Case 1

Diagnosis: Pretibial pruritic papular dermatitis (PPPD)

Clinical Features:

- Forty-four patients described by Annessi
- Pruritic, erythematous, smooth papules
- Thought to be a response to gentle, chronic rubbing, akin to dermal lichen simplex chronicus
- Chronic course, responds to topical steroids, but often recurs after stopping treatment

Microscopic Features:

- Mild orthokeratosis and mild acanthosis, flattening of rete ridge pattern
- Superficial to mid-dermal perivascular lymphocytic inflammation with variable numbers of histiocytes and eosinophils
- Superficial dermal fibrosis with multinucleated fibroblasts

Main differential diagnoses:

- Lichen simplex chronicus
- Lichen amyloidosis

Take Home Message:

PPPD is a distinctive clinical and histologic reaction to persistent rubbing of the skin

Reference:

Dermatoses you’ve probably seen but never heard of

Case 2

Anneli R Bowen, MD

Diagnosis: Insulin-derived nodular amyloidosis

Clinical Features:

- Hard, hyperpigmented nodule at site of insulin injection
- Longstanding insulin-dependent diabetic patients who don’t rotate insulin injection site
- Usually have high or increasing insulin requirements
- Erratic glucose control
- May be associated with acanthosis nigricans-like epidermal hyperplasia

Microscopic Features:

- Dermal nodules of hypereosinophilic, homogenous material that stains with Congo red, showing typical apple-green birefringence under polarized microscopy
- Infiltrate of plasma cells, the presumed source of amyloid deposits
- Amyloid deposit positive for insulin by immunohistochemistry

Take Home Message:

Insulin-derived nodular amyloidosis is a potential complication of insulin injection in diabetics that do not rotate insulin injection site and can result in poor glucose control

Reference:

Dermatoses you’ve probably seen but never heard of

Case 3

Diagnosis: Hyperkeratotic and lichenified dermatosis of the gluteal region (senile gluteal dermatosis)

Clinical Features:
- Common dermatosis of the elderly who spend most of the day sitting
- Thin, male:female 130:7
- Itching or pain of varying intensity, may be asymptomatic
- Brownish plaques on the gluteal cleft of the buttocks – ‘three corners of a triangle’
- Horizontal hyperkeratotic linear ridges a characteristic sign
- Treatment difficult

Microscopic Features:
- Hyperkeratosis
- Acanthosis
- Follicular plugging
- No amyloid deposits

Main differential diagnoses:
- Anosacral amyloidosis (lichen amyloid)
- Irritant/allergic contact dermatitis
- Lichen simplex chronicus
- Mycosis fungoides

Take Home Message:

This is a common dermatosis of the elderly that has received little attention

Reference:
Case 4

Diagnosis: Cutaneous collagenous vasculopathy

Clinical Features:

- Described in 2000 by Salama and Rosenthal
- Telangiectasias usually start in middle age on lower legs, progresses to become more generalized
- Male=female
- May darken over time
- No associated underlying disease, no history of hemorrhage
- Likely under recognized/confused with generalized essential telangiectasia

Microscopic Features:

- Dilated superficial vessels
- Thickened walls (reduplicated basement membrane material)
  - Seen well on PAS and immunohistochemical staining for Collagen IV
  - These characteristic changes can be subtle or focal

Main differential diagnoses:

- Without systemic implications
  - Generalized essential telangiectasia
  - Unilateral nevoid telangiectasia
  - Hereditary benign telangiectasia
- With systemic implications
  - Hereditary hemorrhagic telangiectasia
  - Ataxia telangiectasia
  - Liver disease
  - Increased estrogen states

Take Home Message:

- Benign, progressive, asymptomatic generalized telangiectasia associated with characteristic reduplication of basement membrane histologically

References:

Dermatoses you’ve probably seen but never heard of

Case 5

Diagnosis: Granulomatous pigmented purpuric dermatosis

Clinical Features:

- Uncommon variant of pigmented purpuric dermatosis
- Clinically, lesions resemble other pigmented purpuric dermatoses
- Like other pigmented purpuric dermatoses, most common on lower extremities but may involve trunk
- Tenuous association with hyperlipidemia - requires more study to confirm

Microscopic Features:

- Superficial and sometimes deep dermal perivascular lymphocytes and histiocytes with extravasated erythrocytes.
- May have siderophages

Main differential diagnoses:

- Other pigmented purpuric dermatoses
  - Schamberg’s purpura
  - Lichenoid purpura of Gougerot and Blum
  - Eczematoid purpura of Doucas and Kapetanakis
  - Lichen aureus
  - Purpura annularis telangiectodes
- Capillaritis-like mycosis fungoides

Take Home Message:

Granulomatous pigmented purpuric dermatosis is an uncommon variant of pigmented purpuric dermatoses- possibly associated with hyperlipidemia, may demonstrate deep dermal involvement histologically

Reference:

Dermatoses you’ve probably seen but never heard of  
Summer AAD 2017, New York, NY

Case 1

Diagnosis: Collagenous and elastotic marginal plaques of the hands

Other names: Keratoelastoidosis marginalis, degenerative collagenous plaques of the hands, digital papular calcific elastosis

Clinical Features:

- Five cases reported as ‘degenerative collagenous plaques of the hands’ in 1960
- Male predominance, mean age ~ 70 in a series of 20 patients, ranges from 40 – 80 years
- Bilateral, symmetrical linear plaques at the junction of the dorsal and palmar skin on medial thumb, extending to radial index finger
- Slow progression for many years before presentation, most patients asymptomatic with stiffness occasionally reported
- Etiology unknown, hypotheses include chronic actinic damage, chronic pressure, altered collagen and elastin

Microscopic Features:

- Hyperkeratosis and epidermal acanthosis with loss of rete ridge pattern
- Haphazard arrangement of thickened collagen bundles in the dermis, some oriented perpendicular to the epidermis
- Dermis is relatively acellular and avascular
- Fragmented elastic fibers
- Basophilic elastotic masses – rounded degenerative aggregates of elastic material that may calcify

Take Home Message:

The findings are distinctive clinically and histologically

References:

Dermatoses you’ve probably seen but never heard of

Case 2

Scott R Florell, MD

Diagnosis: Annular lichenoid dermatitis of youth

Clinical Features:

- Twenty-three cases described in 2003, ~ 50 cases described to date
- Lesions present mostly on the torso and groin
- Red macules to patches, become hypopigmented centrally leaving an annular ring with a brown edge
- A few cases have been described in adults
- Clinical differential diagnosis includes mycosis fungoides, morphea, vitiligo, other annular erythemas
- Etiology unknown - Austrian group found Borrelia spirochetes in 11/14 biopsies, suggested ALDY may be an early, superficial stage of morphea
- Treatment empiric, including topical corticosteroids, tacrolimus, phototherapy
- Usually chronic/recurrent clinical course

Microscopic Features:

- Lichenoid interface dermatitis
- Hyperplastic epidermis with thinned, sometimes quadrangular rete
- Exocytosis of lymphocytes
- Keratinocyte apoptosis
- Can be difficult to distinguish from mycosis fungoides; T-cell monoclonality has not been described in ALDY so T-cell gene rearrangement studies could be helpful in some cases

Main Differential Diagnoses:

- Mycosis fungoides
- Inflammatory vitiligo
- Morphea
- Annular erythema

Take Home Message:

Annular lichenoid dermatitis (of youth) shares clinical and histologic features with mycosis fungoides; remember this differential diagnostic consideration with truncal, round to annular clinical lesions with lichenoid histology

References:

Dermatoses you’ve probably seen but never heard of

Case 3

Diagnosis: Circumscribed palmar hypokeratosis

Clinical Features:

- Ten patients described by Perez in 2002
- ~70 cases described to date, mostly in middle age or elderly
- Most patients present with a solitary lesion on the right (dominant) palm, mostly thenar or hypothenar
- Etiology unknown, ? localized disorder of keratinization
- No specific treatment – typically prolonged course

Microscopic Features:

- Abrupt loss and thinning of the stratum corneum

Main Differential Diagnosis:

- Porokeratosis

Take Home Message:

The findings are distinctive clinically and histologically

References:

Dermatoses you’ve probably seen but never heard of

Case 4

Diagnosis: Demodex folliculorum-associated spinulosis

Clinical Features:

- Tiny follicular spicules involving face
- Predisposing factors include increased age, diabetes mellitus, hemodialysis and immunosuppression
- Several cases have been reported in the setting of polycythemia rubra vera and hydroxyurea therapy
- Treatment: topical permethrin, ivermectin, oral ivermectin

Microscopic Features:

- Follicular spicules composed of Demodex folliculorum mites with associated hyperkeratosis
- Minimal associated dermal inflammation

Treatment:

- Crotamiton topical
- Permethrin topical
- Ivermectin topical
- Ivermectin oral

Take Home Message:

*Demodex folliculorum*-associated spinulosus is an important entity to remember when evaluating a patient with facial follicular spicules because it is easy diagnosed and may be easily treatable

References:

Dermatoses you’ve probably seen but never heard of

Case 5

Scott R Florell, MD

**Diagnosis:** White fibrous papulosis of the neck (WFPN)

**Clinical Features:**

- Multiple asymptomatic whitish to yellowish papules on the lateral and posterior neck
- Men and women older than 40 years of age
- Etiology unknown, ? intrinsic cutaneous aging or chronic UV exposure
- No specific treatment

**Microscopic Features:**

- Routine H&E may look like normal skin, may show mild thickening of collagen bundles
- Decreased to absent dermal elastic tissue

**Main Differential Diagnoses:**

- Pseudoxanthoma elasticum-like papillary dermal elastolysis (PXE-PDE)
- Papillary dermal elastosis (PDE)
- Mid-dermal elastolysis
- Linear focal elastolysis

**Take Home Message:**

The findings are distinctive clinically and histologically. White fibrous papulosis of the neck may be part of a spectrum of similar disorders termed ‘fibroelastolytic papulosis,’ including WFPN, PXE-PDE, and PDE.

**References:**