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Case 1

Yoo Jung Kim, MD
Parul Kathuria Goyal, MD
Cuong V. Nguyen, MD

A 73-year-old male is admitted to the hospital with rapidly progressive interstitial lung disease. On physical examination, he is found to have painful palmar papules and ulcerations over the distal fingertips. Which of the following is true regarding his underlying diagnosis?

A. Associated with underlying occult malignancy
B. Cardiac dysfunction is major cause of mortality
C. Ferritin is commonly dramatically elevated
D. Patients typically present with profound myopathy
E. Prognosis for this subtype of patients is excellent

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Explanation: This patient has anti-MDA5 clinically amyopathic dermatomyositis, a subtype of DM associated with rapidly progressive interstitial lung disease causing an overall poor prognosis as well as painful palmar papules with digital ulcerations. This subtype is not commonly associated with underlying malignancy. Ferritin is commonly dramatically elevated in this subtype of DM (C).

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Case 2

Samantha Venkatesh, MD
Pedram Yazdan, MD
Inderjit Kaur Gill, MD

A 35-year-old male presents to dermatology clinic with growing symmetric tender pink-brown papulonodules over the acral and extensor surfaces. Biopsy of the lesion shows a dense neutrophilic infiltrate with leukocytoclasis. Which of the following statements is true regarding this disease?

A. Dapsone is considered first-line treatment
B. DIF demonstrating perivascular fibrinogen is often strongly positive
C. Granuloma formation is seen on pathology at all stages of the disease
D. Lipid deposition is seen on histopathology in early lesions
E. This condition is strongly associated with staphylococcal infection

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Explanation: This patient has erythema elevatum diutinum, a rare and chronic cutaneous small vessel vasculitis that commonly presents with pink-brown papulonodular lesions over the acral and extensor surfaces. Dapsone is considered first-line treatment for this condition (A).

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Case 3

Victor Quan, MD
Xiaolong Alan Zhou, MD, MSc

All of the following are generally accepted features of Rowell syndrome except:

A. IgG/C3 at the BMZ in a granular pattern on DIF
B. Lack of a preceding drug or infectious exposure
C. Lack of acral involvement
D. Photosensitivity
E. Positive ANA in a speckled pattern

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D. **Photosensitivity**
E. Positive ANA in a speckled pattern

Explanation: Rowell syndrome is generally defined by the presentation of erythema multiforme-like lesions in association with lupus erythematosus. Multiple diagnostic criteria have been proposed since the original criteria proposed by Rowell et al. All of the above are generally accepted features of Rowell syndrome except for photosensitivity (D).

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Case 4

Taylor Erickson, MD
Karina Vivar, MD

Which of the following is true about eosinophilic fasciitis-like chronic cutaneous graft-versus-host disease?

A. Favors the extremities, most commonly involving the hands and feet
B. Histopathology normally shows very superficial dermal sclerosis
C. MRI is useful to identify deep fibrosis and fasciitis
D. Presents as an early manifestation of cutaneous cGvHD
E. Reported incidence of 30% among all cases of chronic cutaneous GvHD

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D. Presents as an early manifestation of cutaneous cGVHD
E. Reported incidence of 30% among all cases of chronic cutaneous GVHD

Explanation: Eosinophilic fasciitis-like chronic cutaneous graft-versus-host disease is a rare late manifestation of cGVHD that presents with deep dermal sclerosis of the extremities sparing the hands and feet. MRI is useful in this subtype of cGVHD to identify deep fibrosis and fasciitis (C).

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Case 5

Molly Hales, MD, PhD
Spencer Ng, MD, PhD
Joaquin Brieva, MD

A 56-year-old male presents to dermatology clinic for evaluation of palpable purpura over the lower extremities. He notes that he was recently diagnosed with crusted scabies and treated with oral ivermectin. Punch biopsy shows evidence of leukocytoclastic vasculitis, and DIF shows deposition of IgA. Which of the following labs should be monitored regularly over the next several months?

A. C-reactive protein
B. Erythrocyte sedimentation rate
C. Hemoglobin
D. Serum creatinine
E. White blood cell count

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E. White blood cell count

Explanation: IgA vasculitis has rarely been reported as a post-scabietic dermatosis. Patients diagnosed with IgA vasculitis should undergo long-term monitoring of serum creatinine (D) to monitor for renal involvement.

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Case 6

Zachary Solomon, MD
Lida Zheng, MD

A 16-year-old male with a history of congenital self-healing reticulocytosis presents to the emergency room with somnolence and confusion after a hike. Review of systems is positive for subacute polyuria and polydipsia. MRI of the brain shows a thickened pituitary stalk. Transsphenoidal pituitary biopsy confirms a diagnosis of reactivated Langerhans cell histiocytosis. Genetic analysis of the Langerhans cells from his biopsy would be most likely to show a mutation in which gene?

A. ARAF
B. BRAF
C. MAP2K1
D. ERBB3
E. ERK

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Explanation: BRAF (B) is an important part of the mitogen-activated protein kinase (MAPK) signaling pathway and has been found to be mutated in roughly 55% of LCH cases. This leads to constitutive signaling and activation of ERK. Subsequent to the discovery of pathogenic BRAF mutations, whole exome sequencing has identified other characteristic mutations in the MAPK pathway from LCH lesions including MAP2K1 (in 33-50% of all LCH without BRAF mutations), EBBB3, and ARAF, though these are less common.

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Case 7

Nicole S. Stefanko, MD
Anthony J. Mancini, MD

Which of the following is true regarding Hyper IgE syndrome?

A. Autosomal dominant forms are caused by activating mutations in *STAT3*
B. Autosomal dominant forms are caused by loss-of-function mutations in *DOCK8*
C. Diagnosis is based on histopathologic findings
D. Eosinophilic folliculitis may be the presenting feature in young infants
E. Unlike patients with Wiskott-Aldrich syndrome, those with Hyper IgE syndrome often have platelet dysfunction

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D. **Eosinophilic folliculitis may be the presenting feature in young infants**
E. Unlike patients with Wiskott-Aldrich syndrome, those with Hyper IgE syndrome often have platelet dysfunction

Explanation: Eosinophilic folliculitis may be the presenting feature of Hyper IgE syndrome in young infants (D). Both sporadic and autosomal dominant forms of Hyper IgE syndrome are caused by dominant negative mutations in *STAT3*, while loss of function mutations in *DOCK8* have been implicated in the pathogenesis of the less common autosomal recessive form. Patients do not typically present with platelet dysfunction, and diagnosis is based on clinical presentation, laboratory abnormalities including eosinophilia and elevated serum IgE levels, and/or detection of associated gene mutation.

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Case 8

Joshua Prenner, MD
Xiaolong Alan Zhou, MD, MSc

A 45-year-old male presents with a pruritic maculopapular eruption four weeks after orthotopic liver transplantation. Punch biopsy shows a vacuolar interface dermatitis with scattered necrotic keratinocytes. Which of the following is FALSE regarding this patient's condition?

A. As compared to patients with GVHD following hematopoietic stem cell transplant, this patient's prognosis is significantly worse
B. Skin biopsy typically demonstrates vacuolar interface dermatitis with scattered necrotic keratinocytes
C. Skin involvement is usually the first sign of disease and the patient may develop involvement of other organ systems
D. Treatment may include steroids and other steroid-sparing agents
E. The patient will likely develop elevated transaminases and hyperbilirubinemia

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E. **The patient will likely develop elevated transaminases and hyperbilirubinemia**

Explanation: The patient will not develop elevated transaminases or hyperbilirubinemia (E). Because the patient in question has solid-organ GVHD following a liver transplant, the liver cannot be one of the organs affected by GVHD, since it is the graft. Other organs that may be affected include the skin, GI tract, and bone marrow.

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Case 9

Alyce Anderson, MD, PhD
Joan Guitart, MD

A 25-year-old female presents to dermatology clinic for evaluation of rapidly progressive pustular lesions and ulcerated papulonodules, some with crusting. She notes recent fevers and myalgias. A RPR titer is noted to be highly positive. Which of the following underlying conditions is she most likely to have?

A. Chronic kidney disease
B. Chronic pulmonary disease
C. Hypertension
D. Type II diabetes mellitus
E. Untreated HIV infection

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B. Chronic pulmonary disease
C. Hypertension
D. Type II diabetes mellitus
E. **Untreated HIV infection**

Explanation: This patient has rupioid syphilis, a rare, malignant form of syphilis characterized by rapid progression of pleomorphic round papules, papulopustules with lamellated encrustations. It is most commonly described in association with HIV infection (E).

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Case 10

Edward Li, MD, PhD
Lida Zheng, MD

Which of the following is the classic triad of cutaneous lesions seen with Birt-Hogg-Dubé syndrome?

A. Fibrofolliculomas, pulmonary bulla, chromophobe renal cell carcinoma
B. Fibrofolliculomas, pulmonary bulla, clear cell renal cell carcinoma
C. Fibrofolliculomas, small cell lung cancer, benign renal masses
D. Fibrous papules, pulmonary bulla, clear cell renal cell carcinoma
E. Trichilemmomas, pulmonary bulla, benign renal masses

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Explanation: Birt-Hogg-Dubé syndrome is a rare inherited disorder characterized by multiple benign skin lesions (fibrofolliculomas, trichodiscomas, and acrochordons), pulmonary bulla/cysts, and renal neoplasia (hybrid oncocytic tumor and chromophobe renal cell carcinoma). Choice A is therefore the correct answer.