Dowling-Degos with Acantholysis: A Rare Case of Galli-Galli Disease

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History of Presentation

- 75 year old previously healthy Caucasian female
- 35 year h/o of widespread hyperpigmented macules
  - first appeared on thighs
  - progressed to involve back, neck, and arms
- 2-3 year h/o pruritic, hyperkeratotic, tan-colored papules
  on the extensor and flexural surfaces of extremities, neck, trunk
History of Present Illness

- A previous biopsy done in 2008 had shown focal acantholytic dyskeratosis, and she had been given a diagnosis of Grover’s disease.

- Previous treatments included:
  - Triamcinolone 0.1% cream (transient improvement)
  - A several month course of tetracycline (no help)

Family History:

- Brother with similar papules limited to the lower extremities which appeared in middle age.

Past Medical History:

- Actinic keratoses
- Osteoarthritis
- HTN, hyperlipidemia
- Glaucoma
- Depression
- GERD

Medications:

- HCTZ
- Paroxetine
- Atorvastatin
- Esomeprazole
- Bimatoprost

Physical Exam:

- BP 154/94 HR 88 RR 14
Differential Diagnosis

- Darier’s disease
- Dowling-Degos disease
- EB Simplex with Mottled Pigmentation
- Disseminated superficial actinic porokeratosis
- Atypical Grover’s disease
Diagnosis:
Galli-Galli Disease

Galli-Galli Disease

- Originally reported by Bardach, Gebhart, and Luger in 1982, who described the disease in two brothers
- A rare acantholytic variant of Dowling-Degos Disease, an autosomal dominant genodermatosis
- Approximately 12 cases reported in the literature
Galli-Galli Disease: Clinical Findings

- M>F, age range 15-56 yrs
- Reddish-brown, hyperkeratotic, scaly, pruritic papules
- Confluent, reticulated hyperpigmented macules
- Involvement of trunk, neck, and both flexor and extensor surfaces of the extremities (vs. Dowling Degos)
- No perioral scars, palmar pits, or nail changes
- +/- Peripheral eosinophilia

Gilchrist 2008; Bardach 2001

Galli-Galli disease; A case report with review of the literature

Heidi Gilchrist, MD, Scott Jackson, MD, Lisa Morse, MD, and Lee T. Nesbitt, MD

Clinical DDx of Galli-Galli Disease

- **Dowling-Degos Disease - KRT5**
  - Reticulated hyperpigmentation mainly in flexures, perioral pitted scars, *comedo-like papules* (*‘dark dots’* )

- **EB Simplex with Mottled Pigmentation - KRT5**
  - Widespread reticulated hyperpigmentation, *skin fragility* (present at birth), blistering, progressive *palmoplantar hyperkeratosis*

- **Reticulate Acropigmentation of Kitamura - KRT5**
  - Acral reticulated hyperpigmentation, *palmar pits*, breaks in dermatoglyphs

- **Dyschromatosis Symmetrica Hereditaria - DSRAD**
  - Symmetrical reticulated *hyper- and hypopigmentation* of extremities, usually presents by age 6yo, patients usually *Asian*.

- **Darier’s Disease – ATP2A2**
  - Hyperpigmented, hyperkeratotic papules in *seborrheic distribution*, dorsal hands and *palms* involved, *dystrophic nails*, white papules on *palate*

Galli-Galli Disease: Histopathology

- **Epidermal acanthosis and hyperkeratosis**

- **Elongated epidermal rete ridges with bud-like filiform projections, suprapapillary thinning**

- **Hyperpigmentation of basal layer**

- **Suprabasilar acantholysis → linear clefts**

- **Mixed dermal inflammatory infiltrate**

Sprecher 2007
Pathogenesis

- Mutation in the KRT5 (Keratin 5) gene on chromosome 12q reported in patients with Galli-Galli disease is believed to lead to:
  - abnormal intermediate filament cytoskeleton
  - compromise in the structural integrity of basal layer keratinocytes → suprabasilar acantholysis

- Hyperpigmentation seen in Galli-Galli, Dowling-Degos, and EBS-MP suggests that keratin 5 probably plays a role in melanin trafficking

Liao et al 2007; Sprecher 2007

Treatment

- Treatment of hyperkeratosis is challenging:
  - Tretinoin and urea cream are irritating and often exacerbate the hyperkeratosis
  - Oral retinoids not helpful

- Therapies reported to improve pruritus have been slightly more successful:
  - NBUVB
  - Prednisone or topical corticosteroids
  - Cyclosporine

Gilchrist 2008; Braun-Falco 2001
Summary

• Galli-Galli disease is an acantholytic variant of Dowling-Degos disease

• Galli-Galli disease should be included in:
  - The clinical DDx of reticulate hyperpigmentation
  - The histologic DDx of focal acantholysis +/- dyskeratosis

• Keratin 5 mutations are found in Galli-Galli, Dowling-Degos, and EBS with mottled hyperpigmentation

References


