Case 1  38 year old female

- History of melanoma and dysplastic nevi
- First presented to our clinic in 2006 with a several year history of a pruritic rash involving the arms, legs, chest, back and neck
- Flared several times a year without full resolution
- Clinical suspicion of lichenoid dermatitis
- BX #1: Bowenoid actinic keratosis

B. Hist, full thickness punch biopsy: Lesion-like granular epithelial pattern with prominent epithelial angio... inflammation and mild focus of epidermal spongiosis.

BX# 2 in 2006

Diagnosis?
BX# 6 in 2008

- Clinical: Lichenoid-like eruption, not improving, not confirmed by biopsy
- DDX: Lichenoid process.

BX# 6

- Skin, left arm: Impetiginized crust with areas of superficial epidermal acantholysis and sparse dyskeratosis. (See comment)
- **Comment:** The histologic differential diagnosis includes bullous impetigo or impetiginized Grover’s disease. The presence of sparse dyskeratotic cells militates against an acantholytic process such as pemphigus. There is no evidence of a lichenoid infiltrate.

**Diagnosis?**
BX# 9 in 2013

Clinical History

Erythematous pigmented macule, question CARP, right posterior neck. Biopsy.

Final Pathologic Diagnosis:

Skin, right posterior neck, shave biopsy: Lentigo-like epidermal pattern and some additional broader buds of epithelium.

Diagnosis?

Derm Group Conf 2015

- Skin, left arm: Impetiginized crust with areas of superficial epidermal acantholysis and sparse dyskeratosis.
- No family history and normal nails but... Darier’s???
Let's Review

Diagnosis?

Galli-Galli Disease

Galli-Galli disease is an acantholytic variant of Dowling-Degos disease: Additional genetic evidence in a German family

Dowling-Degos Disease
**Galli-Galli Disease**


**Reticulate Hyperpigmentary Disorders**

- Rare genetic pigmentary abnormalities which include:
  - Reticulate acropigmentation of Kitamura
  - Dowling-Degos disease
  - Galli-Galli disease

**Reticulate Hyperpigmentary Disorders**

- Reticulate acropigmentation of Kitamura
- Dowling-Degos disease
- Galli-Galli disease
- Dyschromatosis universalis hereditaria
- Dyschromatosis symmetrica hereditaria

Take Home

- Flash back
- Clinical Images
- Galli-Galli Disease
  - Darier’s like eruption and hyperpigmented macules
  - Adenoid proliferation of the rete with basal hyperpigmentation and acantholysis

Case 2 11 year old male
- Presented with 3 week history of draining lesions on the left lower leg
- No history of trauma (does have a dirt bike)
- Treated with I&D, clindamycin, and sulfamethoxazole-trimethoprim by PCP


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2 Weeks Later

Special stains for organisms and subsequent suggested tissue culture were negative

Diagnosis?
Pyoderma Gangrenosum?
- No bowel symptoms and endoscopy/colonoscopy negative
- Developed firm subcutaneous nodule on the left chest

Special Stains Negative

4 months from onset
- Admitted for IV solumedrol
- Work-up for autoimmune and systemic granulomatous disease

Diagnosis?
Diagnosis?

Autoimmune and Systemic Granulomatous Disease Work-up
- Microscopic hematuria
- Proteinuria

Granulomatosis with Polyangitis
- Aka Wegener’s granulomatous
- C-ANCA positive in 81% with cutaneous disease
- Most in men in their 4th decade
- Upper and lower airway
- Renal involvement
- Skin involvement in up to 45%
  » May be the first manifestation 15%

Pauci-influenmatory crescentic glomerulonephritis
- Anti-proteinase 3 ab 148.7
- C-ANCA pattern
Consider Wegener’s granulomatous especially with PG-like ulcers of unusual sites like face/neck/ear.

Pyoderma gangrenosum in kids isn’t just IBD
Granulomatosis with polyangitis
- Not just granulomatous vasculitis
- Suppurative/acneiform
- PNGD
- Churg-Strauss necrotizing extravascular granuloma (Winkelmann)

9 months later
Wound care, mycophenolate and prednisone
Thank You