

Northwestern University Chicago Dermatological Society

Maintenance of Certification Questions

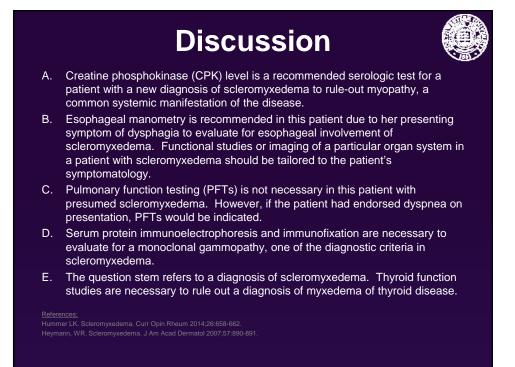
November 9th, 2016

1. A 40-year-old female presents with 2 mm coalescing papules on her hands, neck, glabella and post-auricular areas. She also complains of dysphagia. Biopsy of a representative lesion shows mucin deposition, increased spindle-like fibroblast proliferation, and fibrosis. Which of the following is not part of the recommended work-up of this condition?



- A. Creatine phosphokinase level
- B. Esophageal manometry
- C. Pulmonary function testing
- D. Serum protein immunoelectrophoresis and immunofixation
- E. Thyroid function studies

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- 2. A 50-year-old male presents with new waxy, closely spaced, skin-colored, 3 mm papules involving the glabella and pre-auricular areas. Biopsy and laboratory studies are consistent with scleromyxedema. Which of the following is NOT a serological or histopathological feature consistent with a diagnosis of scleromyxedema?
 - A. Fibrosis and fibroblast proliferation on biopsy
 - B. IgG paraproteinemia
 - C. Mucin deposition on biopsy
 - D. Normal thyroid function testing
 - E. Presence of Anti-DNA topoisomerase I antibody

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- A. Diagnostic histopathological features of scleromyxedema are mucin deposition, fibrosis, and fibroblast proliferation.
- B. Monoclonal gammopathy is a criterion for diagnosis of scleromyxedema. The most commonly seen paraproteinemia in scleromyxedema patients is IgG with prevalence of lambda light chains.
- C. Diagnostic histopathological features of scleromyxedema are mucin deposition, fibrosis, and fibroblast proliferation.
- D. Normal thyroid function testing is a criterion for diagnosis of scleromyxedema as it rules out myxedema associated with thyroid dysfunction.
- E. Presence of Anti-DNA topoisomerase I antibody is not found in scleromyxedema patients. It is found in patients with diffuse cutaneous systemic sclerosis and indicates a high risk of interstitial lung disease.

References: Hummer LK. Scleromyxedema. Curr Opin Rheum 2014;26:658-662. Heymann, WR. Scleromyxedema. J Am Acad Dermatol 2007;57:890-891.

3. A 3-month-old African American female presents with a 12 cm dark brown uniform plaque on her scalp that has been present since birth. Which statement is correct?



- A. On histopathology, the nevomelanocytes do not extend beyond the epidermis and superficial dermis.
- B. The most common mutation is a somatic mutation in the heterotrimeric G protein alpha subunit, GNAQ.
- C. There is approximately a 5% lifetime risk that this patient may develop melanoma.
- D. This lesion can be classified as a medium-sized congenital melanocytic nevus.
- E. This lesion is most commonly seen in individuals with Fitzpatrick skin type 1.

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- 4. A 3-year-old African American male with a history of developmental delay, seizures and hydrocephalus s/p shunt placement presented to the office because his parents were concerned about a large, dark brown "birthmark" on his back. Which statement is correct regarding his condition?
 - A. Neurological symptoms in symptomatic individuals commonly develop before 6 months of age.
 - B. On physical exam, most patients with this condition are likely to have numerous proliferative nodules within a congenital melanocytic nevus.
 - C. On physical exam, most patients with this condition are likely to have a giant congenital melanocytic nevus in a posterior axial location.
 - D. The preferred modality for diagnosis of this condition is a PET-CT scan.
 - E. There is a very strong correlation between evidence of this condition on imaging and neurological symptoms.

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- A. While proliferative nodules are seen within CMN, they are not associated with NCM (option A is false).
- B. The preferred imaging modality is an MRI with gadolinium contrast (option 2 is false).
- C. The patient in this case is suffering from neurocutaneous melanosis (NCM), a rare complication associated most commonly with large congenital melanocytic nevi (CMN). Those at highest risk have CMN larger than 40 cm in their final size in a posterior axial location, multiple satellite nevi and/or numerous medium-sized CMN (option C is correct).
- D. Clinical manifestations are usually seen at a median age of two years (option D is false).
- E. Limited evidence suggests that only approximately 1-10% of patients with evidence of NCM on imaging develop neurological symptoms (option E is false).

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neurocutaneous melanosis in neurologically asymptomatic children. Plast Reconstr Surg. 2001;107(4):933.
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- 5. A 60-year-old male with HIV/AIDS is re-admitted to the hospital for altered mental status. He was discharged three weeks previously for diverticulitis, which was treated with intravenous antibiotics and supportive care. Four months prior, he spent one month in Southern Africa on safari; while there, he encountered cows, goats, and sheep. His cutaneous examination was notable for several scattered verrucous and ulceronecrotic plaques. PCR performed on tissue biopsy revealed *Acanthamoeba* spp., and the patient was diagnosed with disseminated acanthamoebiasis complicated by granulomatous amoebic encephalitis. Which of the following most likely predisposed this patient to developing this infection?

- A. Exposure to livestock
- B. Immunosuppressed status
- C. Recent antibiotic exposure
- D. Recent hospitalization
- E. Travel to Africa

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B. Acanthamoeba spp. is a free-living amoeba that is found worldwide and is ubiquitous in water and soil. Similar to other opportunistic infections, Acanthamoeba spp. requires an immunosupressed host in order to cause disseminated acanthamoebiasis and granulomatous amoebic encephalitis. The amoebic trophozoites establish infection after gaining access to the bloodstream via direct inoculation into skin or via inhalation.

A, C, D, E: Though the patient's recent medical and social history may be relevant to other infections that could be included in the initial differential diagnosis, these factors would not alone predispose an individual to developing disseminated acanthamoebiasis or granulomatous amoebic encephalitis.

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- 6. A 2-day-old female was born with diffuse 2-5mm firm dome-shaped blue-red papules of the scalp, chest, abdomen, and extremities. The infant was also small for gestational age, and physical exam was notable for deafness and hepatosplenomegaly. CT shows periventricular calcifications. The skin lesions are observed to fade into light brown macules over the next several days. If one of the skin lesions were biopsied, what would the histology most likely show?
 - A. CD1a and S100 positive staining cells
 - B. Clonal proliferation of immature cells of neural crest origin
 - C. Dilated vessels in the dermis with thrombosis and endothelium hyperplasia
 - D. Hematopoietic cells in the dermis
 - E. Nuclear and cytoplasmic amphophilic inclusions with chromatin at the nuclear perimeter

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- A. Immunostaining for CD1a and S100 is considered diagnostic in Langerhans cell histiocytosis.
- B. This answer choice describes cutaneous neuroblastoma which is diagnosed on biopsy and with elevated urine VMA (vanillylmandelic acid) and HVA (homovanillic acid).
- C. This answer choice describes the findings seen with multifocal lymphoangioendotheliomatosis, which is not associated with intrauterine growth restriction, periventricular calcifications and sensorineural hearing loss.
- D. This is the correct answer. Dermal extramedullary hematopoiesis is normal during gestation but is persistent in various pathologic states that occur *in utero*. Postnatal extramedullary hematopoiesis is associated with congenital toxoplasmosis, rubella, and CMV, the entity described in this question stem.
- E. The question stem describes a "blueberry muffin baby" caused by congenital CMV infection. Although the classic CMV inclusions do show amphophilic staining and stain positively for CCH2/AD169, the cutaneous lesions of congenital CMV are due to extramedullary hematopoiesis and are not due to direct viral cytopathic change.

References:

Paller AS, Mancini AJ. Hurwitz Clinical Pediatric Dermatology: A Textbook of Skin Disorders of Childhood and Adolescence (4th Ed.). Philadelphia: Elsevier Saunders; 2011.

7. A 52-year-old female with a history of fasting hyperglycemia and an unprovoked pulmonary embolism presents for evaluation of a one-year history of a waxing and waning cutaneous eruption, unresponsive to treatment with topical steroids, vitamin D analogues, and systemic methotrexate. On physical exam, she is noted to have symmetric erythematous, eroded and crusted plaques in the perineum, vulva and inguinal creases. Skin biopsy reveals epidermal pallor, spongiosis, necrotic keratinoctyes, and a perivascular infiltrate of lymphocytes. Which of the following is the next best step in management?



- A. Check ELISA for IgG antibodies to desmoglein 1 and desmoglein 3
- B. Check HIV serologies
- C. Perform a CT scan of the abdomen and pelvis
- D. Perform MRI of the brain to evaluate for metastatic disease
- E. Stop methotrexate and start systemic cyclosporine

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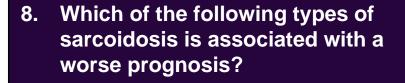




- A. Necrolytic migratory erythema (NME) may present similarly to pemphigus vulgaris with eroded patches and plaques and mucous membrane involvement. NME is not an autoimmune bullous disorder and does not have an association with autoantibodies to desmogleins. For NME, relevant labs include complete blood count, serum glucose, glucagon, zinc and fatty acid levels.
- B. NME may be mistaken for refractory seborrheic dermatitis. Seborrheic dermatitis can be seen with increased frequency and severity in association with HIV.
- C. CT scan of the abdomen and pelvis in order to evaluate for a pancreatic tumor causing glucagonoma syndrome is the appropriate next step once the diagnosis of NME is made.
- D. Approximately 70% of glucagonomas are already metastatic at the time of diagnosis, most commonly to the liver. Metastases to the brain are rare and thus MRI of the brain is not the next best step in evaluation.
- E. Though NME often mimics atopic dermatitis clinically, the biopsy findings of epidermal pallor and necrotic keratinocytes do not support the diagnosis of atopic dermatitis, and thus treatment with cyclosporine would be inappropriate.

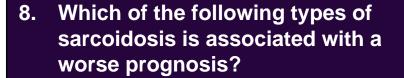
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- B. Ichthyosiform
- C. Lupus pernio
- D. Morpheaform
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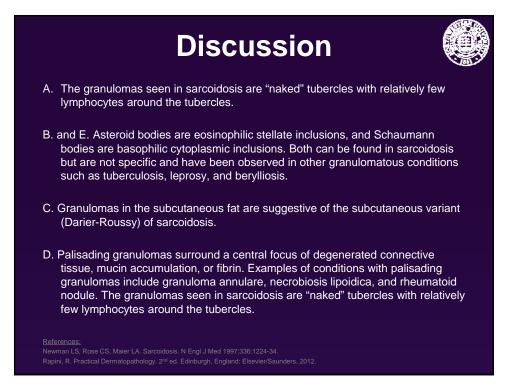
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9. Which of the following histological findings are NOT seen in sarcoidosis:

- A. Epithelioid histiocytes forming "naked tubercles" with minimal lymphocytic infiltrate
- B. Giant cells containing eosinophilic stellate inclusions representing engulfed collagen
- C. Granulomas in the subcutaneous fat
- D. Palisading histiocytes surrounding a central focus of degenerated connective tissue
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- 10. A 1-year-old girl is brought for evaluation of a red to brown papule on the back that has gradually increased in size to a plaque. A biopsy of the lesion would later show an increased number of mast cells. What clinical exam finding should be elicited?
 - A. Applying inward pressure from the border of the lesion induces a dimple formation
 - B. Applying pressure at lesion center causes invagination, which is restored upon pressure release
 - C. Applying tangential pressure at the edge of the lesion causes skin peeling
 - D. Rubbing of the lesion causes redness and edema of the lesion
 - E. Stroking of normal skin causes redness and edema of normal skin

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- A. This describes the dimple sign, which is positive in dermatofibromas.
- B. This describes the buttonhole sign, which is positive in neurofibromas and anetoderma.
- C. This describes the Nikolsky sign, which is positive in intraepidermal blistering disorders like pemphigus vulgaris.
- D. This describes the Darier sign, which is positive in almost all childhood-onset and most adult-onset mastocytosis.
- E. This describes dermatographism, in which physical urticaria is induced in otherwise normal skin.

References: Hartmann K, Escribano L, Grattan C, Brockow K, Carter MC, Alvarez-Twose I, et al. Cutaneous manifestations in patients with mastocytosis: Consensus report of the European Competence Network on Mastocytosis; the American Academy of Allergy, Asthma & Immunology; and the European Academy of Allergology and Clinical Immunology. J Allergy Clin Immunol. 2016;137(1):35-45. Paller AS, Mancini AJ. Hurwitz Clinical Pediatric Dermatology: A Textbook of Skin Disorders of Childhood and Adolescence (5th Ed.). Philadelphia: Elsevier; 2016.

11. A 1-month-old boy is brought for evaluation of numerous scattered red to brown macules, papules, and plaques. A few of the raised lesions have a *peau d'orange* appearance and Darier sign is positive. Which of the following is correct regarding this condition?



- A. It will most likely persist beyond puberty
- B. Histopathology will reveal increased numbers of mast cells
- C. This patient has a germline mutation in the KIT gene
- D. The patient is at high risk for anaphylaxis
- E. A bone marrow biopsy should be ordered

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- A. The vast majority of mastocytosis cases in children are cutaneous and not systemic; therefore a bone marrow biopsy is not normally warranted. Choice E is incorrect.
- B. This child has clinical signs of cutaneous mastocytosis. A lesional skin biopsy would show increased numbers of mast cells, most easily seen with a tryptase antibody stain.
- C. Cutaneous mastocytosis in children most commonly spontaneously resolves around puberty. Choice A is incorrect.
- D. While most children and adults with mastocytosis have an activating mutation in the *KIT* gene, about 25% of children with mastocytosis have no detectable *KIT* gene mutations. Choice C is incorrect as the mutation is not germline.
- E. Childhood-onset mastocytosis has a low risk of anaphylaxis (<10%) compared to adult-onset (50%). Choice D is correct.

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