MOC Questions
CDS - Rush
May 4th, 2016

DEPARTMENT OF DERMATOLOGY
RUSH UNIVERSITY MEDICAL CENTER

American Board of Dermatology

The Chicago Dermatological Society has been approved by the ABD to produce self-assessment activities.

Self-assessment activities are intended to be of value to the practicing dermatologist.

The Chicago Dermatological Society is solely responsible for the content.

Question 1:

A previously healthy 39 year old white female presents with a chief complaint of hair loss. On examination, you note diffuse non-scarring alopecia, few scaly palmar papules and a small, non-inflammatory ulceration on the left 5th distal palmar digit. There are no other cutaneous findings of note. The patient denies any history of muscle pain or weakness. On full ROS she does endorse new onset cough, which has been worsening for the last three weeks.

Of the following, which is the most important NEXT step in management?

A. Referral for pap-smear and mammogram to look for underlying malignancy
B. Send Myositis-Associated and Myositis-Specific antibodies and await results prior to obtaining any additional workup
C. Send serum CPK and Aldolase and await results prior to obtaining any additional workup
D. Referral for urgent pulmonary evaluation, including Pulmonary Function Testing (PFTs)
E. Recommend therapy with Minoxidil 5% and follow up in 6 months for hair loss

Answer 1

D - Referral for pulmonary evaluation, including Pulmonary Function Testing (PFTs)

Commentary:

The patient has characteristic cutaneous findings of diffuse, non-scarring alopecia, scaly palmar papules and non-inflammatory acral ulcerations concerning for MDA-5 antibody positive dermatomyositis. This is supported by lack of myositis on history (MDA-5 DM patients are more likely to be amyopathic or hypomyopathic). MDA-5 DM is associated with increased risk for interstitial lung disease (ILD) which can be rapidly progressive and even fatal. Although answers A, B and C are all correct and can be done as part of further management of this patient, the most important NEXT step in management would be referral for pulmonary evaluation to assess for presence of ILD which can be rapidly progressive in these patients.

References

Gil B, Merav L, Pnina L, Chagai G. Diagnosis and treatment of clinically amyopathic dermatomyositis (CADM): a case series and literature review. *Clin Rheumatol*. 2015 Apr 7.

Kiely PD, Chua F. Interstitial lung disease in inflammatory myopathies: clinical phenotypes and prognosis. *Curr Rheumatol Rep*. 2013 Sep;15(9):359.

Question 2

Which of the following is a risk factor for invasive fungal dermatitis?

A. Vaginal delivery
B. Prolonged hypoglycemia
C. Late gestational age
D. Maternal preeclampsia
E. Hypocomplementemia

Answer 2

A – Vaginal Delivery

Commentary:

Known risk factors for invasive fungal dermatitis include low birthweight, early gestational age, prolonged hyperglycemia, postnatal steroid administration and vaginal delivery.

References:

Passeron T, Desruelles F, Gari-Toussaint M, Dageville C, Lacour JP. Invasive fungal dermatitis in a 770 gram neonate. *Pediatric dermatology*. 21(3):26-261, 2004.

Question 3

Which is the most common extra-cutaneous malignancy associated with Muir Torre Syndrome?

A. Glioblastoma multiforme
B. Uveal melanoma
C. Colonic adenocarcinoma
D. Transitional bladder carcinoma
E. Endometrial carcinoma

Answer 3

C – Colonic Adenocarcinoma

Commentary:

The visceral malignancy most commonly associated with Muir Torre Syndrome is colorectal adenocarcinoma, followed by cancers of urogenital tract, small intestine, and breast.

Reference:

John AM, Schwartz RA. Muir-Torre syndrome (MTS): An update and approach to diagnosis and management. *Journal of the American Academy of Dermatology*. 2016 Mar 31;74(3):558-66.

Question 4

Which of the following predisposes to cutaneous lymphoid nodular hyperplasia?

A. Infection with group A Streptococcus
B. Sjogren's Disease
C. Infection with Borrelia burgdorferi
D. Beta lactam antibiotics
E. Female gender

Question 4:

C, infection with Borrelia burgdorferi

Commentary:

Although the majority of cases of cutaneous lymphoid nodular hyperplasia have no known etiology, previous trauma, tattoos, arthropod bites, vaccinations and infections with Borrelia burgdorferi have been reported. Males are more frequently affected than females.

Reference:

Albrecht S, Hofstadter S, Artsob H. Lymphadenosis benigna cutis resulting from Borrelia infection (borrelial lymphocytoma). *J Am Acad Dermatol.* 1991;24:621-5.

Question 5

What is the change in the average number of future biopsies for patients with widespread atypical nevi at risk for melanoma after total body photography?

A. Increase 2-fold
B. Increase 4-fold
C. Decrease 2-fold
D. Decrease 4-fold
E. No Changes Observed

Question 5:

D - Decrease 4 fold

Commentary: Patients at risk for melanoma experienced a 3.8-
fold reduction in nevus biopsies after total body photography.

Reference:

Truong A, Strazzulla L, March J, et al. Reduction in nevus biopsies in patients monitored by total body photography. *J Am Acad Dermatol.* 2016; S0190-9622(16)01329-3 [Epub Ahead of Print]

Question 6

Kasabach-Merritt Syndrome is most commonly occurs in the setting of which of the following?

A. Infantile hemangiomas
B. Arterio-venous malformations
C. Kaposiform hemangioendothelioma
D. Port wine stain
E. Angiokeratomas

Answer 6

C – Kaposiform hemangioendothelioma

Commentary:

Though Kasabach-Merritt syndrome was first described in association with a "capillary hemangioma", standardization of the medical lexicon and extensive review of histopathology has shown that this coagulopathy is actually associated with tufted angiomas and kaposiform hemangioendotheliomas, not infantile hemangiomas.

Reference:

Enjolras O, Wassef M, Mazoyer E, Frieden IJ, Rieu PN, Drouet L, Taïeb A, Stalder JF, Escande JP. Infants with Kasabach-Merritt syndrome do not have "true" hemangiomas. *The Journal of pediatrics*. 1997 Apr 30;130(4):631-40.

Question 7

- A 23 year-old female presents to your office with a 1 year history of intermittent, edematous papules and plaques on her face and forearms after exposure to cold and windy conditions. She has a positive ice-cube challenge test in your office. You diagnose her with primary acquired cold urticaria and counsel her on the importance of avoiding cold triggers. What percentage of these patients experience at least one additional systemic symptom like hypotension, dizziness, and shortness of breath during flares?

A. 1%

B. 10%

C. 30%

D. 70%

E. 90%

Answer 7

C – 30%

Commentary:

Studies of both adult and pediatric patients have found that approximately 30% of patients with primary acquired cold urticaria will display at least 1 systemic symptom in addition to the cutaneous lesions. The most common symptoms are cardiovascular followed by respiratory. It is important to counsel patients on the risk of potentially serious associated symptoms.

Reference:

Alangari AA, Twarog FJ, Shih MC, Schneider LC. Clinical features and anaphylaxis in children with cold urticarial. *Pediatrics*. 2004;113(4):313-7.

Wanderer AA, Grandel KE, Wasserman SI, Farr RS. Clinical characteristics of cold-induced systemic reactions in acquired cold urticarial syndromes: recommendations for prevention of this complication and a proposal for a diagnostic classification of cold urticarial. *J Allergy Clin Immunol*. 1986;78:417-23.

Question 8

Which interadermal filler is most likely to cause a foreign body reaction?

A. Autologous Fat
B. Bacteria-Derived Hyaluronic Acid
C. Rooster-Comb Derived Hyaluronic Acid
D. Polymethyl-methacrylate (PMMA)
E. Bovine Collagen

Answer 8

D – Poly-methyl methacrylate (PMMA, Artefill)

Commentary

Autologous fat is the least likely to initiate a foreign body reaction, followed by hyaluronic acid, as that molecule is conserved across both tissues and species. Older fillers such as bovine collagen and PMMA have been widely reported to induce foreign body reactions, but PMMA (artefill and artecoll) has been most reports. For that reason this filler is far less popular than the HA based fillers.

Reference:

Alam M, Gladstone H, Kramer EM, et al. ASDS guidelines of care: injectable fillers. *Dermatol Surg.* 2008; 34: S115-48

Lee JM, Kim YJ. Foreign body granulomas after the use of dermal fillers: pathophysiology, clinical appearance, histologic features, and treatment. *Arch Plast Surg.* 2015; 42: 232-9

Question 9

A premature baby in the NICU is noted to have 10 scattered cutaneous vascular lesions in the 2nd week of life consistent with infantile hemangiomas. The baby is hemodynamically stable. What is the most appropriate next step?

A. Head CT
B. Skin biopsy
C. Thyroid panel
D. Abdominal ultrasound
E. Chest x-ray

Answer 9

D – Abdominal Ultrasound

Commentary:

The actual number of cutaneous IH required to precipitate concern remains a floating target, but most people use > 5 as a guideline for hepatic screening based on an a retrospective review of hepatic hemangiomas by Dickie et al. The screening test of choice is a complete abdominal ultrasound with Doppler imaging. Hepatic hemangiomas found through screening ultrasonography are less likely to develop serious clinical sequelae. Although the reasons for this may include detection of hemangiomas that are less likely to progress to symptomatic disease, it appears that it also allows for earlier intervention for more concerning (e.g. diffuse) subtypes. Screening may allow for closer surveillance and earlier treatment before life-threatening progression in a subset of infants with liver hemangiomas, preventing complications and reducing mortality.

References

Rialon KL, Murillo R, Fevurly RD, et al. Impact of Screening for Hepatic Hemangiomas in Patients with Multiple Cutaneous Infantile Hemangiomas. *Pediatr Dermatol*. 2015 Nov-Dec;32(6):808-12.

Dickie BH, Fishman SJ, Azizkahn RG, et al. Hepatic Vascular Tumors. *Seminars in Pediatric Surgery.* 2014;23(4):168-172.

Question 10

What is a potential treatment option for diffuse, systemic cytotoxic T-cell lymphoma?

A. PUVA
B. Topical steroids
C. Romidepsin
D. Local radiotherapy
E. Topical chemotherapy

Answer 10

C – Romidepsin

Commentary:

To date, there is still no definite successful therapy available and the treatment has proven extremely challenging with even the most aggressive modalities proving ineffective in establishing long-term cure. In general, initial treatment with skin-directed therapy including topical chemotherapy, PUVA, and local radiotherapy are show only marginal efficacy. Romidepsin is an anticancer agent approved in 2009 that targets histone deacetylases to induce apoptosis in patients with CTCL. Some providers advocate allogeneic stem cell transplant, but this is controversial.

References

Berti E, Tomasini D, Vermeer MH, et al. Primary Cutaneous CD8-Positive Epidermotropic Cytotoxic T Cell Lymphomas : A Distinct Clinicopathological Entity with an Aggressive Clinical Behavior. *The American Journal of Pathology*. 1999;155(2):483-492.

Nofal A1, Abdel-Mawla MY, Assaf M, et al. Primary cutaneous aggressive epidermotropic CD8+ T-cell lymphoma: proposed diagnostic criteria and therapeutic evaluation. *J Am Acad Dermatol*. 2012 Oct;67(4):748-59.

Gormley RH1, Hess SD, Anand D, et al. Primary cutaneous aggressive epidermotropic CD8+ T-cell lymphoma. *J Am Acad Dermatol*. 2010 Feb;62(2):300-7.

Thank you for participating!

Please return your MOC evaluation form to the registration table before you leave the conference.