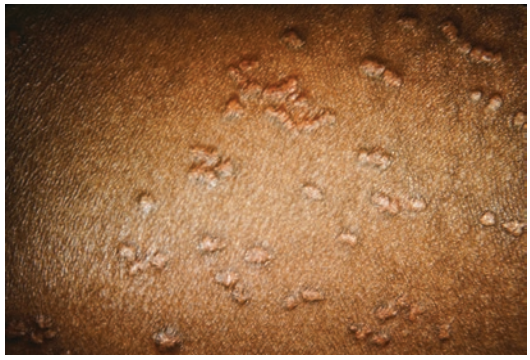
Vitiligo



Sarcoid



Insect bite



Sarcoid

Correct Answer: B

This patient has tinea versicolor, caused by *Malassezia furfur*. It is a superficial fungal infection, usually involving the upper trunk and upper arms. Typical lesions are skin-colored tan-pink macules with scale, often coalescing into large patches. Patients are usually asymptomatic, although mild pruritus may occur. The diagnosis can be made on the basis of the clinical appearance of lesions and their distribution. Potassium hydroxide (KOH) scraping shows numerous round yeasts with short hyphae. A Wood lamp examination may show a characteristic yellow-orange fluorescence. Hypopigmentation may occur, due to inhibition of melanin production by dicarbox-

ylic acid production from metabolism of skin surface lipids by the yeast.

Vitiligo presents with well-demarcated depigmented macules and patches. The distribution is characteristic around the eyes, mouth, genitals, elbows, hands, and feet. It is caused by a loss of melanocytes within the skin. The hair of affected areas can also depigment. Vitiligo is associated with other autoimmune endocrinopathies, most commonly thyroid disease. A family history and review of systems therefore are important parts of the evaluation.

Sarcoidosis is a systemic, granulomatous disease which is rare in children. It is characterized by non-necrotizing granulomas in affected organs, most commonly the lungs. Cutaneous lesions include yellow-brown to red papules and nodules. Erythema nodosum may precede or accompany the disease. Ophthalmology referral is essential to evaluate for uveitis.

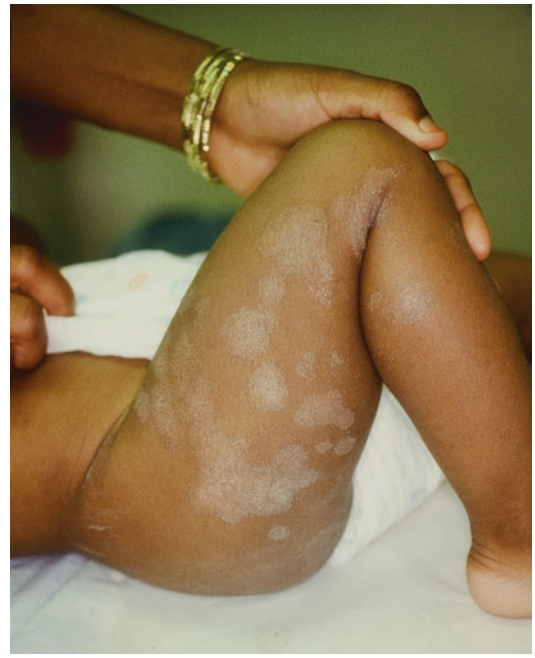
Psoriasis is characterized by sharply demarcated erythematous plaques with silvery scale, most commonly on the scalp, extensor surfaces, and gluteal cleft. Examination of the nails may reveal pitting or onycholysis.

Take-Home Message
Tinea versicolor is a superficial fungal infection causing temporary hypopigmentation. It should be differentiated from tinea corporis and pityriasis rosea as the treatment is different.

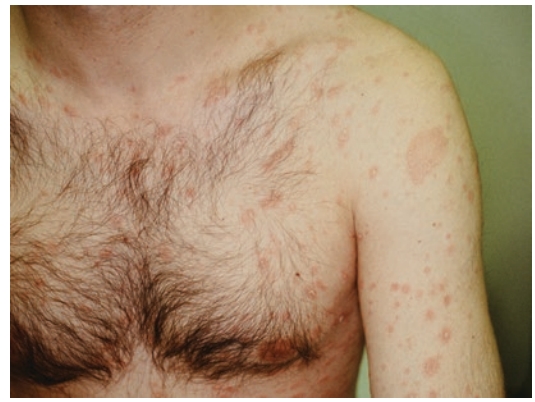
Question 14

A mother brings her 3-month-old girl for evaluation of a rash of a week's duration. There are erythematous patches with scale on the scalp, neck, underarms, and diaper area. The family history is notable for asthma in her older brother. What is the most likely diagnosis?

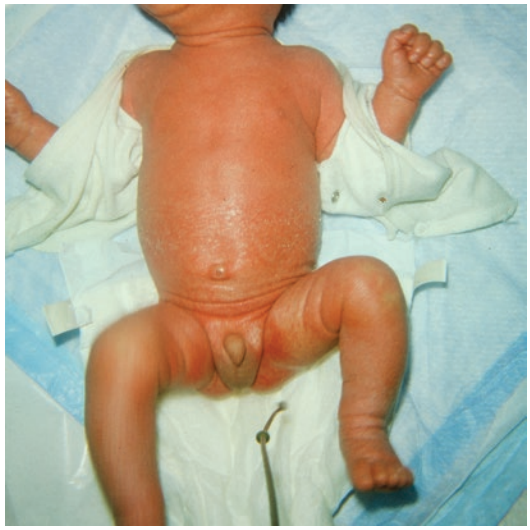
A. Atopic dermatitis
B. Psoriasis
C. Seborrheic dermatitis
D. Langerhans cell histiocytosis
E. Tinea corporis



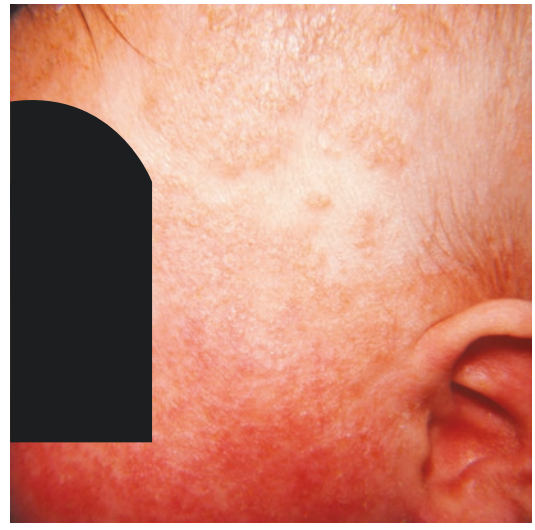
Atopic Dermatitis



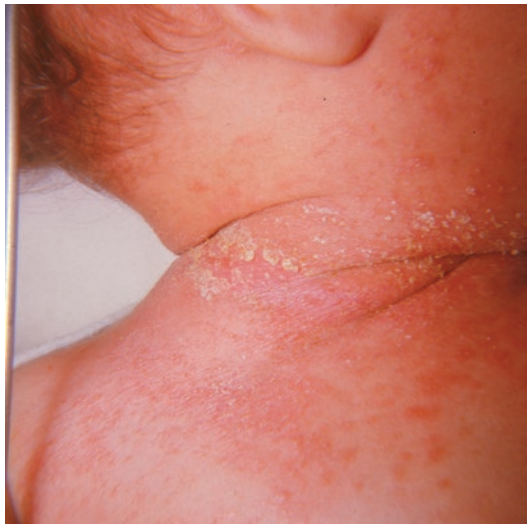
Pityriasis Rosea



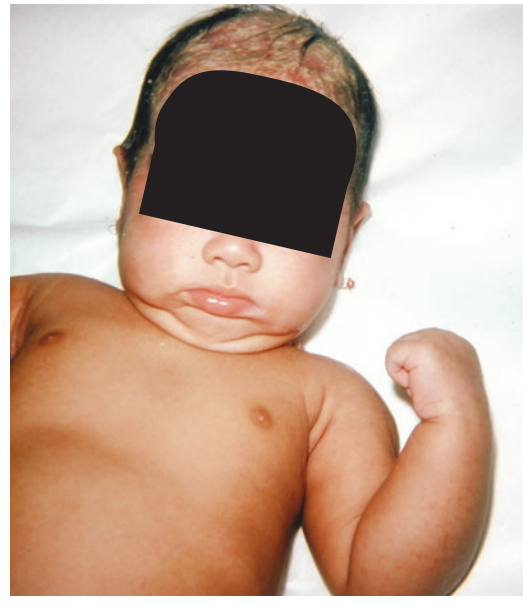
Seborrheic Dermatitis



Seborrheic Dermatitis



Seborrheic Dermatitis



Seborrheic Dermatitis

Correct Answer: C

The rash described is seborrheic dermatitis. It is an erythematous, scaly, symmetric eruption involving hair-bearing (scalp) and intertriginous regions. In infants, it usually presents in the first month of life and incidence peaks at 3 months of age. Many infants with atopic dermatitis have a history of infantile seborrheic dermatitis. Involvement of the diaper area can be a useful clinical clue to the diagnosis of seborrheic dermatitis. In atopic dermatitis, by contrast, the diaper area is usually spared as it is a moist area and the diaper inhibits infants from being able to scratch and worsen the rash.

Atopic dermatitis (AD) is a chronic, pruritic inflammatory skin disease; it is one of the most common dermatoses of infants and children. AD and seborrheic dermatitis may overlap, with features of AD becoming more prominent after 3 months of age. In infants, lesions are symmetrically distributed over the cheeks, forehead, scalp, trunk, and extensor surfaces of the extremities. In children, flexural surfaces are most commonly involved. The course involves remissions and exacerbations. These children may present to the ED with intense pruritus. The majority of children may improve by puberty. Associated conditions include asthma and allergic rhinitis.

Psoriasis presents with discrete, sharply demarcated erythematous plaques with thick scales. The scale is adherent and silvery white leaving pinpoint bleeding when removed (Auspitz sign). The inverse form affects intertriginous areas and can have a similar appearance to seborrheic dermatitis. The lesions are usually symmetric and common sites involved are the scalp, elbows, and knees. Compared to seborrheic dermatitis, psoriasis has a slower response to treatment and requires potent to ultrapotent topical corticosteroid treatment. Systemic steroids should not be used because of risk of rebound or induction of pustular psoriasis.

Langerhans cell histiocytosis (LCH) should be considered in the differential diagnosis of seborrheic dermatitis refractory to conventional treatment. The presence of yellow-red crusted papules, erosions, or purpura should raise concern for LCH. Many organs may be involved including bone, lung, liver, hypothalamus, posterior pituitary, and the lymphatic system. A skin biopsy can distinguish LCH from seborrheic dermatitis.

The classic lesion of tinea corporis is a pruritic, annular lesion with central clearing. The border is raised with an advancing scale and may have papules or vesicles. The lesions may be single or multiple. The diagnosis can be confirmed by KOH examination of skin scrapings. Lesions may be mistaken for nummular eczema, psoriasis, or erythema annulare centrifugum, as well as the herald patch of pityriasis rosea.

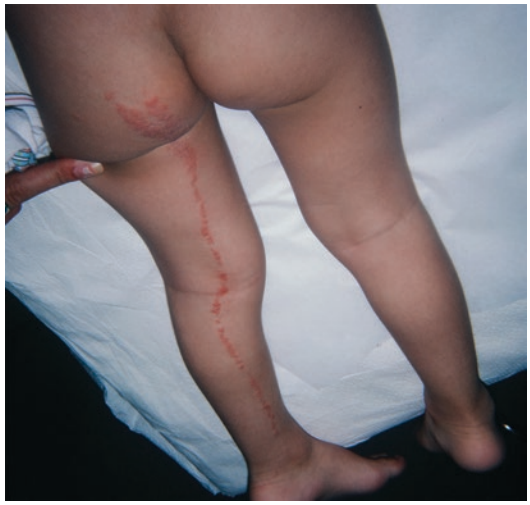
Take-Home Message
Seborrheic dermatitis is an inflammatory skin condition characterized by erythematous, scaly, symmetric eruption that occurs most often in hair-bearing and intertriginous regions.

Question 15

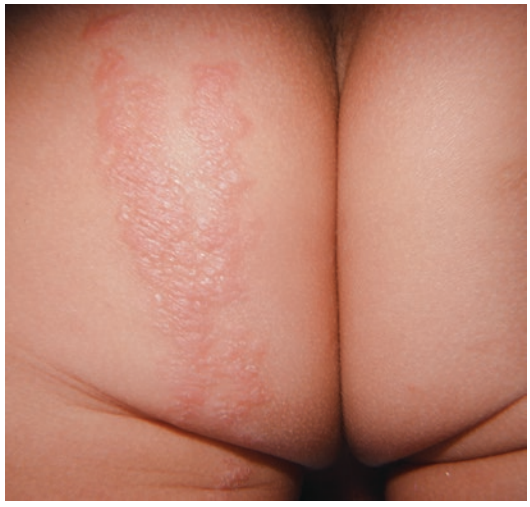
A 5-year-old boy has had a non-pruritic rash on his arm for several days. His mother decided to bring him to the emergency department as it appears to be expanding. On examination, you note flat-topped papules in a linear distribution. What is the best treatment?

A. No treatment
B. Topical vitamin D
C. Topical corticosteroids
D. Tar therapy
E. Oral antihistamines

Correct Answer: A



Lichen striatus



Lichen striatus

This rash is characteristic of lichen striatus. Lichen striatus is a linear, papular eruption of unknown etiology, which is a self-limited condition, most commonly affecting children. This rash begins as 2–4-mm lichenoid papules that coalesce to form a curvilinear band along Blaschko's lines.

The lesions usually occur on the trunk and face and may be erythematous and scaly. Pruritus may occur, especially in children who are also atopic. Nail changes such as longitudinal ridging and nail plate thinning may occur. Since it usually resolves spontaneously, therapy is not required.

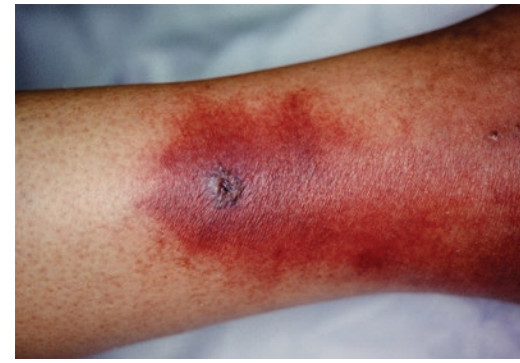
Take-Home Message
Linear, asymptomatic papules are characteristic of lichen striatus.

Question 16

A teenage boy was cleaning the attic with his mother and going through some old clothes. The next morning upon awakening he noticed his arm was painful with blue discoloration. The most likely cause of his symptoms is:

A. A bee sting
B. Erythema migrans
C. Brown recluse spider bite
D. Stevens–Johnson syndrome
E. Pediculosis corporis

Correct Answer: C



Spide bite (brown recluse)

These symptoms are consistent with a brown recluse spider bite. In the United States, *Loxosceles reclusa* is the predominant species. Bites usually occur when spiders become trapped in attics, basements, or storage closets. The initial bite is often painless and therefore may remain unnoticed. Pain usually begins hours later, and an erythematous area with a central pustule or hemorrhagic vesicle may develop. Ultimately, there may be gangrenous ulceration or necrosis. The venom contains sphingomyelinase D, which is mainly cytotoxic, causing local tissue destruction by destroying endothelial cells; it also has a

hemolytic component, which may result in hemolysis. Systemic symptoms such as fever, myalgias, and hemolysis are rare but develop more commonly in children due to toxin burden.

ECM, the cutaneous form of Lyme disease, presents with annular erythema. There is no associated eschar or ulceration. The skin changes are not painful; however, there may be associated fever, fatigue, headache, and arthralgias. A tick bite may not be reported.

Pediculosis corporis, body lice, presents with generalized pruritus. Clinical manifestations include pruritus, excoriations, and small, red macules. The organism lives and reproduces in the lining of clothes and leaves the clothing only for feeding from the skin; therefore, examination of clothing is important. It should be suspected in children with generalized itching, excoriations and poor hygiene. All clothing and bedding should be washed in hot water or dry cleaned. All household contacts should be treated.

Stevens–Johnson syndrome (SJS) is a rare, life-threatening systemic hypersensitivity reaction. It is a T-cell-mediated toxic reaction targeting the basement membrane resulting in separation at the dermal-epidermal junction. Mucous membrane involvement is a characteristic finding. Usually a prodrome of fever, malaise, and sore throat precedes the rash. The palms and soles may be an early site of involvement. Targetoid lesions, dusky macules and patches, bullae, and erythroderma may be seen. The Nikolsky sign may be a helpful clinical clue in patients who have yet to develop epidermal detachment. The distinction between SJS and toxic epidermal necrolysis (TEN) is based on body surface area involvement and is important prognostically. SJS affects ≤10% of the total body surface area and TEN ≥30% of the body surface area. Medications are the most common cause including antibiotics such as sulfonamide, cephalosporins, and anticonvulsants such as carbamazepine, phenytoin, phenobarbital, valproic acid, and lamotrigine. Other agents include nonsteroidal anti-inflammatory drugs. *Mycoplasma pneumoniae* has also been identified in pediatric patients without drug exposure.

Take-Home Message
Most spiders in the United States cause only local pain, redness, and swelling. Body lice are more likely to be found in the clothing seams than on the skin of patients.

Question 17

A 10-year-old girl is brought to the emergency department for evaluation of a painful swelling over her scalp. On examination, the posterior scalp has a boggy plaque with pustules and alopecia. All of the following are true *except*:

A. The condition is contagious
B. Topical steroids should be used to prevent permanent scarring
C. Griseofulvin is effective for most cases
D. Cervical adenopathy is common
E. Permanent alopecia is possible

Correct Answer: B

A tender plaque with pustules and purulent drainage on the scalp of a child describes a kerion, a fungal infection affecting hair follicles. It is characterized by intense inflammation and a boggy, erythematous mass. It is due to a hypersensitivity reaction to the dermatophyte infection. Cervical lymphadenopathy is common, supporting the diagnosis of tinea capitis. Local alopecia and scarring may occur if not promptly treated. Secondary bacterial infection, usually *Staph. aureus*, is common; it should be confirmed with bacterial culture and treated with systemic antibiotics. The initial lesion may be overlooked until alopecia develops. Beware that a kerion may have pustules and crusting and therefore be mistaken for an abscess.

This inflammatory form of tinea capitis requires prompt recognition and treatment with griseofulvin to prevent permanent scarring. Systemic antifungal agents such as griseofulvin for at least 6–8 weeks are required. There is no role for topical corticosteroid therapy; however, prednisone may be used to decrease inflammation and prevent scarring. Tinea capi-

tis is highly contagious; therefore, close contacts, especially prepubescent children, should be evaluated.

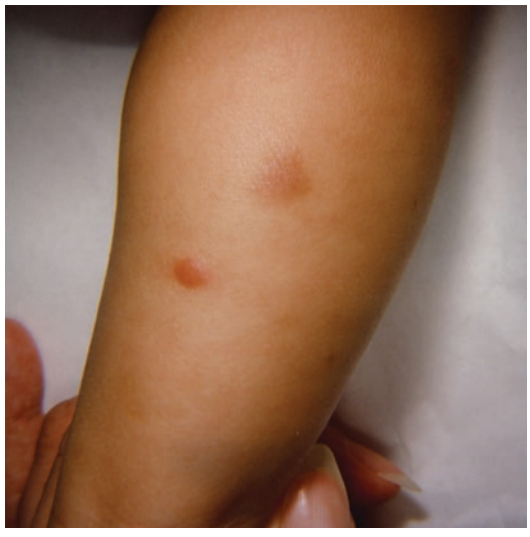
Take-Home Message
The diagnosis of kerion should be strongly considered in children with scalp infections. Prompt initiation of griseofulvin is curative and may prevent permanent hair loss.

Question 18

A 3-year-old who recently came from South America is referred by a local pediatrician for the evaluation of recurrent episodes of abdominal pain and diarrhea. On examination, she has multiple red-brown papules. With the help of an interpreter, you learn that the child was born with these lesions, which sometimes become red and swollen. When you stroke the lesions, a wheal-and-flare reaction occurs. One form of this disorder can:

A. Be treated with acyclovir
B. Result in anaphylaxis
C. Result in scarring
D. Be treated with antibiotics
E. Be treated with topical diphenhydramine

Correct Answer: B



Mastocytosis

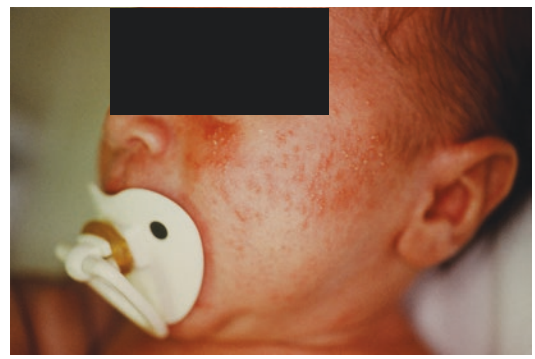
The child has urticaria pigmentosa (UP), the most common form of mastocytosis. Patients with UP have many well-demarcated tan to red-brown papules which react to stroking with a wheal-and-flare reaction. This is known as Darier's sign. The lesions predominantly affect the trunk. Mucous membranes may be involved and the palms and soles are usually spared. It may be present at birth and usually onset occurs in the first 4 years of life. It is usually limited to the skin in children and often resolves by puberty. Measures to avoid histamine release triggers are the mainstay of treatment as these patients may have an increased risk of anaphylaxis. Generally, the prognosis for childhood mastocytosis is favorable.

Take-Home Message
Darier's sign, urtication after brisk stroking of the lesions, is pathognomonic of cutaneous mastocytosis. Widespread cutaneous involvement and positive review of systems warrant evaluation for systemic mastocytosis.

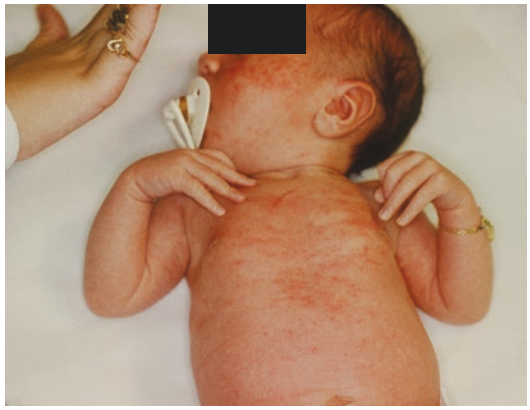
Question 19

A mother brings her 3-week-old infant for evaluation of a rash. She explains the baby had a pustular rash at birth and now has many brown spots. On examination, the baby has diffuse brown macules. The most likely cause of this rash is:

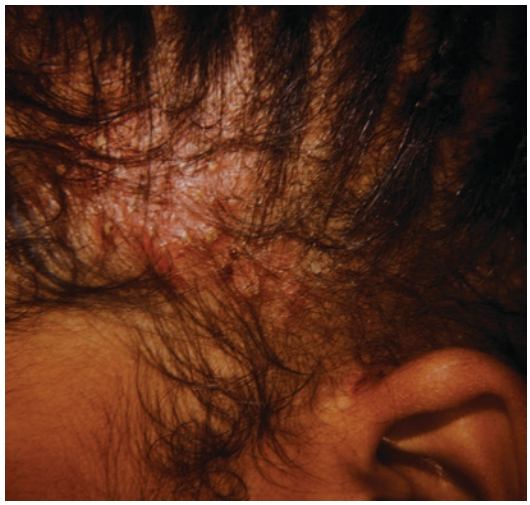
A. Transient neonatal pustular melanosis
B. Erythema toxicum
C. Milia
D. Sebaceous gland hyperplasia
E. Folliculitis



Erythema toxicum



Erythema toxicum



Folliculitis

Correct Answer: A

Transient neonatal pustular melanosis is a benign, self-limited skin disorder which commonly affects black infants. The pustules are present at birth and resolve within the first 24–48 hours of life. After rupture, the pustules leave collarettes of scale and hyperpigmentation, which may persist for weeks to months. Affected areas include the face, neck, and trunk and more rarely the scalp, palms, and soles. Neutrophils can be visualized on wright stain smears of the pustules. No treatment is needed.

Erythema toxicum is a common dermatosis that presents with blotchy erythema and yellow-white pustules in full-term neonates. The palms and soles are usually spared. Lesions typically appear in the first few days of life, waxing and waning over the first couple weeks. Examination of pustule contents with Wright's or Giemsa staining reveals eosinophils.

Milia are small retention cysts that occur on the face of newborns, most commonly on the nose. Filled with keratinocytes and sebaceous debris, they are white-yellow, 1–2-mm papules and require no treatment.

Sebaceous gland hyperplasia is a physiologic phenomenon triggered by maternal androgen exposure. The yellow-colored papules on the face of full-term infants resolve spontaneously.

Bacterial folliculitis is most commonly caused by *Staphylococcus aureus*. Treatment includes antibacterial soap, topical antibiotics, or oral antibiotics in more extensive or recurrent cases. A bacterial culture should be obtained prior to starting oral antibiotics. Twice weekly bleach baths may be helpful to prevent recurrence.

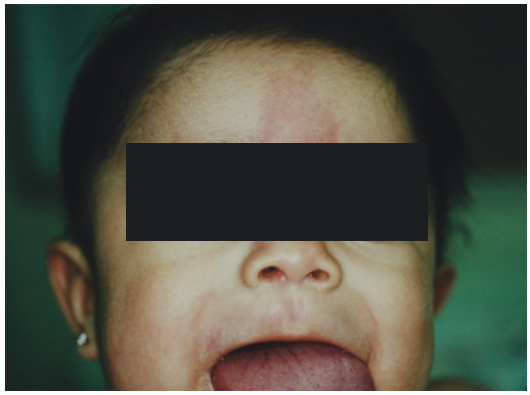
Take-Home Message

Post-inflammatory hyperpigmentation is an important clue to the diagnosis of transient neonatal pustular melanosis and may persist for weeks to months. This condition is seen most commonly in patients with dark skin.

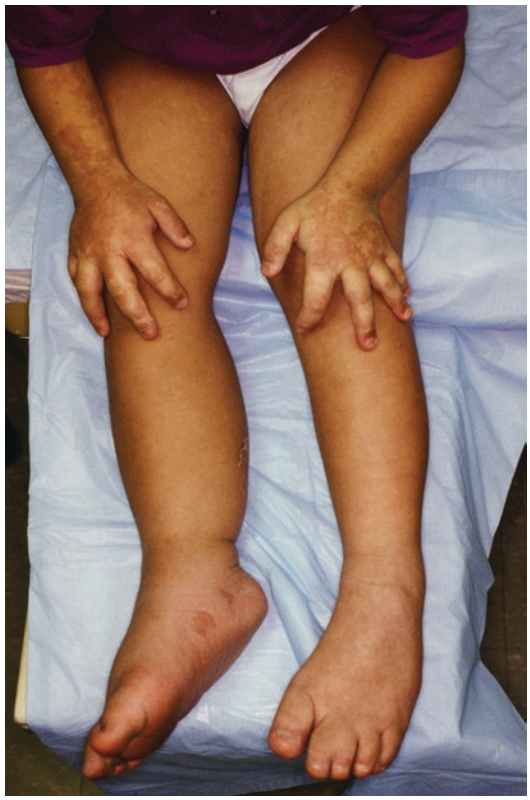
Question 20

A 4-month-old baby presents to the emergency department with a right-sided seizure that initially responds to lorazepam and then recurs. The child has no fever and was fed shortly prior to arrival. On examination, the child has a dark red patch on the right side of the face, involving both the upper and lower eyelids. The remainder of the examination is unremarkable. This child likely has:

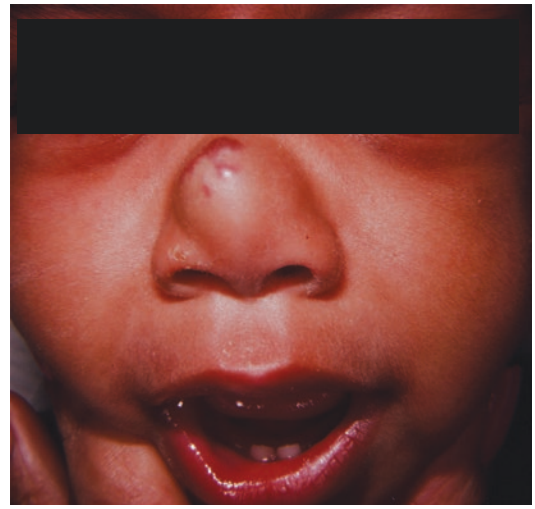
A. Sturge–Weber syndrome
B. Beckwith–Wiedemann syndrome
C. Klippel–Trenaunay syndrome
D. Arteriovenous malformations
E. Infantile hemangioma



Beckwith–Wiedemann



Klippel–Trenaunay



Hemangioma

Correct Answer: A

The presence of seizures and a facial vascular malformation suggests the diagnosis of Sturge–Weber syndrome. The classic triad includes a facial capillary malformation called a port-wine stain (PWS), leptomeningeal angiomatosis, and glaucoma. The PWS is a pink to red, non-blanching patch typically involving the upper face in the cutaneous distribution of the ophthalmic or maxillary division of the trigeminal nerve. It is usually present at birth. With age, the malformation may darken and develop hypertrophy. There is an increased risk for associated neuro-ocular complications if the lesion involves the cutaneous distribution of the ophthalmic division of the trigeminal nerve (i.e., forehead and upper eyelid). Seizures are often the first symptom to appear and are typically focal but may become generalized. The seizures usually develop before the age of five. Early referral to ophthalmology with close follow-up is recommended to screen

for glaucoma, which may be detected at birth. The presence of acute glaucoma with a cloudy cornea is an infantile emergency.

Klippel–Trenaunay syndrome (KTS) consists of PWS, anomalous veins, and progressive overgrowth of the affected extremity. This is present at birth and usually involves a lower extremity. The PWS is generally localized to the hypertrophied area. Thick-walled venous varicosities may become apparent when the child starts ambulating.

Beckwith–Wiedemann syndrome (BWS) is the most common overgrowth syndrome characterized by macroglossia, organomegaly, cardiomyopathy, and neonatal hypoglycemia due to islet cell hyperplasia. It is associated with prenatal and postnatal overgrowth. A capillary malformation of the upper central face including the bilateral upper eyelids is common. There is an increased risk of malignancy, particularly Wilms's tumor and hepatoblastoma. The placentas are large, and pregnancies are often complicated by polyhydramnios.

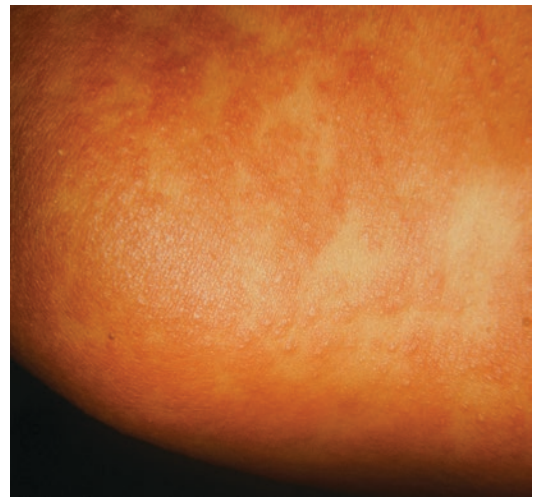
Arteriovenous malformations (AVMs) are vascular malformations with arterial and venous components. On exam they may be warm and pulsatile and have a bruit. Imaging is essential prior to considering therapeutic intervention. The most severe complication includes cardiovascular compromise.

Take-Home Message
A facial port-wine stain requires consideration of Sturge–Weber Syndrome and evaluation by neurology and ophthalmology.

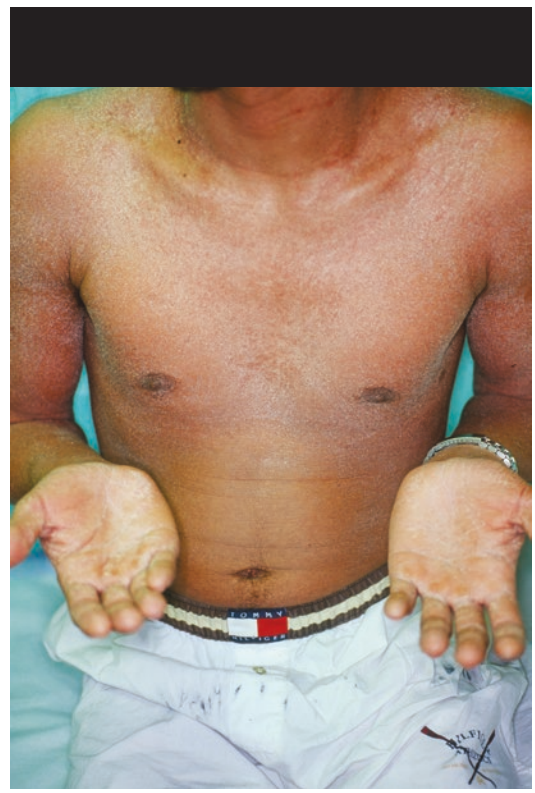
Question 21

A previously healthy 14-month-old infant is brought to the emergency department for evaluation of a fever at 1 am. The child is irritable and has right-sided crackles to auscultation. The chest X-ray shows a pneumonia and IV antibiotics were administered. In the ED, the child continues to be febrile and develops a rash. On examination, there are irregularly shaped ecchymoses and purpuric plaques with surrounding erythema on the bilateral legs. The diagnosis is:

A. Immune thrombocytopenic purpura
B. Purpura fulminans
C. Drug hypersensitivity reaction
D. Child abuse
E. Cutis marmorata



Drug hypersensitivity



Drug hypersensitivity

Correct Answer: B

Purpura fulminans (PF) is a rare clinical manifestation of disseminated intravascular coagulation (DIC). It causes ecchymotic patches and plaques and hemorrhagic necrosis of the skin due to thrombosis of the cutaneous microvasculature. Causes include severe infections such as streptococcus, meningococcus, and pneumococcus. It may also occur as a post-infectious phenomenon with varicella and scarlet fever. The congenital form due to protein C or S deficiency presents in the neonatal period. Treatment should focus on the underlying cause, in addition to supportive measures.

Immune thrombocytopenic purpura (ITP) classically occurs in a previously healthy child following a viral illness. Symptoms may include petechiae, epistaxis, and gingival bleeding. The diagnosis requires the exclusion of other causes of thrombocytopenia. The condition resolves in weeks to months and does not recur. The treatment is determined by the severity of the thrombocytopenia. In general, patients with platelet counts higher than 20,000/mm^3 who remain symptom-free may not require hospitalization. Hospitalization is required for children with platelet counts below 20,000/mm^3 with significant mucous membrane bleeding or with life-threatening hemorrhage.

A morbilliform eruption is the most common drug hypersensitivity reaction. Small pink to red macules and papules usually begin on the face and spread from head to toe on the trunk and extremities. Mucous membranes are typically spared. It occurs 7–14 days after the initial exposure to the offending agent. On subsequent exposure, the eruption will occur more quickly. Antibiotics are among the most common agents, particularly penicillin, sulfonamides, and cephalosporins.

Ecchymoses and purpura in young children raise the question of nonaccidental trauma. However, the presentation is not consistent with nonaccidental trauma in this child.

Cutis marmorata is a transient, netlike, mottling of the skin due to variable vascular constriction and dilation. This refers to physiologic livedo reticularis in response to cold. Livedo reticularis is most commonly seen on the legs. It usually resolves with increasing age.

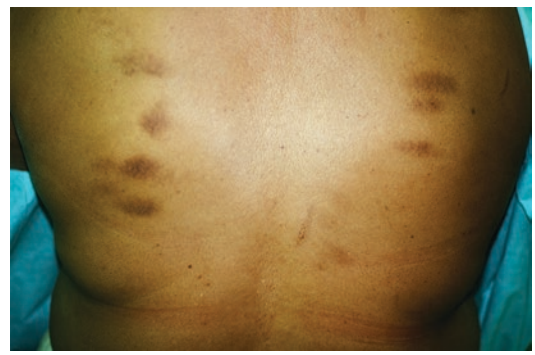
Take-Home Message
Purpura fulminans is the cutaneous presentation of disseminated intravascular coagulation in the setting of sepsis or thrombophilia. This cutaneous finding is a sign of life-threatening illness.

Question 22

An anxious resident calls you to evaluate a child with fever and runny nose as well as a rash on the back. He is worried about physical abuse because the child has multiple linear ecchymotic streaks on the back. The most likely cause of this skin finding is:

A. Gridding
B. Cupping
C. Child abuse
D. Coining
E. Acupressure

Correct Answer: D



Traumatic purpura

In general, bruises on buttocks, trunk, and back are unusual in accidental trauma. However, the pattern described is suggestive of coining or spooning which is a cultural practice, where heated oil is applied to the skin followed by repeated rubbing of the skin with a coin, or other

object, to produce petechiae or ecchymoses. It is a common practice in Southeast Asia to improve circulation or to relieve common symptoms including fever and headache.

Cupping is another remedy that is believed to aid in symptoms such as abdominal pain, fever, and poor appetite. Heated cups create a vacuum on the skin producing characteristically shaped ecchymoses. The technique is used by various cultural groups including Russians, Asians, and Mexicans.

Gridding is a remedy that is most commonly practiced in Russian cultures as well as Ukraine and other eastern European countries. Gridding is the practice of painting the back with iodine in a crisscross pattern. This results in a hyperpigmented grid-like pattern on the back. It is used to treat respiratory illness because the topical application of iodine is thought to relieve cough and congestion. All of these lesions can be mistaken for physical abuse.

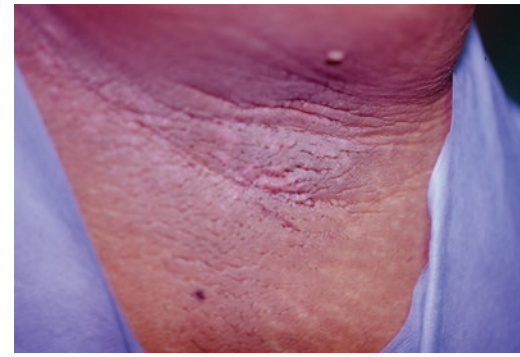
Acupressure and acupuncture generally do not leave cutaneous signs of treatment. The presence of petechiae or ecchymoses should raise the question of nonaccidental trauma, especially in the pediatric population. Increased cultural competency is required to understand the mechanism of these cutaneous findings.

Take-Home Message
Complementary alternative medicine practices can result in cutaneous findings such as circular bruising and linear purpuric streaks that can be mistaken for nonaccidental trauma.

Question 23

A 13-year-old girl is brought to the emergency department for evaluation of fatigue, weakness, and recurrent episodes of vomiting and diarrhea. On examination, you note hyperpigmentation, most prominently on the palms, soles, and skin folds. Her skin is notably darker than her mother's. Her findings are consistent with:

A. Acanthosis nigricans
B. Addison's disease
C. McCune–Albright syndrome
D. Classic congenital adrenal hyperplasia
E. Polycystic ovary syndrome



Acanthosis nigricans

Correct Answer: B

This child has findings consistent with Addison's disease, primary adrenocortical insufficiency. Acute Addisonian crisis is one of the most severe endocrine emergencies manifesting during episodes of acute stress, infection, or trauma. It is characterized by hypovolemia, hypotension, and acute cardiovascular collapse due to renal sodium wasting, hyperkalemia, and loss of vascular tone. An increased pigmentation of the skin and mucous membranes is also present. In addition, increased pigmentation may be noted in existing nevi. Hyperpigmentation occurs due to increased levels of proopiomelanocortin, a precursor to melanocyte-stimulating hormone. As the pigmentation may be subtle, comparison of the patient to other family members may be useful in highlighting the skin findings. Decreased aldosterone production leads to hyponatremia and hyperkalemia. There is also an increased renin level. The diagnostic criteria include low cortisol level and failure to rise with ACTH stimulation. Treatment includes immediate restoration of the intravascular volume with administration of intravenous saline as well as dexamethasone or hydrocortisone.

Acanthosis nigricans (AN) is characterized by hyperpigmentation and hyperkeratosis in intertriginous areas, most commonly the neck and

axillae. It usually manifests at puberty but can occur in childhood with early development of obesity. AN is a cutaneous marker for metabolic disturbance, most commonly insulin resistance.

The triad of McCune–Albright syndrome (MAS) consists of fibrous dysplasia of the bones (polyostotic fibrous dysplasia), patchy cutaneous pigmentation, and endocrinopathies. Precocious puberty is the most common endocrinopathy; other associated disorders include pituitary adenomas (secreting growth hormone), hyperthyroid goiters, and adrenal hyperplasia. In MAS, the café au lait macules have irregular borders, described as the "coast of Maine," and present early in life.

Females with classic congenital adrenal hyperplasia (CAH) due to 21-hydroxylase deficiency present with ambiguous genitalia at birth. Boys generally have no obvious physical signs; hyperpigmentation and/or penile enlargement may be present. Precocious puberty may occur in patients not on appropriate treatment.

Polycystic ovary syndrome presents with hyperandrogenism, irregular menses, and polycystic ovaries. Cutaneous signs of hyperandrogenism include acne vulgaris, hirsutism, and androgenetic alopecia.

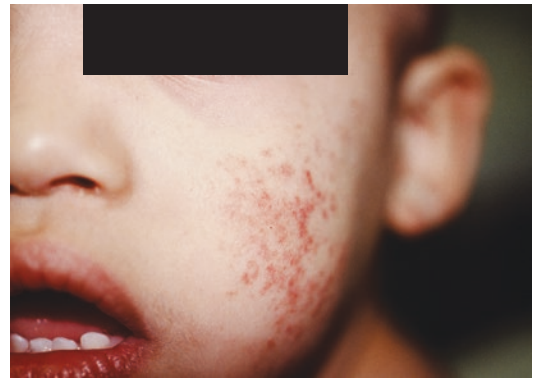
Take-Home Message
Addison disease is a potentially life-threatening condition. Hyperpigmentation is present in nearly all patients with Addison disease.

Question 24

A 5-year-old boy presents with a history of coughing for 4 days. He is well appearing and has a fine petechial rash around the eyes and on the cheeks. Which of the following is true?

A. The rash is most likely due to abnormal platelets.
B. He most likely has a normal platelet count.
C. The petechiae are unrelated to coughing.
D. He should be referred to the ophthalmologist.
E. He should be hospitalized for observation.

Correct Answer: B



Purpura – Valsalva

Petechiae are non-blanching less than 3 mm. The presence of petechiae does not always indicate serious illness. Prolonged crying, forceful coughing, and vomiting increase intravascular pressure and can result in endothelial damage. This can also result from strenuous activities or exercise. The petechiae usually localize to the face, upper chest, or arms (usually above the nipple line).

Idiopathic thrombocytopenic purpura following a viral illness can occur in an otherwise healthy and well-appearing child; however, the petechiae do not localize only to the upper body.

Petechiae caused by infection usually involve gram negative organisms, especially *Neisseria meningitidis*, and rickettsiae. A child presenting with a petechial rash and fever should be evaluated promptly for such life-threatening illnesses. With the appropriate history, certain viral illnesses such as dengue fever, Ebola, and yellow fever should be considered. Other causes of generalized petechiae include malignancies such as leukemia and other that cause thrombocytopenia, medications, radiation, autoimmune disorders, and vasculitis.

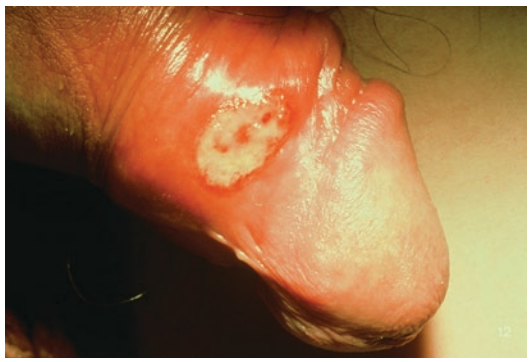
Take-Home Message
Increased intravascular pressure can cause petechiae. The presence of a petechial rash in a febrile child should raise the suspicion of meningococcal infection.

Question 25

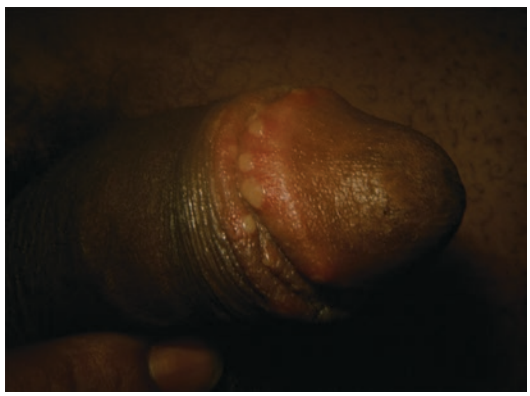
Which of the following associations is INCORRECT?

A. Painless genital chancre: herpes simplex
B. Erythema migrans: *Borrelia burgdorferi*
C. Pastia's lines: scarlet fever
D. Slapped cheeks: erythema infectiosum
E. Blueberry muffin: congenital rubella

Correct Answer A



Chancre (primary syphilis)



Primary herpes simplex

Genital ulcers have a broad differential diagnosis including infectious and inflammatory etiologies. Syphilis is a systemic, sexually transmitted disease caused by the spirochete *Treponema pallidum*. Primary syphilis is manifested as a single, painless ulcer called "chancre," which develops 0–90 days, on average 3 weeks, following exposure. It begins as a small red papule or a crusted superficial erosion.

The chancre can be found most commonly on the penis, vulva, anus, or cervix and is associated with regional lymphadenopathy. The treatment of choice at this stage is a single, intramuscular dose of benzathine penicillin, 2.4 million units.

Herpes simplex infection is characterized by grouped vesicles on an erythematous base. Genital herpes is most commonly caused by herpes simplex virus type-2 (HSV-2). Primary infection is usually painful and symptomatic, with associated systemic symptoms such as tender regional lymphadenopathy, fever, headache, and malaise. Recurrent episodes can have a prodrome of itching or pain, or be asymptomatic. When vesicles rupture, characteristic punched-out and scalloped ulcerations remain. HSV PCR can be performed on the base of the ulcer to confirm the diagnosis if needed.

Lyme disease is caused by the spirochete *Borrelia burgdorferi* and is the most common vector-borne disease in North America. The initial cutaneous manifestation of Lyme disease is a solitary erythematous patch called erythema chronicum migrans. The erythema spreads centrifugally after originating at the site of the bite of the Ixodes tick. The average interval between the tick bite and the appearance of the skin lesion is approximately 1–2 weeks. Erythema migrans does not involve the mucous membranes, palms, or soles. Diagnosis is based on history of exposure to ticks in an endemic area, clinical presentation, and serology. However, many patients with documented Lyme disease may not report a tick bite.

Scarlet fever is caused by infection with group A β-hemolytic streptococcus, which produces streptococcal pyrogenic exotoxins (SPEs). The exanthem of scarlet fever presents with small, erythematous

papules and has been described as “sandpapery”; the erythema blanches with pressure. It involves the trunk and extremities and may be accentuated in skin folds. The petechiae may appear in a linear distribution along the creases, forming Pastia lines. The skin may be pruritic, but it is not tender. Examination of the oropharynx may reveal exudative tonsils and palatal petechiae.

Erythema infectiosum is caused by human parvovirus B19. The rash begins on the face with a bright red, nontender rash most prominent on the cheeks, with circumoral pallor producing the classic “slapped-face” appearance. The rash then spreads to the trunk and proximal extremities. Central clearing of lesions gives the rash a lacy, reticulated appearance. This tends to be more prominent on extensor surfaces, sparing the palms and soles. Mild prodromal symptoms may occur a week prior to rash onset. The patients are afebrile and do not appear ill.

Congenital rubella can present with a characteristic, bluish, papular eruption, termed a “blueberry muffin” rash, which represents dermal hematopoiesis. Other associated anomalies include cataracts, deafness, cardiac defects, and CNS abnormalities. The fetus is most susceptible to infection in the first 16 weeks of gestation.

Take-Home Message

A painless genital ulcer, known as a chancre, is the characteristic lesion of primary syphilis. Herpetic ulcers are characterized by a scalloped border. Typical herpetic lesions present as a cluster of small erythematous painful vesicles, which quickly ulcerate.