MOC Self-Assessment Questions

MAY 2015 CHICAGO DERMATOLOGICAL SOCIETY CONFERENCE

HOST – RUSH UNIVERSITY

1. Dysregulation of the metabolism of which of the following should be considered in the management of a patient with porphyria cutanea tarda?

1. Copper
2. Calcium
3. **Iron**
4. Magnesium
5. Vitamin E

1.

Explanation: Understanding the oxidative role iron plays in decreasing the function of heme synthesis underlies key principles in the treatment of porphyria cutanea tarda. Especially in the setting of iron overload, phlebotomy is the treatment of choice with a goal of maintaining ferritin levels below 20.

References:

Caballes FR, Sendi H, Bonkovsky HL. Hepatitis C, porphyria cutanea tarda and liver iron: an update. Liver Int. 2012 Jul;32(6):880-93.

2. A 57-year-old man with a history of chronic hepatitis C infection develops non-inflamed blisters and erosions on the dorsal hands associated with milia formation. A biopsy from lesional skin demonstrated a non-inflammatory sub-epidermal blister, consistent with porphyria cutanea tarda. Based on this information, treatment was initiated with hydroxychloroquine 200 mg po daily. After initiating treatment, the patient called complaining of right upper quadrant pain and fever. Laboratory analysis demonstrated significant elevation of AST and ALT. Which of the following provides the best explanation for the patient's presentation after initiation of hydroxychloroquine?

1. Mobilization of hepatic iron stores
2. **Too rapid release of stored hepatic porphyrins**
3. Acute biliary obstruction
4. Hydroxychloroquine hypersensitivity reaction
5. Reactivation of hepatitis C virus

2.

Explanation: Aside from phlebotomy, treatment with antimalarials has been shown to reduce cutaneous

manifestations of porphyria cutanea tarda by decreasing iron stores of toxic intermediate porphyrins

formed in the heme synthesis pathway. It is critical that these drugs be given at lower doses than might be

used for other indications, as too rapid release of porphyrin intermediates may cause an acute hepatitis. It

is recommended that hydroxychloroquine be given at 200 mg twice weekly.

References:

Caballes FR, Sendi H, Bonkovsky HL. Hepatitis C, porphyria cutanea tarda and liver iron: an update. Liver Int. 2012 Jul;32(6):880-93.

3. Which features are most suggestive of a diagnosis of alopecia areata on dermoscopic evaluation for hair loss?

1. Fibrotic white dots and absent hair shafts
2. Hair shaft thickness heterogeneity and an increased proportion of vellus hairs
3. Chaotic arrangement of multiple broken hair shafts
4. **Yellow dots and broken tapered hair shafts**
5. Large yellow-brown dots and peripheral thick arborizing vessels

3.

Explanation: Trichoscopy can be a powerful tool when differentiating etiologies for alopecia. Alopecia areata demonstrates yellow dots and characteristic broken tapered hair shafts, known as "exclamation mark hairs," as a result of the cessation of mitotic activity in the hair shaft. Answer choice A describes cicatricial alopecia. Answer choice B describes androgenetic alopecia. Answer choice C describes trichotillomania. Answer choice E describes the follicular plugging seen in discoid lupus erythematosus lesions.

References:

Mubki T, Rudnicka L, Olszewska M, Shapiro J. Evaluation and diagnosis of the hair loss patient: Part II; trichoscopic and laboratory evaluations. J Am Acad Dermatol. 2014 Sep;71(3):431.e1-431.e11.

4. The pathogenesis of alopecia areata is most likely directly mediated by which cell type?

1. Neutrophil
2. Mast cell
3. Macrophage
4. CD4+ T cell
5. **CD8+ T cell**

4.

Explanation: As new therapeutic agents are developed for this challenging disease, it is important to understand that its pathogenesis is primarily mediated by effector cytotoxic T cells as they stage an attack on follicle-resident melanocytes.

References:

Madke B, Doshi B, Khopkar U, Dongre A. Appearances in dermatopathology: The diagnostic and the deceptive. Indian J Dermatol Venereol Leprol. 2013 May-Jun;79(3):338-48.

5. Which of the following is the most characteristic on histopathology of sclerosing lipogranuloma?

1. Touton giant cells
2. **Variably sized cystic vacuoles**
3. Stellate fibroblasts
4. Blue/black rod-shaped organisms in the cytoplasm of phagocytes
5. Fibrotic plaques of the tunica albuginea

5.

Explanation: As sclerosing lipogranuloma represents a foreign body reaction to an oil-based substance, the expected granulomatous reaction is punctated by oil-containing vacuoles of varying sizes, giving the so-called "Swiss cheese" appearance of lesions histologically. Foreign-body giant cells may be seen. As the response is not fibrotic, an increase in fibroblasts is not expected.

References:

Madke B, Doshi B, Khopkar U, Dongre A. Appearances in dermatopathology: The diagnostic and the deceptive. Indian J Dermatol Venereol Leprol. 2013 May-Jun;79(3):338-48.

6. Which of the following is the most commonly observed clinical feature of sclerosing lipogranuloma of the penis?

1. Erythema
2. Phimosis
3. Fistulas
4. **Ulceration**
5. Malodorous drainage

6.

Explanation: Understanding the clinical presentation of a sclerosing lipogranuloma will lead to earlier suspicion for the diagnosis, earlier biopsy of affected skin, and more rapid management. Although lesions may be erythematous, even skin-colored lesions of sclerosing lipogranuloma are likely to demonstrate ulceration. Fistulas may be seen in a smaller portion of patients.

References:

Pehlivanov G, et al. Foreign?body granuloma of the penis in sexually active individuals (penile paraffinoma). J Euro Acad of Dermatol Venereol. 2008 Jul;22(7):845-51.

7. Primary infection with blastomycosis is most often contracted via

1. Inhalation of aerosolized pigeon droppings
2. Direct cutaneous inoculation via canine bite
3. **Inhalation of spores from moist soil**
4. Oral consumption of minute quantities of guano
5. Direct cutaneous inoculation from puncturing trauma

7.

Explanation: Blastomycosis is endemic to the Mississippi and Ohio river valleys, Great Lakes region and the southeastern united states. The lungs are typically the first site of infection, via inhalation of spores in moist soil. Primary cutaneous inoculation of blastomycosis is extremely unusual. Cryptococcus may be contracted from bird excreta. Histoplasmosis may be contracted via inhalation of organisms, which may arise from bird, fowl and bat droppings.

References:

Klein BS, et al. Isolation of Blastomyces dermatitidis in soil associated with a large outbreak of blastomycosis in Wisconsin. N Engl J Med. 1986 Feb 27;314(9):529-34

8. Appropriate treatment for immunosuppressed patients with blastomycosis typically involves

1. Amphotericin
2. Efinaconazole
3. Itraconazole
4. **Amphotericin + Itraconazole**
5. Surgical debridement

8.

Explanation: Blastomycosis requires systemic antifungal treatment. While mild to moderate non-CNS disease may be treated alone with itraconazole, severe or progressive disease should be treated with amphotericin B. In the immunocompromised population, a combination of amphotericin B and itraconazole is recommended.

References:

Chapman SW, et al. Clinical practice guidelines for the management of blastomycosis: 2008 update by the Infectious Diseases Society of America. Clin Infect Dis, 2008 Jun 15;46(12):1801-12.

9. Which histopathologic features are most suggestive of a diagnosis of interstitial granulomatous drug reaction?

1. **Piecemeal fragmentation of collagen and elastic fibers**
2. "Top heavy" infiltrate and abundant mucin
3. Granuloma with tissue eosinophilia
4. Acute necrosis of collagen fibers surrounded by histiocytes
5. Evidence of vasculitis

9.

Explanation: Interstitial granulomatous drug reaction must be differentiated from interstitial granulomatous dermatitis and from granuloma annulare. Interstitial granulomatous dermatitis and drug reaction both involve a "bottom heavy" interstitial infiltrate of histiocytes and neutrophils and lack dermal mucin. Interstitial granulomatous drug reaction may be distinguished by a vacuolar interface dermatitis and prominent eosinophils. Granuloma annulare is often a "top heavy" dermal process accompanied by abundant mucin deposition.

References:

Magro CM, et al. The interstitial granulomatous drug reaction: a distinctive clinical and pathological entity. J Cutan Pathol. 1998 Feb;25(2):72-8.

10. Which of the following is most likely to be decreased in a patient with zinc deficiency?

1. Hemoglobin
2. **Alkaline Phosphatase**
3. Erythrocyte Sedimentation Rate
4. Albumin
5. C-Reactive Protein

10.

Explanation: Rapid identification of zinc deficiency is critical to allow for repletion. While the zinc level may be low, in some cases, it may be low-normal or even normal. Measuring alkaline phosphatase levels may serve as a surrogate marker for zinc status, as it requires zinc as a co-factor.

References:

Lott JP, Reeve J, Ko C, Girardi M. Periorificial dermatitis and erosive inguinal plaques in a 57-year-old woman. Acquired zinc deficiency acrodermatitis enteropathica (ADE). JAMA Dermatol. 2013 Mar; 149(3):357-63.

11. Of the following, which is the most likely to contribute to acquired zinc deficiency?

1. Non-Hodgkin lymphoma
2. **Gastric bypass surgery**
3. Chronic hepatitis C
4. Internal malignancy
5. Copper supplementation

11.

Explanation: Acquired zinc deficiency results from nutritional zinc deficiency secondary to poor dietary intake or malabsorption and can be seen in a variety of conditions, including alcoholism, anorexia, gastrointestinal disorders (eg, inflammatory bowel disease, celiac sprue, pancreatic insufficiency, and bowel resection), adherence to fad diets high in phytates, and receipt of zinc-deficient parenteral nutrition. High zinc intake may lead to decreased copper absorption.

References:

Lott JP, Reeve J, Ko C, Girardi M. Periorificial dermatitis and erosive inguinal plaques in a 57-year-old woman. Acquired zinc deficiency acrodermatitis enteropathica (ADE). JAMA Dermatol. 2013 Mar; 149(3):357-63.

12. Hair loss may be seen in approximately what proportion of patients with secondary syphilis?

1. **5-10%**
2. 20-25%
3. 45-50%
4. 65-70%
5. >75%

12.

Explanation: Although representing a small portion of patients, recognizing moth eaten alopecia as a sign for secondary syphilis is important as it may be the presenting sign of syphilis.

References:

Mubki T, Rudnicka L, Olszewska M, Shapiro J. Evaluation and diagnosis of the hair loss patient: Part II; trichoscopic and laboratory evaluations. J Am Acad Dermatol. 2014 Sep;71(3):431.e1-431.e11

13. Which presentation is most consistent with alopecia associated with systemic lupus erythematosus?

1. Exclamation mark hairs, black dots, lack of erythema
2. Linear band of scarring hair loss across frontal hair line
3. **Dyspigmentation, telangiectasias, incomplete hair loss in patches**
4. Pustulosis with scarring and tufted follicles
5. Diffuse thinning across the scalp with retained hair lines

13.

Explanation: Alopecia remains a part of the diagnostic criteria for systemic lupus erythematosus. Dyspigmentation, telangiectasias and incomplete hair loss in patches in combination should raise suspicion for underlying lupus. Answer choice A describes alopecia areata. Answer choice B describes frontal fibrosing alopecia, a presentation of lichen planopilaris. Answer choice D describes folliculitis decalvans. Answer choice E describes telogen effluvium.

References:

Ye Y, Zhao Y, Gong Y, et al. Non-scarring patchy alopecia in patients with systemic lupus erythematosus differs from that of alopecia areata. Lupus. 2013 Dec;22(14):1439-45.

14. A 42-year-old HIV positive man presented with a painful erythematous pustule on the back with surrounding erythema and induration. A culture of the contents from the pustule revealed a coagulase-negative staphylococcus later identified as Staphylococcus lugdunensis. Which of the following is the most reasonable next step?

1. Consider the result a contaminant
2. Consider the result demonstrative of normal skin flora
3. Prescribe mupirocin 2% ointment
4. **Prescribe an oral antibiotic based on sensitivity results**
5. Reculture the lesion

14.

Explanation: Aerobic culture results of coagulase-negative staphylococcus are often interpreted as reflective of commensal organisms and overlooked as possible causative agents of cutaneous infection.

References:

Donoghue S, Vekic D, Wehrhahn M, Whitfield M. Staphylococcus lugdunensis: case report and discussion. Australas J Dermatol. 2014 Nov;55(4):301-3.

15. Although blastomycosis-like pyoderma is often caused by more than one bacterial species, which is the most commonly isolated species?

1. Escherichia coli
2. Pseudomonas aeruginosa
3. Streptococcus pyogenes
4. Candida albicans
5. **Staphylococcus aureus**

15.

Explanation: Blastomycosis-like pyoderma may present with large verrucous plaques with or without pustules. Bacteria such as S. aureus, P. aeruginosa, Proteus, E. coli, or streptococci may be isolated; however, S. aureus is the most commonly isolated. Antibiotics appropriate for the organism isolated are curative; however, response may be delayed and prolonged therapy required.

References:

Lee YS, Jung SW, Sim HS, et al. Blastomycosis-like pyoderma with good response to acitretin. Ann Dermatol. 2011 Aug;23(3):365-8.

16. In addition to the association with cystic fibrosis, aquagenic wrinkling of the palms has been reported in patients taking which class of medication?

1. Antihistamines
2. SSRIs
3. **NSAIDs**
4. Opioids
5. Calcium channel blockers

16.

Explanation: To avoid unnecessary genetic testing, understanding that NSAIDs have been associated with aquagenic wrinkling is important.

References:

Arkin L, Flory J, et al. Pediatr Dermatol. 2012 Sep-Oct;29(5):560-6.

17. A punch biopsy obtained from a patient with a diffuse eruption of 2-3 mm firm skin-colored papules reveals deposition of mucin in the upper and mid dermis with increased collagen deposition. Which lab test will most likely be abnormal in this patient?

1. Hemoglobin
2. Liver function panel
3. C-reactive protein
4. Creatinine
5. **Immunofixation electrophoresis**

17.

Explanation: Increased dermal fibroblasts with increased collagen deposition and mucin deposition are seen in scleromyxedma and nephrogenic systemic fibrosis. Scleromyxedema may occur in the setting of monoclonal gammopathy, which would be evident on an immunofixation electrophoresis.References:

Applebaum D, Daulat S, Duvic M. JAMA Dermatol. 2014;150(8):893-894.

18. Which of the following should be considered in the initial work up of a patient with MDA-5 positive dermatomyositis?

1. Colonoscopy
2. Chest plain films
3. Mammography
4. **Pulmonary function testing**
5. Papanicolaou smear

18.

Explanation: Anti-MDA-5 (CADM-140) antibodies in dermatomyositis have been associated with increased risk for interstitial lung disease. The best tests to assess for lung involvement in these patients are CT chest and pulmonary function testing. Chest plain films are not sensitive enough to be used in this capacity.References:

Koichi Y, Aya Y, Megumi U, et al. A case of anti-MDA5-positive rapidly progressive interstitial lung disease in a patient with clinically amyopathic dermatomyositis ameliorated by rituximab, in addition to standard immunosuppressive treatment. Mod Rheumatol. 2015 Mar 12:1-5.

19. A 34-year-old woman newly diagnosed with dermatomyositis demonstrates positive titers against TIF1-ʔ. Which of the following tests is the most appropriate to order in this patient?

1. Liver function tests
2. **CA-125 level**
3. Calcium
4. Thyroid function tests
5. HbA1C

19.

Explanation: Anti-TIF1-γ (p155) antibodies in dermatomyositis have been associated with increased risk for an associated internal malignancy. In women with dermatomyositis, the most commonly associated internal malignancy has been ovarian cancer; therefore, measuring CA-125 levels in this patient would be prudent.References:

Valenzuela A, Chung L, Casciola-Rosen L, Fiorentino D. Identification of clinical features and autoantibodies associated with calcinosis in dermatomyositis. JAMA Dermatol. 2014 Jul;150(7):724-9.

20. Which variant of morphea has been associated with the most frequent disease recurrence?

1. Guttate
2. Plaque
3. **Linear**
4. Profunda
5. Generalized

20.

Explanation: Even after adequate treatment has led to remission, morphea may recur, requiring re-initiation of treatment. Linear morphea has been shown to be the subtype most likely to recur, suggesting that regular follow up even after adequate treatment and remission is warranted.

References:

Mertens JS, Syger MM, Kievit W, et al. Disease recurrence in localized scleroderma: a retrospective analysis of 344 patients with paediatric- or adult-onset disease. Br J Dermatol. 2015 Mar;172(3):722-8.

21. Which of the following antibodies is most commonly associated with an overlap between dermatomyositis and scleroderma?

1. **Anti-Ku**
2. Anti-Jo1
3. Anti-CADM140
4. Anti-PL-7
5. Anti-p155

21.

Explanation: Anti-Ku and anti-U1RNP antibodies may be seen in patients with dermatomyositis who have features overlapping with another connective tissue disease. Specifically, anti-Ku antibodies have been associated with an overlap between dermatomyositis and systemic sclerosis. Anti-Jo1 and anti-PL-7 antibodies are both seen with the anti-synthetase syndrome. Anti-p155 (TIF1-γ) antibodies are associated with internal malignancy.

References:

Suzuki S, Yonekawa T, Kuwana M, et al. Clinical and histological findings associated with autoantibodies detected by RNA immunoprecipitation in inflammatory myopathies. J Neuroimmunol. 2014 Sep 15;274(1-2):202-8.