M. Yadira Hurley, MD
Professor Dermatology and Pathology
Department of Dermatology
Director of Dermatopathology
Saint Louis University

Abbott Laboratories – I(Grants/Research Funding); AbbVie – I(Grants/Research Funding); Actelion – I(Grants/Research Funding), SP(H); Amgen – I(Grants/Research Funding); Anacor Pharmaceuticals, Inc. – O(Grants/Research Funding); AstraZeneca – I(Grants/Research Funding); Centocor Ortho Biotech Inc. – I(Grants/Research Funding); Genentech, Inc. – I(Grants/Research Funding); Pfizer Inc. – I(Grants/Research Funding); Regeneron – I(Grants/Research Funding); XOMA (US) LLC – I(Grants/Research Funding);
The most likely diagnosis is:

A. Angiosarcoma
B. Basal cell carcinoma
C. Cutaneous B-cell lymphoma
D. Lymphomatoid Papulosis
E. Mycosis fungoides
The most likely diagnosis is:

A. Angiosarcoma
B. Basal cell carcinoma
C. **Cutaneous B-cell lymphoma**
D. Lymphomatoid Papulosis
E. Mycosis fungoides
Cutaneous B-cell Lymphoma

- Majority of cutaneous lymphomas are T cell lymphomas, but B-cell lymphomas account for 20-30% of all cutaneous lymphomas.

- Classification of cutaneous lymphomas published by the World Health Organization (WHO) and the European Organization for Research and Treatment of Cancer (EORTC) (2005)

**WHO–EORTC CLASSIFICATION OF CUTANEOUS B-CELL LYMPHOMAS WITH PRIMARY CUTANEOUS MANIFESTATIONS**

- Primary cutaneous marginal zone B-cell lymphoma[*]
- Primary cutaneous follicle center lymphoma
- Primary cutaneous diffuse large B-cell lymphoma, leg type
- Primary cutaneous diffuse large B-cell lymphoma, other
- Intravascular large B-cell lymphoma

*Includes cases previously designated as primary cutaneous immunocytoma and primary cutaneous plasmacytoma.
Primary Cutaneous B-cell Lymphoma

- Primary Cutaneous B-cell Lymphoma:
  - Follicle center lymphoma and marginal zone B-cell lymphoma: indolent clinical behavior
  - Diffuse large B-cell lymphoma, leg type and diffuse large B-cell lymphoma, other: intermediate clinical behavior

- Follicle center lymphoma:
  - Most lesions arise on the head and neck followed by trunk
  - Usually single erythematous plaques or tumors that increase with size overtime
  - 95% 5 year survival
Angiosarcoma

- Blue to violaceous nodules or plaques on face and scalp of the elderly

*Journal of the American Academy of Dermatology 2005; 53:744-745*
*Basal cell carcinoma

- Papular nodular lesion with a pearly translucent edge
Lymphomatoid papulosis

- Crops of papules and nodules at different stages of development; become necrotic and ulcerated
*Mycosis Fungoides

- Patches and plaques with scale that can progress to tumors
The most likely diagnosis is:

A. Blastomycosis
B. Coccidiomycosis
C. Keratoacanthomas
D. Myiasis
E. Sporotrichosis
The most likely diagnosis is:

A. Blastomycosis
B. Coccidiomycosis
C. Keratoacanthomas
D. Myiasis
E. Sporotrichosis
Sporotrichosis

- *Sporothrix schenckii* (dematiaceous, dimorphic fungus)
- Lymphangitic, localized (fixed) and disseminated
  - Lymphangiectatic accounts for 75% of cases
    - Single ulcer or nodule followed by subcutaneous nodules along the course of local lymphatics
    - Upper limbs are the most common site of involvement
- Percutaneous implantation of infected vegetable matter
Systemic mycosis: Infection develops in lungs and followed by skin / other organs

- **Blastomycosis** (North American)
  
  *Blastomyces dermatitidis*
  
  1. Primary cutaneous: crusted verrucous nodule, may show lymphangitic spread
  2. Disseminated
     - Verrucous and granulomatous lesions with thick crust and pustules along advancing edge
  3. Pulmonary

- **Coccidioidomycosis**
  
  *Coccidioides immitis*
  
  1. Most commonly acute self-limited pulmonary infection
  2. Disseminated infection occurs especially in immunocompromised
     - Verrucous plaque usually on the face or pustular lesions
  3. Rarely lymphangitic nodules
Blastomycosis
Keratoacanthomas: dome shaped papule or nodule with central keratin plug

- Keratoacanthoma centrifugum marginatum (solitary)
- Multiple keratoacanthomas
  - Ferguson Smith type (OMIM132800): adolescence, heal leaving a scar
  - Eruptive Grzybowski type: multiple can be in the hundreds
Myiasis

- Furuncle like presentation that culminates in ulceration
- Infestation of live human tissues by the larvae of flies in the order Diptera
- Predilection for exposed surfaces (feet and forearms)

New onset pruritic eruption that is not associated with fever or lymphadenopathy. The most likely diagnosis is:

A. DRESS (drug rash with eosinophilia and systemic symptoms) syndrome
B. Fixed drug eruption
C. Morbilliform drug eruption
D. Toxic epidermal necrolysis
E. Urticaria
New onset pruritic eruption that is not associated with fever or lymphadenopathy. The most likely diagnosis is:

A. DRESS (drug rash with eosinophilia and systemic symptoms) syndrome
B. Fixed drug eruption
C. Morbilliform drug eruption
D. Toxic epidermal necrolysis
E. Urticaria
Morbilliform Drug Eruption

• ‘Drug exanthem,’ macules and papules in a confluent measles-like pattern
• Most common drug reaction involving the skin
• Begins on torso or head and neck and migrates peripherally
• Can occur with first exposure often 8-10 days into treatment and recurs within 24 hours of reexposure
  – Requires initial exposure for sensitization
• Resolves 1-2 weeks
Most common drug is antibiotics
- Beta-lactams and sulfa drugs

Reactions to drugs can be allergic or non allergic
- Four types of allergic reactions
  1. Type I: IgE mediated – urticaria, angioedema and anaphylaxis
  2. Type II: cytotoxic – drug-induced thrombocytopenia
  3. Type III: circulating immune complexes – LCV, serum sickness
  4. Type IV: delayed hypersensitivity – drug exanthems, ACD
**DRESS Syndrome**

- Drug-induced hypersensitivity syndrome
  - phenytoin hypersensitivity syndrome
- Morbilliform cutaneous eruption with fever, lymphadenopathy, hematologic abnormalities, and multiorgan manifestations
  - Facial edema
- Anticonvulsants and sulfonamides
- Lymphocyte activation, drug metabolic enzyme defects, eosinophilia, and human herpesvirus-6 reactivation
- Later onset and longer duration than other drug reactions, latent period of 2 to 6 wks
- May have significant multisystem involvement: hematologic, hepatic (10% mortality, renal, pulmonary, cardiac, neurologic, gastrointestinal, endocrine
*Fixed Drug Eruption*

- One or more tender red plaques that resolve with PIPA
- Genital or oral mucosa
- 1-8 hours of drug ingestion
- Recurs in same location with re-exposure
TEN

- Rapid onset of symmetric, painful eruption
- >30% BSA
- Peak at 3 days
- Positive Nikolsky sign
- Immune and metabolic pathogenesis
Urticaria

- Type 1: IgE mediated
- Angioedema is a deep variant with mucous membrane involvement
- May be precursor to anaphylaxis
- Requires sensitization and can occur within minutes of re challenge
The most likely diagnosis is:

A. Dyshidrotic dermatitis
B. Erythema multiforme
C. Scabies
D. Syphilis
E. Tinea corporis
The most likely diagnosis is:

A. Dyshidrotic dermatitis
B. Erythema multiforme
C. Scabies
D. Syphilis
E. Tinea corporis
Erythema multiforme

- Abrupt onset of targetoid lesions favoring acrofacial regions; remain fixed ≥7 days
  - Central dusky purpura, elevated edematous pale ring, and surrounding macular erythema
- EM minor – little/no mucosal involvement or systemic symptoms
- EM major – severe mucosal involvement and systemic symptoms
- Herpes simplex virus infection most common precipitator
- Histopathology: Individual necrotic keratinocytes under a basket-weave stratum corneum with vacuolar interface dermatitis
*Stevens-Johnson syndrome and *Toxic epidermal necrolysis

- **SJS/EM major** -> Mycoplasma infections and sulfonamides
  - Two of more mucosal surfaces with less than 10% TBSA
- **TEN** -> anticonvulsants and NSAIDS
  - >30% TBSA
  - Histopathology: Full thickness epidermal necrosis
Dyshidrotic dermatitis (eczema)/ pompholyx

- Small deep seated vesicles palms and fingers (sides)
- Mistaken pathogenesis: Hypersecretion of sweat with retention in the acrosyringium
- Pompholyx is a spongiotic dermatitis of acral skin that results in retention of vesicles

*Scabies: papulovesicular type
- Vesicles at end of very fine wary dark lines
- Interdigital folds, palms, wrists, nipples and male genitalia

*Syphilis: secondary (maculopapular or papulosquamous)
- Palms – involve the skin lines
- Characteristic palmar and plantar involvement are indurated yellow-red spots

*Tinea corporis: circular lesion with elevated boarder and central clearing, dry and scaly
*Scabies*
*Secondary syphilis
40 year old male with a history of onychomycosis presents with a new eruption. The most likely diagnosis is:

A. Erythema annulare centrifugum
B. Erythema migrans
C. Erythrokeratodermia variabilis
D. Ichthyosis linearis circumflexa
E. Tinea corporis
40 year old male with a history of onychomycosis presents with a new eruption. The most likely diagnosis is:

A. Erythema annulare centrifugum
B. Erythema migrans
C. Erythrokeratodermia variabilis
D. Ichthyosis linearis circumflexa
E. Tinea corporis
Figurate erythemas

- Erythema annulare centrifugum
  - Annular or polycyclic lesions that grows slowly with *trailing scale* at the inner boarder of annular erythema
    - Days-months; frequently idiopathic (dermatophyte infections)

- Erythema marginatum (rheumatica)
  - Migratory polycyclic eruption of acute rheumatic fever (no scale); associated with carditis; precedes arthritis

- Erythema gyratum repens

- Erythema migrans
*Erythema Gyratum Repens*

- Rapidly migratory concentric “wood-grain” rings
- Undulating wavy bands with trailing scale
- Paraneoplastic: lung carcinoma #1

*Journal of the American Academy of Dermatology 2006; 54:745-762*
*Erythema Migrans*

- **Borrelia species**

- Tick bite leaves a small red macule or papule, several days after the bite there is gradual expansion of the redness

- Advancing border is red to blue-red and free of scale
Erythrokeratodermia variabilis
- Erythematous patches with bizarre configurations that are sharply demarcated that change over time
- Red brown keratotic plaques, often polycyclic, fixed in location

Ichthyosis linearis circumflexa
- Serpiginous red plaques with double-edge scale
- Netherton syndrome (atopic, trichorrhexis invaginata)

*Tinea corporis
Erythrokeratodermia variabilis

This patient also has gastrointestinal bleeding. The most likely diagnosis is:

A. Cowden syndrome
B. Cutaneous Crohn’s disease
C. Hereditary hemorrhagic telangiectasia
D. Peutz-Jeghers syndrome
E. Pyostomatitis vegetans
This patient also has gastrointestinal bleeding. The most likely diagnosis is:

A. Cowden syndrome
B. Cutaneous Crohn’s disease
C. Hereditary hemorrhagic telangiectasia
D. Peutz-Jeghers syndrome
E. Pyostomatitis vegetans
Hereditary hemorrhagic telangiectasia

- Many ethnic groups with prevalence varying among populations
- Recurrent epistaxis from mucosal telangiectasia is a common presenting feature, often within the first two decades of life, frequently before cutaneous lesions
- Telangiectases are also located on the lips, oral mucosa, upper extremities, nail beds, and trunk
  - Cutaneous telangiectases rarely cause significant bleeding
- Gastrointestinal tract telangiectases and AVMs - significant bleeding in 16% of patients
- Pulmonary AVMs in 15% of patients leading to respiratory complaints and neurologic complications, including emboli and stroke
- Other neurologic findings include CNS AVMs resulting in migraines and seizures
- AVMs are also found in the liver
Crohn’s disease:

- Often contiguous with external mucous membranes (oral and perianal)
  - Labial swelling
  - Granulomatous erosion perianal
- Metastatic lesions
- Cobblestonning hard palate and tongue

Pyostomatitis vegetans:

- Similarities in associations to PG
- Characterized by pustules and ulcerations
- Treatment of the underlying IBD

Journal of the American Academy of Dermatology 2013 68, 211.e1-211.e33 DOI: (10.1016/j.jaad.2012.10.036)
*Peutz–Jeghers

- Dark brown to blue macules in the perioral or periocular areas and the buccal mucosa; hyperpigmented macules on the fingers
- Associated cancer: Duodenal, colon, breast, pancreas, stomach, small bowel, cervix, uterus, ovary, testes, and thyroid

*Cowden syndrome:

- Trichilemmomas, papillomatous papules, acral and plantar keratosis, lipomas, and mucocutaenous neuromas
- Associated cancer: Colon, thyroid, endometrial, and breast cancers and renal cell carcinoma
The most likely diagnosis is:

A. Hypertrophic lichen planus
B. Prurigo nodularis
C. Psoriasis
D. Tinea corporis
E. Stasis dermatitis
The most likely diagnosis is:

A. Hypertrophic lichen planus
B. Prurigo nodularis
C. Psoriasis
D. Tinea corporis
E. Stasis dermatitis
Lichen planus

- Common – pruritic, purple, polygonal papules with flexor distribution and koebnerization
- Wickham striae – fine reticulated network of white lines
  - Corresponds to areas of periadnexal hypergranulosis
- Nail changes in 5%
  - Pterygium formation is characteristic (trauma, connective tissue disorders): Proximal nail fold fuses with nail bed
  - Onychorrhexis, onycholysis, trachyonychia (20 nail dystrophy in children)
- Mucous membrane disease has many forms
  - can progress into squamous cell carcinoma
- Difficult to distinguish from lichenoid drug eruptions
  - Can have eosinophils (Gold, ACE inhibitors)
• 2/3 of patients will spontaneously remit in 1 -2 years
• Likely is an autoimmune reaction against antigens on lesional keratinocytes
  – Associated with Hepatitis C (geographic region and oral involvement)
  – Dental metals?!
• Variants:
  » actinic, acute, annular, atrophic, bullous, LP pemphigoides, hypertrophic, keratosis lichenoides chronica, linear, LP-LE, LP pigmentosus, erosive, vulvovaginal gingival syndrome
• LP, lichen planopilaris, lichenoid drug eruption
- **Prurigo nodularis**: firm papules or nodules with lichenification and excoriations
- **Psoriasis**: well demarcated plaques with silver-white scale
• *Tinea corporis: Latin word “gnawing worm or moth,” annular lesions with central clearing

• *Stasis dermatitis: papulosquamous plaques with pigment abnormality, lower legs
The most likely diagnosis is:

A. Leukocytoclastic vasculitis
B. Livedoid vasculopathy
C. Rocky mountain spotted fever
D. Traumatic injury
E. Stasis dermatitis
The most likely diagnosis is:

A. Leukocytoclastic vasculitis  
B. Livedoid vasculopathy  
C. Rocky Mountain spotted fever  
D. Traumatic injury  
E. Stasis dermatitis
Livedoid vasculopathy

- Livedoid vasculitis, atrophie blanch, PURPLE (painful purpuric ulcers with reticular pattern of the lower extremities)
  - No leukocytoclastic vasculitis present

- Focal painful, purpuric lesions of the lower extremities that ulcerate and heal with small, stellate, white scars ('atrophie blanche')
  - Atrophie blanche also occurs in patients with stasis dermatitis and ulceration

- Most cases have no underlying disease
  - Rule out primary or secondary hypercoagulable states

- Therapy: low-dose aspirin, nifedipine, pentoxifylline, heparin

- Histology: thickened blood vessel walls, thrombosis, hemorrhage
Leukocytoclastic vasculitis

- Palpable purpura
*Rocky mountain spotted fever

- Small red macules on ankles and wrists that become papular, petechial and hemorrhagic and spread to trunk

- Lymphocytic vasculitis -> leukocytoclastic vasculitis

Traumatic injury – ‘trauma induced skin disease’

*Stasis dermatitis
  - Papulosquamous plaques with hemosiderosis and purpura
The most likely diagnosis is:

A. Allergic contact dermatitis
B. Cutaneous lupus
C. Fixed drug eruption
D. Seborrheic dermatitis
E. Keratoacanthoma
The most likely diagnosis is:

A. Allergic contact dermatitis

B. Cutaneous lupus

C. Fixed drug eruption

D. Seborrheic dermatitis

E. Keratoacanthoma
Acute, subacute, discoid, panniculitis

Cutaneous Lupus
- Discoid lupus
- Hypertrophic lupus erythematous (LE)
- Tumid
- Lupus panniculitis
- Chilblain LE
- Subacute cutaneous LE (papulosquamous, annular)
- Drug induced
Discoid lupus

- Discoid lupus
  - Young adult women > men
  - Dull red macules, eventually develop scale, scarring, atrophy, and hyperpigmentation/hypopigmentation
  - Scalp, ears, nose
  - Scale has “carpet tack“
  - Can transform into squamous cell carcinoma
  - Generalized forms rare
  - >95% stay localized to skin
*Allergic contact dermatitis: erythema, vesicles, bullae, superficial scale, pruritic

*Fixed drug eruption

*Seborrheic dermatitis: Erythematous, greasy scaly patches that do not show atrophy, alopecia, dilated follicles or follicular plugs

*Keratoacanthoma: Crateriform tumor on actinically damaged skin
*Allergic contact dermatitis*
*Seborrheic dermatitis
*Keratoacanthoma*
This patient complains of blisters and pruritus. The most likely diagnosis is:

A. Bullous pemphigoid  
B. Cicatricial pemphigoid  
C. Dermatitis herpetiformis  
D. Pemphigus vulgaris  
E. Scabies
The most likely diagnosis is:

A. Bullous pemphigoid
B. Cicatricial pemphigoid
C. Dermatitis herpetiformis
D. Pemphigus vulgaris
E. Scabies
Bullous pemphigoid

- Tense bulla with red urticarial base
- Elderly population
- Subepidermal bulla with eosinophils
- BP230 and BP180 (hemidesmosome plaque)
- Linear IgG and C3 by direct immunofluorescence
- Therapy: topical steroids, tetracycline, immunosuppressants
- Oral involvement in 20%
Cicatricial pemphigoid

- Mucous membrane pemphigoid
- Predilection for oral and ocular mucous membranes with a tendency to scar
- Skin involved 25%
- Heterogenous group of diseases: BP180, type VII collagen, epiligrin (Laminin 5)
• Bullous tinea
• *Dermatitis herpetiformis
  – pruritic papules, vesicles, erosions
  – symmetric involvement of extensor surfaces, such as the elbows, knees, shoulders, and buttocks
  – intense pruritus
• *Pemphigus vulgaris
  – suprabasilar vesicle = flaccid, erosions
• *Scabies
30 year-old female with a history of oral erosions and a new onset rash. The most likely diagnosis is:

A. Linear IgA disease
B. Pemphigoid gestationis
C. Pemphigus erythematosus
d. Pemphigus foliaceous
e. Pemphigus vulgaris
30 year-old male with a history of oral erosions and a new onset rash. The most likely diagnosis is:

A. Linear IgA disease
B. Pemphigoid gestationis
C. Pemphigus erythematosus
D. Pemphigus foliaceous
E. Pemphigus vulgaris
Pemphigus vulgaris

- Flaccid bullae that form painful erosions
- Oral involvement occurs first in 60% of cases
- Nikolsky sign
- Prior to systemic corticosteroids mortality was high
- Suprabasilar bullae with acantholysis
- DIF: epidermal intercellular IgG and C3
- Antibody to Desmoglein 3
- Pemphigus vegetans – variant with vegetative plaques
Pemphigus vulgaris

Pemphigus vegetans
**Linear IgA disease**

- Discrete bullae that occur in clusters
  - Cluster of jewels
- Adults: trunk and limbs
- Child (chronic bullous disease of childhood): facial and perineal lesions
Pemphigoid gestationis

- Herpes gestationis
- Urticarial plaques within the periumbilical area which progress to a generalize tense, bullous eruption
- Aberrant MHC class II placental molecules homologous to BPAg2 stimulate maternal antibody production
Pemphigus erythematosus

- Erythematous scaly plaques
- Butterfly distribution
- Association with other autoimmune diseases
*Pemphigus foliaceous*

- Recurrent crops of vesicles that easily rupture
- Mucous membranes rarely involved
- Autoantibodies to Desmoglein 1
- Drug induced: penicillamine
- Fogo selvagem: endemic form
The most likely diagnosis is:

A. Allergic contact dermatitis
B. Granuloma annulare
C. Lichen planus
D. Polymorphous light eruption
E. Psoriasis
The most likely diagnosis is:

A. Allergic contact dermatitis
B. Granuloma annulare
C. Lichen planus
D. **Polymorphous light eruption**
E. Psoriasis
Polymorphous light eruption

- Most common form of photosensitivity
- Papular or erythematopapular
- Female > Male, ? Etiology, Juvenile spring eruption of the ear
- Occurs within 1-3 days after sun exposure (UVA and/or UVB induced)
- Spring
- 20% have + ANA; Screen and follow these patients for systemic lupus erythematosus
**Allergic contact dermatitis**
- Pruritic eczematous, scaly plaques accentuated by papulovesicles

**Granuloma annulare**
- Papules that spread peripherally with central involution
- Dermal process

**Lichen planus**

**Psoriasis**
- Sharply demarcated papulosquamous plaques
Granuloma annulare, generalized
The most likely diagnosis is:

A. Acute intermittent porphyria
B. Bullous pemphigoid
C. Pemphigus vulgaris
D. Porphyria cutanea tarda
E. Hand eczema
The most likely diagnosis is:

A. Acute intermittent porphyria
B. Bullous pemphigoid
C. Pemphigus vulgaris
D. Porphyria cutanea tarda
E. Hand eczema
Porphyria cutanea tarda

- Most common of the porphyrias
- Photosensitivity
- Bullae without inflammation, erosions, fragile skin, milia, hypertrichosis, hyperpigmentation
- Sclerodermoid changes can occur late
- Associations: liver disease, hepatitis C, alcoholism, hemochromatosis, HIV
- Uroporphyrinogen decarboxylase
Porphyria cutanea tarda

- Non-inflammatory subepidermal bullae
- PAS-positive thickening of blood vessel walls
- Fluorescence of urine
- 24-hour urine collection
  - Uroporphyrins >> Coproporphyrins
- Treatment: Phlebotomy, antimalarials
Acute intermittent porphyria

- No skin findings
- Neurological, psychiatric, and abdominal symptoms

*Bullous pemphigoid

- Tense bullae on an erythematous base

*Pemphigus vulgaris

- Flaccid vesicles and erosions, no milia
Hand eczema

- Pompholyx, dyshidrotic eczema
- Vesicular eruption of palms and soles
- Deep seated vesicles that lead to scaly patches
The most likely diagnosis is:

A. Acrodermatitis enteropathica
B. Necrolytic acral erythema
C. Necrolytic migratory erythema
D. Pellagra
E. Scurvy
The most likely diagnosis is:

A. Acrodermatitis enteropathica
B. Necrolytic acral erythema
C. Necrolytic migratory erythema
D. Pellagra
E. Scurvy
Necrolytic acral erythema

- Eroded, erythematous to violaceous patches, tender flaccid blisters and erosions which result in hyperkeratotic plaques in older lesions
  - Dorsal > volar surfaces
- Rare, Associated with Hepatitis C virus
- Histology resembles acrodermatitis enteropathica and necrolytic migratory erythema
- Zinc(?), fatty acids, glucagon levels normal
- Treatment of underlying hepatitis or hyperalimentation
Acrodermatitis enteropathica

- Autosomal recessive defect in zinc transporter protein ZIP4
- Infancy after weaning
- Alopecia, diarrhea and dermatitis
- Periorificial and acral eczematous eruption
*Necrolytic migratory erythema*

- Glucagonoma syndrome – glucagon secreting islet tumor of pancreas
- Waves of extending annular erythema and superficial epidermal necrosis leaving crusted erosions with resolution in 10 to 14 days
- Trunk, groin, perineum, thighs, and buttocks
- Histologically similar to other deficiency states (acrodermatitis enteropathica)
**Pellagra**

- Multisystem nutritional disorder
- Inadequate amounts of niacin (nicotinic acid)
- Dermatitis, diarrhea, dementia, death
- Burning erythema in sun-exposed areas followed by hyperpigmentation and epithelial desquamation
Scurvy

- Deficiency of vitamin C (ascorbic acid)
  - Formation of collagen
  - Normal hair growth
- Perifollicular hemorrhage, corkscrew hairs, bleeding gingivitis
The patient has a history of diabetes mellitus and complains of pruritic lesions. The most likely diagnosis is:

A. Calcinosis cutis
B. Calciphylaxis
C. Kyrle’s disease
D. Elastosis perforans serpiginosa
E. Folliculitis
The patient has a history of uncontrolled hypertension and complains of pruritic lesions for years. The most likely diagnosis is:

A. Calcinosis cutis
B. Calciphylaxis
C. Kyrle’s disease
D. Elastosis perforans serpiginosa
E. Folliculitis
Primary perforating dermatosis:
- Kyrle’s disease, perforating folliculitis and reactive perforating collagenosis
  - Dermal substances extruded through epithelium with little damage to surrounding structures

Classic KD is traditionally described in young to middle-aged adults and is often associated with diabetes mellitus
- 2- to 8-mm dome-shaped papules with a central keratotic plug, some coalescing into plaques
- most frequently distributed on the extremities.
- lesions may be follicular or extrafollicular
- linear arrangement is common.

Perforating disorders observed in ESRD (acquired perforating dermatosis) share features with the primary perforating dermatoses
*Calcinosis cutis:

- metastatic calcification: calcium deposition in normal subcutaneous and cutaneous tissues
- firm papules, plaques, and nodules
- chalky discharge may be extruded

*Calciphylaxis:

- firm, bilaterally symmetric, painful purpuric plaques or nodules, often with a reticulated pattern resembling livedo reticularis (retiform purpura)
- angulate or stellate ulcers with eschar or frank gangrene

Journal of the American Academy of Dermatology 2000 43, 975-986
DOI: (10.1067/mjd.2000.110651)
*Elastosis perforans serpiginosa:*

- neck, upper extremities, and face
- primary lesions manifests as slightly erythematous or flesh-colored keratotic papules in arcuate or serpiginous pattern

*Folliculitis:*

- Follicular based red papules and pustules
- Etiology: infection, occlusion, irritation, drugs
  - Steroid folliculitis

*Journal of the American Academy of Dermatology 2015 73, 891-893DOI: (10.1016/j.jaad.2013.03.010)*
The most likely diagnosis is:

A. Darier’s disease
B. Erythema ab igne
C. Lichen amyloidosis
D. Lichen planus
E. Pretibial myxedema
The most likely diagnosis is:

A. Darier’s disease
B. Erythema ab igne
C. Lichen amyloidosis
D. Lichen planus
E. Pretibial myxedema
Darier’s disease

- Autosomal dominant, mutations in ATP2A2 gene encoding endoplasmic reticulum Ca\(^{++}\) ATPase (SERCA2)
- **Keratotic, greasy papules**, seborrheic areas, nail changes (v-shaped nicks, red and white lines), oral mucosal white papules, acral keratoses
- Histology: acantholytic dyskeratosis, suprabasilar clefting, corps ronds and grains
Erythema ab igne
- Reticulate hyperpigmentation in livedo reticularis pattern from heat

Lichen amyloidosis
- Small discrete, pruritic waxy papules with predilection for extensor lower extremities
Lichen amyloid

Journal of the American Academy of Dermatology 1997; 37:923-928
*Lichen planus

*Pretibial myxedema
  • Sharply circumscribe nodular lesions, diffuse non-pitting edema, or elephantiasis-like thickening of the skin
  • Anterior aspect of lower legs with sparing of dorsum of feet
*Pretibial Myxedema

60 year old male presents with a persistent, pruritic eruption which involves the trunk and buttocks. The most likely diagnosis is:

A. Follicle center lymphoma
B. Lymphomatoid papulosis
C. Marginal zone B-cell lymphoma
D. Mycosis Fungoides
E. Psoriasis
60 year old male presents with a persistent, pruritic eruption which involves the trunk and buttocks. The most likely diagnosis is

A. Follicle center lymphoma
B. Lymphomatoid papulosis
C. Marginal zone B-cell lymphoma
D. Mycosis Fungoides
E. Psoriasis
Mycosis Fungoides

- Clinically and pathologically distinct form of cutaneous T-cell lymphoma
  - CD3+, CD4+, CD8-
- Progression from patches to plaques to tumors
- 50% of all primary cutaneous lymphomas
- Male predominance
- Trunks, thighs, breast of women
- Indolent course with slow progression
- Subtypes: folliculotrophic, pagetoid reticulosis, granulomatous slack skin
Mycosis Fungoides

Journal of the American Academy of Dermatology 2009; 60:359-375
**Follicle Center Lymphoma**

- Single erythematous plaque to tumor
- Most commonly on head and neck then trunk
Lymphomatoid Papulosis

- Crops of papules and nodules at different stages of development
- Become necrotic and ulcerated

*Marginal zone B-cell lymphoma

- Lesions occur predominantly on the arms and trunk but many other sites can be involved
- Frequently multifocal red to purple papules and nodules
*Psoriasis*

- Well-circumscribed erythematous patches with silvery white scale
The most likely diagnosis is:

A. Extragenital lichen sclerosus
B. Plaque morphea
C. Scleredema
D. Scleromyxedema
E. Systemic sclerosis
The most likely diagnosis is:

A. Extragenital lichen sclerosus
B. Plaque morphea
C. Scleredema
D. Scleromyxedema
E. Systemic sclerosis
Morphea (Localized scleroderma)

- Scleroderma is a broad term used to denote a subset of fibrosing disorders
- Morphea, also referred to as localized scleroderma
  - fibrosing condition that is limited to the skin, subcutaneous tissue, underlying bone, and rarely—when present on the face and head—the underlying central nervous system
- Systemic scleroderma, or systemic sclerosis, is a systemic fibrosing disorder
- Early lesions present as erythematous to dusky violaceous patches and plaques
- Resolve into sclerotic, hairless, anhidrotic plaques with varying PIPA
- Plaque morphea is the most common presentation in adults
- Linear morphea is the most common presentation in children and often presents with fibrosis of underlying tissues, resulting in additional morbidity
<table>
<thead>
<tr>
<th>Classification</th>
<th>Included subtypes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Plaque</td>
<td>Morphea en plaque, guttate, atrophoderma of Pasini and Pierini, keloidal, and lichen sclerosis et atrophicus</td>
</tr>
<tr>
<td>Generalized</td>
<td>Defined as involving 2 or more body areas</td>
</tr>
<tr>
<td>Bullous</td>
<td>Linear morphea of the extremities, en coup de sabre, and progressive facial hemiatrophy</td>
</tr>
<tr>
<td>Linear</td>
<td>Morphea profunda, subcutaneous morphea, eosinophilic fasciitis, and pansclerotic morphea</td>
</tr>
</tbody>
</table>
*Lichen sclerosus*

- Genital or extragenital

- Extragenital lichen sclerosus
  - most common on the neck, shoulders, and upper portion of the trunk
  - flat, white, polygonal papules, and slight atrophic white plaques

Scleromyxedema

- Generalized and sclerodermoid lichen myxedematosus, papular mucinosis
- Firm papules with areas of induration that are due to mucin deposition with an increase in dermal collagen
  - 2-3 mm firm waxy papules with a strikingly linear array
Scleredema

- Scleredema adultorum of Buschke
- Non-pitting induration of skin
- Posterior neck, shoulders, upper trunk and face
- Variants: post infection, insidious course, DM

Journal of the American Academy of Dermatology
Vol. 65, Issue 4, Pages e115-e117
Systemic sclerosis (Systemic scleroderma)

- **Limited cutaneous systemic sclerosis**
  - affects the distal extremities, causing sclerodactyly
  - associated with history of Raynaud phenomenon, telangiectasias, gastrointestinal involvement
  - risk of isolated pulmonary artery hypertension

- **Diffuse cutaneous systemic sclerosis**
  - proximal (above the knee and elbow) skin thickening
  - sclerodactyly
  - explosive onset of debilitating Raynaud phenomenon
  - telangiectasias and gastrointestinal involvement
  - risk of interstitial lung fibrosis and renal crisis.

*Journal of the American Academy of Dermatology 2011 65, 1-12 DOI: (10.1016/j.jaad.2010.08.038)*