Great Cases and Great Discussion

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CASE 1

- 43-year-old African American man; 7-month history of recurrent tender skin lesions of face, trunk, and extremities
- Small brown crusted papules; lesions resolve and new ones appear
- Laboratory: Eosinophilia of 2.62 K/ml (normal 0-0.7 K/ml).
- Infectious work up negative
What is the diagnosis?
Lymphomatoid Papulosis

*But*…

Clinical Course

- Patient had persistent eosinophilia up to 5.58 K/ml; developed new LyP skin lesions
- FISH of peripheral blood demonstrated fusion of *FIP1L1*-platelet-derived growth factor receptor-alpha (*PDGFRA*) [4q12; *CHIC2* deletion] gene
- Bone marrow biopsy showed hypercellularity with increased eosinophils with atypical features
- Cytogenetic analysis of bone marrow demonstrated *FIP1L1*-*PDGFRA* fusion gene
- Treatment with imatinib (Gleevec) 400mg PO daily; no new LyP lesions have developed
**FIP1L1-PDGFRA Fusion Gene**

- Factor Interacting with PAPOLA and CPSF1
  - PAPOLA: Poly(A) polymerase alpha
  - CPSF 1: Cleavage and polyadenylation specificity factor subunit 1
- Chromosomal deletion on 4q12; fusion of genes FIP1L1 and PDGFRA (platelet-derived growth factor receptor, alpha)
- Constitutively activated tyrosine kinase; cells multiply
- Causes chronic eosinophilic leukemia responsive to treatment with tyrosine kinase inhibitors
Discussion

- Association between LyP and chronic eosinophilic leukemia with the *FIP1L1-PDGFRα* fusion gene
- LyP associated with other hematolymphoid conditions: Hodgkin lymphoma, anaplastic large cell lymphoma, MF and hypereosinophilic syndrome
- Fourth report of *FIP1L1-PDGFRα* rearrangement gene occurring with LyP
- Also seen with systemic mastocytosis
- Important to remember; rare association with LyP treatable with Gleevec

CASE 1
Diagnosis

*FIP1L1-PDGFRα*-associated Chronic Eosinophilic Leukemia and Lymphomatoid Papulosis
Which of the following disorders has not been associated with lymphomatoid papulosis?

a. Hodgkin’s disease  
b. Large cell anaplastic lymphoma  
c. Hypereosinophilic syndrome  
d. Mycosis fungoides  
e. Post-transplant lymphoproliferative disorder
CASE 2

Case History

• 10 year-old Hispanic female presented gradually enlarging asymptomatic “pimple” on her nose for 6 weeks
• Medical history unremarkable
• 1 cm solitary red-to-violaceous firm non-tender papule with irregular surface and telangiectases present on left alar rim
Similar Case
Diagnosis?
NEUROTHEKEOMA

Greek *theke,* "sheath"

- Age range 15 months - 84 years
  mean = 28 years
- Male: Female = 1:2
- Body site distribution: UE 34%, head/neck 29%, trunk 17%, LE 10%
- Average diameter 1.2 cm
- Two forms:
  - Traditional with neural morphology and mucin
  - Cellular
Cellular Neurothekeoma

- Head and neck region; adolescents and young adults
- May be confused with Spitz nevus or melanoma; may have atypical cells with mitoses
- S100 negative; NKI-C3 (CD 63) +
- Differentiation controversial; nerve sheath (Schwann cell), perineural fibroblast
- Fascicles and nests of plump spindle and epithelioid cells; lobulated growth pattern
- Symmetrical; spares epidermis
- Cells with abundant amphophilic cytoplasm; ill-defined cell borders
- Relatively rare; must think of in proper setting
CASE 2
Diagnosis
Cellular Neurothekeoma

Which of the following is true regarding cellular neurothekeoma?

a. The lesion is comprised of epithlioid cells arranged in nests
b. It may histologically simulate melanoma
c. Special stains for S-100 protein are negative
d. It was previously thought to be of muscle differentiation
e. All of the above
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CASE 3
Clinical History

- Two-year-old girl; African American and Sioux Indian heritage; referred for erythrokeratoderma variabilis (EKV)
- History: global developmental delay, hypotonia, hyporeflexia, asthma, right eye cataract
- Prenatal history: untreated maternal Graves’ disease; neonatal jaundice
- Generalized xerosis, accentuated flexural skin markings, circinate desquamation of the trunk and extremities
- Unusual facial features, truncal dermal melanocytosis; sacral dimples.
- Slightly elevated acylcarnitine levels; normal karyotype, thyroid function, electromyography studies, and serum amino acids
Histology

- Diffuse infiltrate of dermal foamy macrophages
- PAS negative; CD68 +; CD1a, CD117 –
- Peripheral blood smear: lipid droplets in circulating granulocytes and monocytes
- Electron microscopy: non-membrane bound, cytoplasmic lipid vacuoles in macrophages, fibroblasts, mast cells, keratinocytes and circulating granulocytes and monocytes
Peripheral Blood smear

Keratinocyte Electron Microscopy
Diagnosis?

CASE 3
Diagnosis

Neutral Lipid Storage Disease with Ichthyosis
(Chanarin-Dorfman Syndrome)
Chanarin-Dorfman Syndrome

- Rare; autosomal recessive
- Abnormal accumulation of neutral lipids in skin, granulocytes and monocytes, central nervous system, muscle, gastrointestinal tract and kidneys
- Genetic defect at ABHD5 (chromosome 3p21.33): lipid droplet associated protein; activator of lipolysis and mobilization of intracellular fat stores
- Clinical features: congenital non-bullous ichthyosis with EKV-like findings, myopathy, cataracts, sensorineural deafness and global developmental delay

Chanarin-Dorfman Syndrome

- Diagnosis confirmed by lipid droplets within granulocytes and monocytes on peripheral blood smear (Jordan’s anomaly)
- Course variable; some die due to multi-organ failure; others survive into adulthood
- Emollients, keratolytics, and systemic retinoids improve ichthyosis
- Dietary restriction of fat may improve clinical outcome
Which of the following is true regarding Chanarin-Dorfman syndrome?

a. It is a mucopolysaccharidosis
b. It is inherited in autosomal dominant fashion
c. Jordan’s anomaly, lipid within monocytes and granulocytes, is diagnostic
d. All patients die within a few years of diagnosis
e. None of the above
Case 4

A 27-year-old white woman presented with a left elbow mass. At 12 years old, she developed unexplained bilateral cataracts. At 15 years, she developed enlarging tumors on the Achilles tendons. At 20 years, lesions developed on tibial tuberosities and extensor surface of the hands. At 27, when she presented, she had developed headaches, mild dementia and unsteady gait. She has a sister with similar symptoms.

Physical Examination

- Multiple firm nodules on extensor surface of right hand, elbows, knees; largest on Achilles tendons
- Poor attention, concentration and impaired memory, mild spasticity of lower extremities, diffuse hyporeflexia, positive Babinski’s sign, wide-based ataxic gait
- EEG abnormal, with bursts of high voltage slow waves
- MRI of brain: marked cerebellar enlargement
Slow wave bursts
Histopathology

- Dense, diffuse, infiltrate of histiocytes in dermis and superficial subcutis
- Some multinucleated and replete with foamy cytoplasm
- Prominent cholesterol clefts
Laboratory Data

- CBC, Chem 20, LFT's: normal
- Lipids: Cholesterol 206, Triglycerides 224, LDL 133
- Other initially ordered studies WNL
Is there any other test you would order?

Dermatopathologist Ordered:

- **Cholestanol** 4.09 -- 30 times normal (nl: 0.2 mg/dl)
- Urine capillary gas chromatography positive for increased bile alcohol glucuronides (bile acid precursors)
Diagnosis:
Cerebrotendinous Xanthomatosis

Clinical Features

- Autosomal recessive; mutation in CYP27A1 gene located on chromosome 2q33-qter
  - Cytochrome P450, Family 27, Subfamily A, Polypeptide 1
  - Involved in bile acid biosynthesis
- Xanthomata, cataracts and progressive CNS abnormalities beginning in adolescence and young adulthood
- Due to deposition of *cholestanol* in brain, liver and skin
- Eventually fatal if not treated; stigmata reversible with timely treatment so important to recognize
- Kudos to Dr. Alun Wang at Tulane who made the diagnosis
Pathophysiology

- Decreased 26-hydroxylation of cholestane diol
- Decreased levels of chenodeoxycholic acid which is a negative regulator
- Lack of negative feedback on HMG CoA reductase and 7-hydroxylation
- Increased metabolites converted to cholestanol; deposit in tissue

Cerebrotendinous Xanthomatosis

- Diagnosis made by measuring plasma and urine cholestanol.
- Treatment with chenodeoxycholic acid life saving. Without treatment neurologic dysfunction and death will occur.
  - CDA not FDA approved at the time; had to be obtained from Germany. Still difficult to get
- Patient and sister are on CDA and have responded well. EEG’s now stabilized and blood and urine levels of cholestanol have decreased.
Cerebrotendinous Xanthomatosis

*Important Points*

- Disease rare but think about when faced with individual with tendinous or tuberous xanthomas
- If not recognized, severe and possibly irreversible sequelae may ensue
- You, the dermatologist or dermatopathologist, can be a hero and come to the rescue!

Which of the following is true regarding cerebrotendinous xanthomatosis?

a. Lipid is deposited in the brain and liver  
b. The diagnosis may not be apparent for many years  
c. Chenodeoxycholic acid administration may be lifesaving  
d. Tendinous xanthomas are characteristic  
e. All of the above
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Case History

- 41 year old Caucasian man with multiple ulcers, skin thickening and loss of eyebrows
- Ulcers began in 2009 on knees reportedly following loss of sensation following back injury in 2003
- Noted skin thickening with generalized loss of skin sensation on abdomen and loss of eyebrows three years ago
- Examination revealed thin yellow plaques on both cheeks; leonine facies, saddle nose deformity, bilateral loss of eyebrows, slate gray color of skin of face, infiltrated plaques on nose with telangiectasias
- Subcutaneous nodules of arms, chest and abdomen
- Angulated ulcers with granulation tissue on knees
Histology

• Diffuse infiltrate of epithelioid histiocytes in the dermis
• Some elongated granulomas
• Histiocytes with abundant foamy cytoplasm
What other tests would you do?
Diagnosis

Hansen’s Disease
Lepromatous Form with Features of Lucio’s Phenomenon

Lucio’s Phenomenon

• Severe cutaneous necrosis reaction; occurs in lepromatous Hansen’s patients either untreated or inadequately treated
• Infiltrated areas evolve with central necrosis and subsequent ulceration
• Thrombo-occlusive process due to bacterial liposaccharides causing macrophages to release TNF and IL-1; endothelial cells produce prostaglandins, IL-6, coagulation factor III with thrombi and secondary necrosis
• ENL: immune complex mediated LCV
• This case suggests direct vascular compromise secondary to extensive bacterial infection
Which of the following is true regarding diffuse lepromatous Hansen’s disease?

a. The condition may histologically resemble a xanthoma  
b. Ulcerations may develop  
c. Patients may develop a “La Bonita” form due to infiltration and diminution of normal rhytides  
d. Lucio’s phenomenon is due to a thrombotic process  
e. All of the above
Case History

- 45 year old Guatemalan man presented to charity health clinic for longstanding verrucous lesions of lower legs
- Process had been gradually increasing in size and extent
- History of working in construction where he had to stand in brackish water while digging trenches
- Otherwise healthy
- An incisional biopsy was performed
What is your diagnosis? What animal other than man is affected?
Lobomycosis

- Also known as keloidal blastomycosis and Lobo’s disease
- Endemic in rural South and Central America; acquired by trauma
- Affects bottlenosed dolphins; has been reported in Florida
- Caused by *Lacazia loboii* (previously *Loboa loboii*)
- Keloidal, verrucous nodules, plaques, and tumors
- Fungus grows as globose cells connected by narrow neck forming branching chains
- Diffuse granulomatous inflammation with abundant histiocytes and giant cells
- Pseudoepitheliomatous hyperplasia and intraepidermal abscesses typically absent
- Fungus 7-14 μm (average 9-10 μm) in diameter
- Treatment surgical excision
Lobomycosis

Blastomycosis
Lobomycosis:

a. Is a disease found only in man
b. Is caused by a dimorphic fungus
c. Is endemic in Texas
d. Is best treated with oral antifungal agents
e. Is morphologically similar to blastomycosis
Case 7

Case History

• 25 year-old Honduran male with 4 – 6 month history of left nasal congestion and obstruction
• Had low-volume epistaxis approximately once a week over number of years
• Works as a painter and occasionally performs sandblasting work
Diagnosis?
Rhinosporidiosis

- Endemic to India and Sri Lanka; occ'l in South America, US
- Exposure when bathing in stagnant water where animals also bathe
- Pruritic papule evolves into erythematous polypoid mass in nasal mucosa; obstruction, discharge of pus, organisms, cysts, pseudocysts and mucus with white dots on erythematous background resulting in strawberry-like appearance
- *Rhinosporidium seeberi*: large, endosporulating organism; clusters with a fish parasite near the animal-fungal divergence known as “DRIP” clade (*Dermocystidium*, rosette agent, *Ichthyophonus*, and *Psorospermium*), now known as Mesomycetozoa
- Sporangia develop from individual spores; increase up to 300 micrometers; contain up to 16,000 spores
- PAS-positive, GMS staining organisms >100 micrometers
- Histologic ddx: Cocci: much smaller spherules
- Wide surgical excision treatment of choice
**Rhinocerebral Rhinosporidiosis**

- *Rhinosporidium seeberi*: large, endosporulating organism
- Clusters with a fish parasite near the animal-fungal divergence known as “DRIP” clade (*Dermocystidium*, rosette agent, *Ichthyophonus*, and *Psorospermium*), now known as *Mesomycetozoa*
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- PAS-positive, GMS staining organisms >100 micrometers
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- Wide surgical excision treatment of choice
Rhinosporidiosis:

a. Is seen most commonly in India and Sri Lanka
b. Is caused by an organism of the DRIP clade
c. Is manifest by the development of strawberry-like lesions
d. Organisms highlighted with PAS and GMS stains
e. All of the above
Rhinosporidiosis:

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CASE 9
Clinical History

- 69-year-old Caucasian female; history of non-Hodgkin's B cell lymphoma in remission
- One-year history of episodic blistering on face, upper extremities, elbows
- Lesions begin as "whelps" and progress to tense, painful blisters within hours
- Direct immunofluorescence negative
- At time of development of lesions, stage IV low-grade nodal marginal zone B-cell non-Hodgkin's lymphoma diagnosed
- Clinically, 1x1 cm tense blister with surrounding erythema and focal crusting on forearm; several red papules and inflamed blister with a red and purpuric base on medial left lower leg
Histology

- Dense nodular mixed infiltrate of lymphocytes, abundant eosinophils
- Confluent epidermal necrosis; massive ballooning degeneration and reticular alteration
- *In situ* hybridization studies for Epstein-Barr virus (EBV) negative
- Immunohistochemical stains: CD3+ and CD4+ T cells; occasional CD20 and CD79a B cells; CD56 stain negative
Diagnosis?

Diagnosis

Exaggerated Insect Bite-like Reaction in association with Nodal Marginal Zone B-cell Non-Hodgkin's Lymphoma
Discussion

• Hypersensitivity to mosquito bites, hydroa vacciniforme-like papulovesicular eruption and paraneoplastic or non-specific immunobullous reactions all reported in association with hematologic neoplasms
• “Insect bite-like reaction" coined due to majority of patients not recalling insect bites
• Most commonly with CLL
• Pathogenesis unclear; may be seen with chronic EBV infection
• May respond to prednisone, dapsone and chemotherapy to treat underlying hematologic disorder
• Do not confuse with immunobullous disease

Which of the following is true regarding exaggerated bullous arthropod assault reactions?

a. They are seen more commonly in immunocompromised individuals
b. Fire ant stings are common causes
c. Dapsone may be effective treatment
d. Vasculitis is seen histologically
e. Both A and C
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Case History

• 71 year old Hispanic male with fever, hypotension
• History of end stage liver disease and diabetes
• Process developed after ingesting raw oysters
• Bilateral lower extremity edema and dusky violaceous patches with scattered hemorrhagic bullae
• Laboratory Data: thrombocytopenia, renal failure, and mild coagulopathy
Diagnosis?

*Vibrio vulnificus* Infection
Case Discussion

• *Vibrio vulnificus*: Gram negative bacterium
• Causes rapidly progressing life threatening infections; associated with raw seafood consumption or direct inoculation with contaminated marine environment
• May present as septicemia or wound infection
• Mortality rate >50%
• Often seen in patients with pre-existing liver disease, alcoholism, other underlying chronic illnesses or immunosuppression
• Over 90% of patients have cutaneous findings
  – Vesicles, bullae, edema, gangrene, cyanosis, purpura, macules and patches, papules, wheals and/or pustules
• Important to remember and not confuse with immunobullous disease

*Vibrio vulnificus* infection:

a. Is caused by an organism related to the cause of cholera
b. May cause widespread bullous eruption
c. Associated with raw seafood ingestion
d. Acquisition of infection may be hindered by horseradish and pepper sauce
e. All of the above
Vibrio vulnificus infection:

a. Is caused by an organism related to the cause of cholera
b. May cause widespread bullous eruption
c. Associated with raw seafood ingestion
d. Acquisition of infection may be hindered by horseradish and pepper sauce
e. All of the above
• 40 year-old male with three week history of asymptomatic eruption involving the trunk and extremities
• Denied associated symptoms except for a mild headache
• Numerous 3-4 mm pink, slightly indurated papules without surface change, diffusely present on the neck, trunk and extremities. Face, palms and soles were spared.
What is your diagnosis? What other studies might you order?

Additional Data

• Laboratory: Reactive RPR with a titer of 1:512 and a reactive confirmatory FTA-Abs
• Immunostain for spirochetes positive (two biopsy specimens)
Diagnosis

Secondary Syphilis
Take Home Points: Syphilis

- Histopathologic diagnosis requires good clinical information
- Both clinical and histologic features can be subtle
- Do not rely on abundant plasma cells for the diagnosis
- Immunostains for spirochetes are much more reliable than traditional silver stains

Secondary syphilis:

- Histologically may demonstrate a sparse perivascular dermatitis with few plasma cells
- Is best diagnosed using the Warthin-Starry silver stain
- Is declining in incidence
- Other forms include rupial and lues maligna
- A and D
Secondary syphilis:

a. Histologically may demonstrate a sparse perivascular dermatitis with few plasma cells
b. Is best diagnosed using the Warthin-Starry silver stain
c. Is declining in incidence
d. Other forms include rupial and lues maligna
e. A and D
Case Discussion

• 57 year old Hispanic male with pulmonary lymphomatoid granulomatosis treated with Rituxan and EPOCH (etoposide, vincristine, doxorubicin, prednisone, and cyclophosphamide)

• Extremely tender, violaceous plaque of right malar area; lesion began as small pink, crusted papules that coalesced

• Had recently been intubated with tape used to keep endotracheal tube in place
Diagnosis?
Diagnosis

Invasive Fusarium and Aspergillus Infection in a Patient with Lymphomatoid Granulomatosis

Case Discussion

- Neutropenia primary risk factor
- Other factors: trauma with intravenous catheters, tape, occlusive dressings, chronic administration of corticosteroids and cutaneous graft-versus-host-disease
- Aspergillus, Candida, and Fusarium species most common causes of disseminated fungal infections with cutaneous manifestations in cancer patients
- Fusarium fungemia more likely to present with skin lesions than other fungal pathogens
- More than one fungal species may cause disease
- High mortality rate: Fusarium 80%; Aspergillus 75%
- Early diagnosis essential
Fusarium vs Aspergillus

- Aspergillus: Hyphae branch at 45°
- Fusarium: random branching, some at 90°
- Generally requires culture
- Unusual to get more than one pathogen in same skin lesion

Invasive opportunistic fungal infections:

a. Most commonly caused by *Fusarium*, *Candida* and *Aspergillus*
b. Neutropenia most important risk factor
c. *Fusarium* most likely to present with skin lesions
d. 45 degree branching characteristic of *Aspergillus*
e. All of the above
Invasive opportunistic fungal infections:

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b. Neutropenia most important risk factor
c. *Fusarium* most likely to present with skin lesions
d. 45 degree branching characteristic of *Aspergillus*
e. All of the above

CASE 13
• 42-year-old African-American male; 7-month history of asymptomatic eruption of bilateral extensor forearms and dorsal hands
• 1-3mm firm, ivory, waxy papules without scale or erythema; similar linearly-arranged papules on the flank
• Laboratory data: negative HIV, hepatitis C; UPEP/SPEP equivocal gamma-globulin of 1.66 mg/L (reference range 0.50 - 1.50mg/L) with M spike of 1.16mg/L and IgG-κ light chain of 34.1 mg/L (reference range, 3.3-19.4 mg/L)
Colloidal iron staining
Diagnosis?

Diagnosis

Atypical Lichen Myxedematous
Localized Atypical Lichen Myxedematosus

- Several variants: acral persistent papular mucinosis, discrete papular lichen myxedematosus, juvenile and adult self-healing papular mucinosis, papular mucinosis of infancy, nodular lichen myxedematosus
- Acral persistent papular mucinosis: hands and extensor distal forearms; more common in females
- Discrete papular lichen myxedematosus: symmetrical eruption of proximal limbs and trunk associated with HIV
- Etiology unclear; mucin comprised of acid glycosaminoglycans; thought to be reactive

Localized Atypical Lichen Myxedematosus

- Histologically triad of mucin deposition, fibroblast proliferation, and fibrosis
- Localized atypical forms have no progression to scleromyxedema; this case unusual as it had paraproteinemia – Paraproteinemia strongly associated with scleromyxedema
- Scleromyxedema: progressive, disabling often fatal course
- Localized lichen myxedematosus: good prognosis, does not require treatment
Which of the following is true regarding lichen myxedematosus?

a. All forms are associated with paraproteinemia
b. All forms eventuate into scleromyxedema
c. Some forms have been associated with HIV infection
d. The mucin is similar to that seen in follicular mucinosis
e. The histologic differential diagnosis includes follicular mucinosis
CASE 14

Case History

- 59 year-old woman presented in January, 2010 with acute onset several tender papules on the digits of hand
- Past history significant for excision of malignant melanoma in situ of right index finger 2 months prior to presentation
- Several small, tender red to violet papules of dorsal and palmar aspects of right index and long fingers
- Papillary dermal edema with superficial and deep perivascular and periadnexal lymphoid infiltrate particularly concentrated around eccrine sweat glands
Diagnosis?

Diagnosis

Pernio (Chilblains)
Pernio (Chilblains)

- Painful or pruritic acral lesions
- Present 12-24 hrs after cold exposure
- Usually resolve in 2-3 weeks
- Some cases associated with lupus*
- Usually seen in Winter months but may be seen in warmer months if patient has poor circulation (Raynaud’s) or handles cold objects (works in cold environment)


Perniosis:

- Commonly demonstrates lymphocytic vasculitis histologically
- Seen exclusively in winter months
- Demonstrates a mixed infiltrate with abundant eosinophils
- May be associated with cryofibrinogenemia
- None of the above
Perniosis:

a. Commonly demonstrates lymphocytic vasculitis histologically
b. Seen exclusively in winter months
c. Demonstrates a mixed infiltrate with abundant eosinophils
d. May be associated with cryofibrinogenemia
e. None of the above
Case Discussion

• 65-year-old Caucasian female; 15-year history of non-pitting, non-pruritic, recurrent bilateral upper lid swelling, R>L
• Diagnosed with rosacea and eyelid “allergies”
• Mild right “mechanical” upper lid ptosis; bilateral upper lid swelling
• Brawny induration; no pain, pruritus or sensory deficit
Diagnosis?

Diagnosis

Melkersson-Rosenthal Syndrome
Case Discussion

- Unknown etiology
- May be confused with angioedema or thyroid-associated ophthalmopathy.
- May exist as triad of facial palsy, facial edema and furrowed tongue
- Most cases perioral but may be periorcular
- Histologically sarcoidal granulomas adjacent to dilated lymphatic vessels in edematous stroma classic
- Histiocytes may be seen within lymphatics as in this case
- DDx includes sarcoidosis and cutaneous Crohn’s disease

Which of the following is true regarding Melkersson-Rosenthal disease?

a. Scrotal tongue is required to make the diagnosis
b. Histologically may simulate sarcoidosis and cutaneous Crohn’s disease
c. May show granulomatous vasculitis
d. May clinically simulate angioedema
e. Both B and D
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August 2016 Weather Forecast

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Who Should Attend?

• Dermatologists
• Pathologists
• Dermatopathologists
• Dermatology residents (discounted rates)
• Physician assistants and extenders
• Spouses and family!

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Contact Me for Questions

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- www.dermpath.com