MOC Self-Assessment Questions

NOVEMBER 2015

CHICAGO DERMATOLOGICAL SOCIETY CONFERENCE

HOST – NORTHWESTERN UNIVERSITY

Use your “clicker” to indicate your answer to the following questions.

You can obtain a copy of the slides, including references, on the protocol book page of the CDS website after the meeting.

You will receive credit for 8 MOC questions today.

1. Which of the following mycosis fungoides (MF) variants or mimics carries the worst prognosis?

1. **Folliculotropic mycosis fungoides**
2. Granulomatous slack skin (GSS)
3. Lymphomatoid papulosis (LyP)
4. Pagetoid reticulosis
5. Primary cutaneous anaplastic large cell lymphoma (PCALCL)

1.

Folliculotropic mycosis fungoides carries the worst prognosis of the conditions listed above, with an estimated 5-year survival rate of 80%. In contrast, granulomatous slack skin (GSS) is an extremely rare variant of MF that presents clinically with pendulous, lax skin in the intertriginous zones but can involve other sites as well. This variant does not portend any adverse prognostic significance and in fact is thought to have near 100% 5-year survival. Lymphomatoid papulosis (LyP) is a CD30+ lymphoproliferative disorder (LPD). Although there is debate regarding its nosology, most consider it a very low-grade form of CTCL. Although associated in some cases with other hematologic malignancies, LyP itself is thought to represent a largely benign process. Pagetoid reticulosis is another rare variant of mycosis fungoides. It is characterized clinically by a localized and indolent patch or plaque, often on an extremity, and histologically by a strictly intraepidermal proliferation (hence "pagetoid") of the atypical T cells. Like granulomatous slack skin, pagetoid reticulosis is thought to be a benign variant of mycosis fungoides that essentially never causes disease-specific death. Primary cutaneous anaplastic large cell lymphoma (C-ALCL), along with LyP, constitutes the remainder of the CD30+ lymphoproliferative disorder (LPD) spectrum. It does demonstrate extracutaneous spread in some cases and carries an estimated 95% 5-year survival rate, worse than LyP, GSS, and pagetoid reticulois but better than folliculotropic MF.

REFERENCES:

1. Willemze R. Cutaneous T-Cell Lymphoma. In: Bolognia JL, Jorrizo JL, Schaffer JV, eds. Dermatology. 3rd ed. Philadelphia, PA: Elsevier,Inc; 2012:chap 120.

2. Willemze R, Jaffe ES, Burg G, et al. WHO-EORTC classification for cutaneous lymphomas. Blood 2005;105:3768-85.

2. Which of the following is thought to be a risk factor for the development of radiation-induced morphea?

1. Age at time of radiation
2. Radiation dose
3. **Systemic sclerosis**
4. Prior radiation recall
5. Breast cancer

2.

It is believed that systemic sclerosis is a relative risk factor for developing an exaggerated post-irradiation fibrosis. Age and radiotherapy parameters such as total radiation dose, dose per fraction and severity of acute reaction do not seem to be significant risk factors for developing radiation-induced morphea (RIM). Breast cancer appears to be associated with RIM but there have been no studies as of yet to confirm it as a risk factor.

Reference(s):

1. Bleasel NR, Stapleton KM, Commens C, Ahern VA. Radiation-induced localized scleroderma in breast cancer patients. Australas J Dermatol. 1999;40:99–102.

3. A 2-year-old female presents for evaluation of erythematous, scaling and crusted plaques on the scalp, post-auricular skin and inguinal folds that have persisted for 6 months. She has been unresponsive to treatment with topical ketoconazole. Which of the following is true regarding this condition?

1. This condition is frequently associated with HIV infection.
2. A PET-CT scan is an important initial step in the work-up of this condition.
3. The prognosis for this condition is excellent and it often resolves spontaneously.
4. **Representative histopathology would be expected to stain positive for S100 and CD1a.**
5. This condition is associated with polyclonal expansion of bone marrow-derived granulocytes in the skin

3.

The patient in this scenario most likely carries a diagnosis of Langerhans cell histiocytosis (LCH). Immunostaining for S100 and CD1a is considered diagnostic in LCH. While seborrheic dermatitis can be seen with increased frequency and severity in association with HIV, LCH is not seen with increased frequency in the patients with HIV. A skeletal survey is recommended in the initial work-up of LCH due to high frequency of lytic bone lesions in this condition. A PET-CT scan is not considered first-line in evaluation due to the increased amount of radiation delivered to the child. Any further imaging would be based on associated symptoms and is not typically part of the initial evaluation. The prognosis in LCH is highly variable and depends on the extent of organ involvement found on work-up. In general, multisystem disease portending a worse overall prognosis and is an indication for systemic treatment. While it remains controversial as to whether this condition represents a true malignancy, the infiltrate in LCH has been shown to be of monoclonal origin. It is composed of bone marrow-derived Langerhans cells, which are dendritic cells that function in antigen presentation.

Reference(s):

1. Salter EK, High WA. Langerhans cell histiocytosis: A review of the current recommendations of the Histiocyte Society. Pediatr Dermatol. 2008;25(3):291-295.

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

4. A 22-year-old female is diagnosed with localized granuloma annulare. Which of the following histopathologic patterns would be expected on biopsy of a characteristic lesion?

1. Giant cells and asteroid bodies with noncaseating granulomas in the superficial and deep dermis
2. Necrobiosis with palisading and interstitial granulomas in a horizontally layered fashion involving the entire dermis
3. **Necrobiosis surrounded by palisading or interstitial granulomas with mucin deposition involving the superficial and mid dermis**
4. Giant cells with noncaseating granulomas in the superficial and deep dermis
5. Necrobiosis with giant cells and palisading granulomas in the deep dermis and subcutis

4.

The noninfectious granulomatous skin conditions share similar histopathologic patterns, with the hallmark feature being a predominantly histiocytic inflammatory infiltrate. Despite their similarities, these disorders can be distinguished by location and pattern of the granulomatous infiltrate, as well as by the presence or absence of necrobiosis (degeneration of collagen). Granuloma annulare (GA) classically demonstrates foci of necrobiosis in the superficial and mid dermis, surrounded by the histiocytic infiltrate. The presence of mucin also helps to distinguish this disorder. In contrast, sarcoidosis classically demonstrates the sharply demarcated epithelioid granulomas with few surrounding lymphocytes ("naked" granulomas), as well as giant cells with eosinophilic stellate inclusion bodies (asteroid bodies). In cutaneous Crohn's disease, similar granulomas are found, however the infiltrate is surrounded by numerous lymphocytes and lacks the asteroid bodies. Necrobiosis lipoidica and rheumatoid nodules have degenerated collagen, as in GA. However, necrobiosis lipoidica shows the classic "tiered" pattern to its granulomatous infiltrate. Rheumatoid nodules can be distinguished by the deep location of the granulomas on histology and by its distinct clinical features.

Reference(s):

1. Kamil ZS, et al. Non-infectious granulomatous conditions of the skin: a clinicopathological approach to diagnosis. J Clin Pathol 2015;21:Epub ahead of print.

2. Thornsberry LA, & English III, JC. Etiology, diagnosis, and therapeutic management of granuloma annulare: an update. Am J Clin Dermatol 2013;14:279–290.

3. Keimig EL. Granuloma annulare. Dermatol Clin 2015;33:315–329.

4. Ball NJ, et al. The histiologic spectrum of cutaneous sarcoidosis: a study of twenty-eight cases. J Cutan Pathol 2004;31(2):160-8.

5. A middle-aged Caribbean male presents with a 6-month history of a diffuse eczematous eruption. He has no other significant past medical history. He was previously diagnosed with mycosis fungoides stage IB, but was unresponsive to topical steroids and narrowband-UVB. What virus is most likely implicated in this patient's disease?

1. Epstein Bar virus
2. Human immunodeficiency virus
3. **Human T-lymphotrophic virus type-1**
4. Merkel cell polyomavirus
5. Human herpes virus 8

5.

HTLV-1 is a virus that is endemic to the Caribbean, Japan, South America, and Africa and is the first virus implicated in causing malignancy in humans. Adult T-cell leukemia/lymphoma (ATLL) has varied cutaneous manifestations and should be suspected if the patient is of Caribbean or Japanese origin and has an unusual presentation of T-cell lymphoma. All the other options have been implicated in causing malignancy in humans, but are usually seen in immunocompromised patients and do not typically present in a CTCL-like picture.

Reference(s):

1. McGill NK, Vyas J, Shimauchi T, Tokura Y, Piguet V. HTLV-1-associated infective dermatitis: updates on the pathogenesis. Exp Dermatol. 2012;21:815-21.

2. Marchetti MA, Pulitzer MP, Myskowski PL, et al. Cutaneous manifestations of human T-cell lymphotrophic virus type-1-associated adult T-cell leukemia/lymphoma: a single-center, retrospective study. J Am Acad Dermatol. 2015;72:293-301.

3. Shimoyama M. Diagnostic criteria and classification of clinical subtypes of adult T-cell leukaemia-lymphoma. A report from the Lymphoma Study Group (1984-87). Br J Haematol. 1991;79:428-37.

4. Karube K, Aoki R, Sugita Y, et al. The relationship of FOXP3 expression and clinicopathological characteristics in adult T-cell leukemia/lymphoma. Modern pathology: an official journal of the United States and Canadian Academy of Pathology, Inc 2008;21:617-25.

6. For localized cutaneous infections caused by Mycobacterium chelonae, what is the empiric antibiotic of choice?

1. Amoxicillin
2. **Clarithromycin**
3. Clarithromycin + linezolid
4. Clarithromycin + gentamycin
5. Linezolid

6.

Mycobacterium chelonae is a gram positive bacterium. It is one of the pathogens that cause atypical mycobacterium infection characterized by the clinical presentation of painful, nonhealing subcutaneous nodules and abscesses. For limited cutaneous infections of M. chelonae, the treatment of choice is clarithromycin 500 mg for 6 months. Surgical debridement may also be helpful in limited cutaneous disease. Recent evidence suggests linezolid as a promising oral therapy in cases where clarithromycin alone fails. Combination therapy of clarithromycin or azithromycin and gentamicin may be used for disseminated disease involving the ocular or pulmonary systems, or in cases of treatment failure with a macrolide alone.

Reference(s):

1. Wallace RJ Jr, Tanner D, Brennan PJ, Brown BA. Clinical trial of clarithromycin for cutaneous (disseminated) infection due to Mycobacterium chelonae. Ann Intern Med. 1993 Sep 15;119(6):482-6.

2. Wallace RJ Jr, Brown-Elliott BA, Ward SC, Crist CJ, Mann LB, Wilson RW. Activities of linezolid against rapidly growing mycobacteria. Antimicrob Agents Chemother. 2001 Mar;45(3):764-7.

7. Which of the following is a feature of congenital and erosive vesicular dermatosis healing with reticular scarring?

1. **Preterm birth**
2. Twin gestation
3. Forceps delivery
4. Maternal gestational diabetes
5. Cesarian delivery

7.

Congenital erosive and vesicular dermatosis (CEVD) is a rare disorder that presents at birth with vesicles, erythema, erosions, crusts, and ulceration involving at least 75% of the skin surface. Lesions heal within the first few months of life as supple, reticulated scars. The mode of inheritance of CEVD is thought to be sporadic with amniotic banding, intraunterine infections or a developmental defect of skin possibly playing a role. Infants with CEVD believed to be at risk for HSV infection; however, HSV infection is not thought to play a role in the pathogenesis of CEVD. In a recent report of 28 cases of CEVD, associated features included: preterm birth (76%), nail abnormalities (46%), hyperthermia/hypohidrosis (46%), history of maternal chorioamnionitis (43%), neurodevelopmental and ophthalmological abnormalities (36%) and tongue atrophy (29%).

Reference(s):

1. Tlougan BE, Paller AS, Schaffer JV, Podjasek JO, Mandell JA, Nguyen XH, Spraker MK, Hansen RC. Congenital erosive and vesicular dermatosis with reticulated supple scarring: unifying clinical features. J Am Acad Dermatol. 2013 Dec;69(6):909-15.

8. A 52-year-old male presents to your dermatology clinic for his yearly total body skin exam. His past medical history is notable for hypertension, hyperlidemia, obesity, psoriasis and Hepatitis C. He remarks that he is in his last week of treatment for Hepatitis C with pegylated interferon a-2a (IFN), ribavirin (RBV) and sofosbuvir. On examination, you are least likely to note which of the following:

1. **Alopecia areata**
2. Eczematous eruption
3. Folliculitis
4. Psoriasis flare
5. Xerosis

8.

Alopecia areata is a rare side effect that can occur in setting of treatment with IFN. One case series of 152 patients being treated with IFN and RBV showed an incidence rate of just 0.7% (1 patient). The other cutaneous findings are seen more commonly: eczematous eruption (9.2%), xerosis (2.6%), psoriasis (new onset or flare – 2.0%) and folliculitis (1.3%).

Reference(s):

1. Li Z, Zhang Y, An J, Feng Y, Deng H, Xiao S and Ji F. Predictive factors for adverse dermatological events during pegylated/interferon alpha and ribavirin treatment for hepatitis C. J Clin Vir. 2014; 60(3):190-195.

Thank you for participating!

TURN IN YOUR SURVEY SHEET TO THE REGISTRATION COUNTER!

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