MOC Self-Assessment Questions

JUNE 2015 CHICAGO DERMATOLOGICAL SOCIETY CONFERENCE

HOST – LOYOLA UNIVERSITY

1. Langerhan cells stain positive with which of the following?

1. PAS
2. **CD1a**
3. CD20
4. Giemsa
5. Fite

1.

Answer: B.

CD1a stains Langerhans cells. PAS stains glycogen and mucin. CD 20 is a B cell marker. Giemsa classically is a maker for mast cells. Fite is an acid fast stain for leprosy.

Favara B, Feller A et al. Classification of Histiocytic Disorders. Medical and Pediatric Oncology. 1997. 29:157-166.

2. A patient presents with diabetes insipidus, exophthalmos and cranial bone lesions. Based the historical subclassifications of Langerhans cell histiocytosis, what is the diagnosis?

1. Letter-Siwe disease
2. **Hand-Schuller-Christian disease**
3. Eosinophilic granuloma
4. Congenital self healing reticulohistiocytosis
5. Infantile fibromatosis

2.

Answer: B.

Hand-Shuller Christian presents as a triad of diabetes insipidus, exophthalmos and bone lesions, most often of the cranium. Letter-Siwe is characterized by diffuse skin involvement , bone and visceral involvement. Eosinophilic granuloma is characterized by solitary bone lesions. Congenital self healing reticulohistiocytosis presents with diffuse skin involvement with good prognosis.

Bolognia J, Jorizzo J, Schaffer J. Dermatology. China: Elsevier, 2012.

3. Histopathological examination of a lesion on the forearm demonstrates a band-like lymphoid infiltrate, hypergranulosis, destruction of the basal layer, and numerous Civatte bodies. The most likely diagnosis is:

1. Lichen Striatus
2. PLEVA
3. **Lichen Planus**
4. PMLE
5. Erythema Multiforme

3.

Answer C:

Lichen planus is characterized by a lichenoid interface dermatitis with destruction of the basal layer, Civatte bodies, and a sawtooth rete ridge pattern. Lichen planus is not typically associated with parakeratosis or eosinophils.

Friedman DB, Hashimoto K. Annular atrophic lichen planus. J Am Acad Dermatol. 1991;25:392-394.

4. Which of the following medications has level A evidence as a highly effective systemic therapy for refractory lichen planus of the skin?

1. **Acitretin**
2. Metronidazole
3. Chloroquine
4. Azathioprine
5. Thalidomide

4.

Answer A:

Acitretin is the only agent against lichen planus that has achieved level A evidence as a highly effective systemic therapy. It was shown to be highly effective in a placebo-controlled double-blind study including 65 patients with a 64% response rate compared to 13% in the placebo group. It is typically administered at a dose of 0.5 to 0.7mg/kg body weight until remission is achieved followed by 0.3 to 0.5mg/kg body weight thereafter. It can be used as a monotherapy or in combination with topical or systemic steroids.

Manousaridis I, Manousaridis K, Peitsch WK, Schneider SW. Individualizing treatment and choice of medication in lichen planus: a step by step approach. J Dtsch Dermatol Ges. 2013;981-991

5. The most common subtype of mastocytosis seen in the pediatric population is?

1. **urticaria pigmentosa**
2. mastocytoma
3. diffuse cutaneous mastocytosis
4. telangiectasia macularis eruptiva perstans
5. leukemia

5.

Answer A:

Mastocytosis begins during childhood in 2/3 of cases. Most commonly, pediatric mastocytosis is characterized by urticaria pigmentosa, with mastocytomas and diffuse cutaneous mastocytosis being much less prevalent. TMEP is more frequent in adult mastocytosis and is exceedingly rare in the pediatric population.

6. What percentage of pediatric mastocytosis progress to systemic disease?

1. **1-5%**
2. 15-20%
3. 40-50%
4. 75-80%
5. 90-100%

6.

Answer A:

About 3% of pediatric mastocytosis cases are aggressive and progress to mast cell leukemia or mast cell sarcoma with systemic disease. As a result, long term follow up of affected patients is extremely important, especially given the >50% mortality in children with mast cell leukemia or sarcoma.

Previous 2 questions from: Méni C, Bruneau J, Georgin-Lavialle S et al. Paediatric mastocytosis: a systematic review of 1747 cases. Br. J. Dermatol., 2015 vol. 172(3) pp. 642-51.

7. Which of the following cutaneous signs correlates with disease activity in juvenile dermatomyositis?

1. Gottron's papules
2. **Nail fold capillary paucity**
3. Heliotrope rash
4. Widespread erythema
5. Pruritus

7.

Answer B:

Reduced nail fold capillary density has been shown to be a highly sensitive diagnostic marker for juvenile dermatomyositis but also a measure of disease activity over time. The other cutaneous findings listed did not show statistically significant correlation with disease activity.

Schmeling H, Stephens S, Goia C et al. 'Nailfold capillary density is importantly associated over time with muscle and skin disease activity in juvenile dermatomyositis' Rheumatology 2011;50:885-893.

8. Which of the following autoantibodies are associated with calcinosis cutis in patients with juvenile dermatomyositis?

1. **Anti-p140**
2. Anti-Mi-2
3. Anti-Jo
4. ANA
5. Anti-U1-RNP

8.

Answer A:

Anti-p140 antibody was significantly associated with the presence of calcinosis cutis in juvenile dermatomyositis. Anti-Mi2 is associated with cutaneous disease but milder muscle disease. ANA is found to be positive in 65-70% of cases of juvenile dermatomyositis but has not been correlated with clinical presentation. Anti-U1-RNP is usually found in overlap syndromes. Anti-Jo has not been correlated with any significant clinical finding.

Gunawardena H, Betteridge ZE, McHugh NJ: Myositis-specific autoantibodies: their clinical and pathogenic significance in disease expression. Rheumatology (Oxford).48:607-612 2009.

9. Which of the following is most commonly located in a midline location on the neck in a pediatric patient:

1. Second Branchial Cleft Cyst
2. **Bronchogenic Cyst**
3. Schwannoma
4. Dermoid Cyst
5. Epidermal Cyst

9.

Answer B:

Bronchogenic Cyst: Bronchogenic cysts are small, solitary cysts or sinuses, most typically located in a midline location in the region of the suprasternal notch or over the manubrium. Branchial cleft cysts occur over the sternocleinomastoid muscle. Dermoid cysts are commonly found superolateral to the lateral eyebrow. Schwannomas and epidermal cysts can occur on any surface of the body.

Reference: Gaikwad N, Uttam Sathe N, Wadkar G, et al. Schwannoma of the Cervical Vagus Nerve in a Child: A Case Report. 2013. Indian J Otolaryngol Head and Neck Surgery;65(1):S188-S191

10. Histological features of Schwannomas include which of the following:

1. Small, regularly spaced vessels in Antoni B areas
2. Non-encapsulated
3. Myxoid Antoni A and Palisading Verocay body Antoni B areas
4. **Hypercellular Antoni A and Hypocellular Antoni B areas**
5. Hypocellular Antoni A and Hypercellular Antoni B areas

10.

Answer D:

Hypercellular Antoni A and Hypocellular Antoni B areas: Antoni A and B tissue types represent distinct histologic architectural patterns that aid in the histopathologic diagnosis of schwannomas. Type A tissue is highly cellular and demonstrates nuclear palisading and associated Verocay bodies, which reflects their prominent extracellular matrix and secretion of laminin. Type B tissue is loosely organized with myxomatous and cystic changes and may represent degenerated Antoni A tissue.

Reference: Behuria S et al. Diagnosis and Management of Schwannomas Originating From the Cervical Vagus Nerve. Ann R Coll Surg Engl. 2015;97(2):92-97

11. Pyogenic arthritis, pyoderma gangrenosum and acne syndrome (PAPA) involves dysregulation between pyrin and PSTPIP1 protein interaction. What other condition shares a common pathoetiology?

1. Job's Syndrome
2. Still's Disease
3. **Familial Mediterranean Fever Syndrome**
4. Crohn's Disease
5. Mixed cryoglobulinemia

11.

Answer C:

PSTPIP1 binds to pyrin and mutations in pyrin result in familial Mediterranean fever. Since disease-associated mutations in PSTPIP1 enhance pyrin binding, PAPA syndrome and FMF are thought to share a common pathogenesis.

References:

Waite AL, Schaner P, Richards N et al. Pyrin modulates the intracellular distribution of PSTPIP1. PLoS One 2009; 4:e6147

Dinarello CA, Van der Meer JWM. Treating inflammation by blocking interleukin-1 in humans. Semin Immunol 2013; 25:469-84

12. What type of arthritis is associated with PAPA syndrome (Pyogenic arthritis, pyoderma gangrenosum and acne)?

1. Classical rheumatoid arthritis
2. Bowel-associated arthropathy
3. Progressive erosive seronegative arthritis
4. **Sterile erosive arthritis**
5. Gram positive inflammatory arthritis

12.

Answer D:

Polyarthritis is frequently associated with pyoderma gangrenosum. A literature review involving 133 cases of PG reported associations with three separate phenotypes of arthritis including classical rheumatoid arthritis, bowel-associated arthropathy, and progressive erosive seronegative arthritis.

Arthritis associated with PAPA syndrome is specifically a sterile erosive arthritis

Reference:

DeFilippis EM, Feldman SR, Huang WW. The genetics of pyoderma gangrenosum and implications for treatment: a systemic review. Br J Dermatol 2014

13. Eosinophilic pustular folliculitis is most commonly found in which of the following populations?

1. Japanese males
2. Neonates and infants
3. **HIV-positive individuals**
4. Bone marrow transplant patients
5. Chemotherapy recipients

13.

Answer C:

The three clinical types of eosinophilic pustular folliculitis include classic EPF (Ofuji's disease) occurring in Japanese middle-aged adults, infantile EPF occurring in neonates and infants, and immunosuppression-associated EPF including HIV-positive individuals and HIV-negative patients including those with hematologic or lymphoproliferative disorders, following bone marrow transplant, and chemotherapy-induced. EPF associated with HIV infection is now the now the most common clinical type.

Eosinophilic pustular folliculitis in patients with acquired immunodeficiency syndrome. Report of three cases. J Am Acad Dermatol 14(6):1020-1022, 1986.

Human immunodeficiency virus-associated eosinophilic folliculitis. A unique dermatosis associated with advanced human immunodeficiency virus infection. Arch Dermatol 127(2):206-209, 1991.

Eosinophilic folliculitis: Before and after the introduction of antiretroviral therapy. Arch Dermatol 141(10):1227-1231, 2005.

14. Which medication is considered first line oral therapy for classic eosinophilic pustular folliculitis (Ofuji's disease)?

1. Isotretinoin
2. Metronidazole
3. Anti-retroviral therapy
4. Minocycline
5. **Indomethacin**

14.

Answer E:

Topical steroids and calcineurin inhibitors are generally the first approach to treatment of all types of eosinophilic pustular folliculitis. NSAIDs, in particular indomethacin, is recommended as first-line therapy for classic EPF. Anti-retroviral treatments is considered first line therapy for HIV-associated EPF. Other second line treatments which have been used with some success include metronidazole, isotretinoin, and minocycline.

Eosinophilic pustular folliculitis: A comprehensive review of treatment options. Am J Clin Dermatol 5(3):189-197, 2004

Therapeutic effectiveness of various treatments for eosinophilic pustular folliculitis. Acta Derm Venereol 89(2):155-159, 2009

Eosinophilic pustular folliculitis (Ofuji's disease): Indomethacin as a first choice of treatment. Clin Exp Dermatol 26(2):179-181, 2001

15. Which of the following medications has been associated with the development of eosinophilic pustular folliculitis?

1. **Cyclophosphamide**
2. Hydralazine
3. Sulfamethoxazole/trimethoprim
4. Lisinopril
5. Prednisone

15.

Answer A:

EPF associated with drug therapy has been reported with minocycline, indeloxazine hydrochloride, carbamazepine, allopurinol with and cyclophosphamide. The case report involving cyclophosphamide involves cyclophosphamide alone or exhibiting a synergistic effect with 5-fluorouracil.

Eosinophilic pustular folliculitis induced by chemotherapy. J am Acad Dermatol 54(4):729-730, 2006.

16. A child presents with inflammatory tender pustules on the legs that ulcerate. Biopsy shows neutrophilic dermatosis consistent with pyoderma gangrenosum. What is the most likely underlying systemic condition?

1. Irritable bowel syndrome
2. **Ulcerative colitis**
3. Crohn's disease
4. Diabetes mellitus
5. Arthritis

16.

Answer B:

Commonly, pediatric patients with PG have underlying systemic disease. Arthrtis, bowel disease and hematologic malignancy are most frequently seen. In one study ulcerative colitis is seen in about 25% of patients with PG, while Crohn's occurred in ~15%. Similarly, only 3% of patient's with Crohn's disease develop PG. Arthritis is commonly associated with PG in adults, but is much less common in children (<15%). Irritable bowel syndrome and diabetes mellitus are not associated with PG.

Graham JA, Hansen KK, Rabinowitz LG, Esterly N. Pyoderma gangrenosum in infants and children. Pediatr Dermatol 1994 Mar; 11(1): 10-7.

17. First line therapy for moderate to severe pyoderma gangrenosum is:

1. **Prednisone and local wound care**
2. Topical tacrolimus and local wound care
3. Prednisone and adjuvant dapsone
4. Intralesional kenalog
5. Methotrexate

17.

Answer A:

Topical steroids and, more recently, calcineurin inhibitors may be effective in early or mild PG, but are not effective in severe lesions. Prednisone is favored as first line therapy due to its rapid onset of action, well-known side effect profile and minimal cost. Pain control should also be addressed.

Miller J, Yenzer BA, Clark A, Jorizzo JL, Feldman SR. Pyoderma gangrenosum: a review and update on new therapies. J Am Acad Dermatol 2010 Apr; 62(4): 646-54.

18. A 70 year old man presents with juicy erythematous papules and nodules on the head and neck, and skin biopsy shows a predominantly B-cell infiltrate. He occasionally feels fatigued but the remainder of his review of systems is negative. Which of the following is the most likely diagnosis?

1. Acute lymphocytic leukemia
2. Sweet's syndrome
3. **Chronic lymphocytic leukemia**
4. Neutrophilic eccrine hidradenitis
5. Hodgkin disease

18.

Answer C:

Of the answer choices, the patient fits the most common profile of a patient with chronic lymphocytic leukemia due to his age, presentation, and histopathological findings. Acute lymphocytic leukemia is commonly found in the pediatric population. Sweet's syndrome and neutrophilic eccrine hidradenitis both may be associated with malignancy and are characterized by neutrophilic, not B cell infiltrates. Hodgkin disease is most commonly seen in young adults, and limited cutaneous involvement does not usually occur with systemic symptoms of the malignancy; otherwise, disseminated cutaneous lesions are a sign of established advanced disease.

References:

Weisshaar E, Fleischer AB, Bernhard JD, Cropley TG. Pruritus and dysesthesia. In: Bolognia JL, Jorizzo JL, Rapini RP, editors. Dermatology. 3rd ed. Mosby:London. 2012.

Davis MD, Perniciaro C, Dahl PR, et al. Exaggerated arthropod-bite lesions in patients with chronic lymphocytic leukemia: a clinical, histopathologic, and immunopathologic study of eight patients. J Am Acad Dermatol. 1998;39:27–35.

Winfield HL, Smoller BR. Other lymphoproliferative and myeloproliferative diseases. In: Bolognia JL, Jorizzo JL, Rapini RP, editors. Dermatology. 3rd ed. Mosby:London. 2012.

19. Juvenile dermatomyositis can be treated with IVIG. IVIG therapy should not be administered to patients with which one of the following?

1. **IgA deficiency**
2. Kawasaki syndrome
3. Severe combined immune deficiency (SCID)
4. Patients who recently received a vaccination
5. Hematologic malignancy

19.

Answer A:

Patients with IgA deficiency and anti-IgA antibodies are at greater risk of anaphylaxis or hypersensitivity to IVIG. IVIG therapy is used to treat Kawasaki syndrome and SCID. Patients who recently received a vaccination and are undergoing IVIG therapy may need to have titers checked and vaccination repeated as they may not respond. However, recent vaccination is not a contraindication to IVIG therapy.

Burks AW, Sampson HA, Buckley RH. Anaphylactic reactions after gamma globulin administration in patients with hypogammaglobulinemia. Detection of IgE antibodies to IgA.

N Engl J Med. 1986;314(9):560.

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