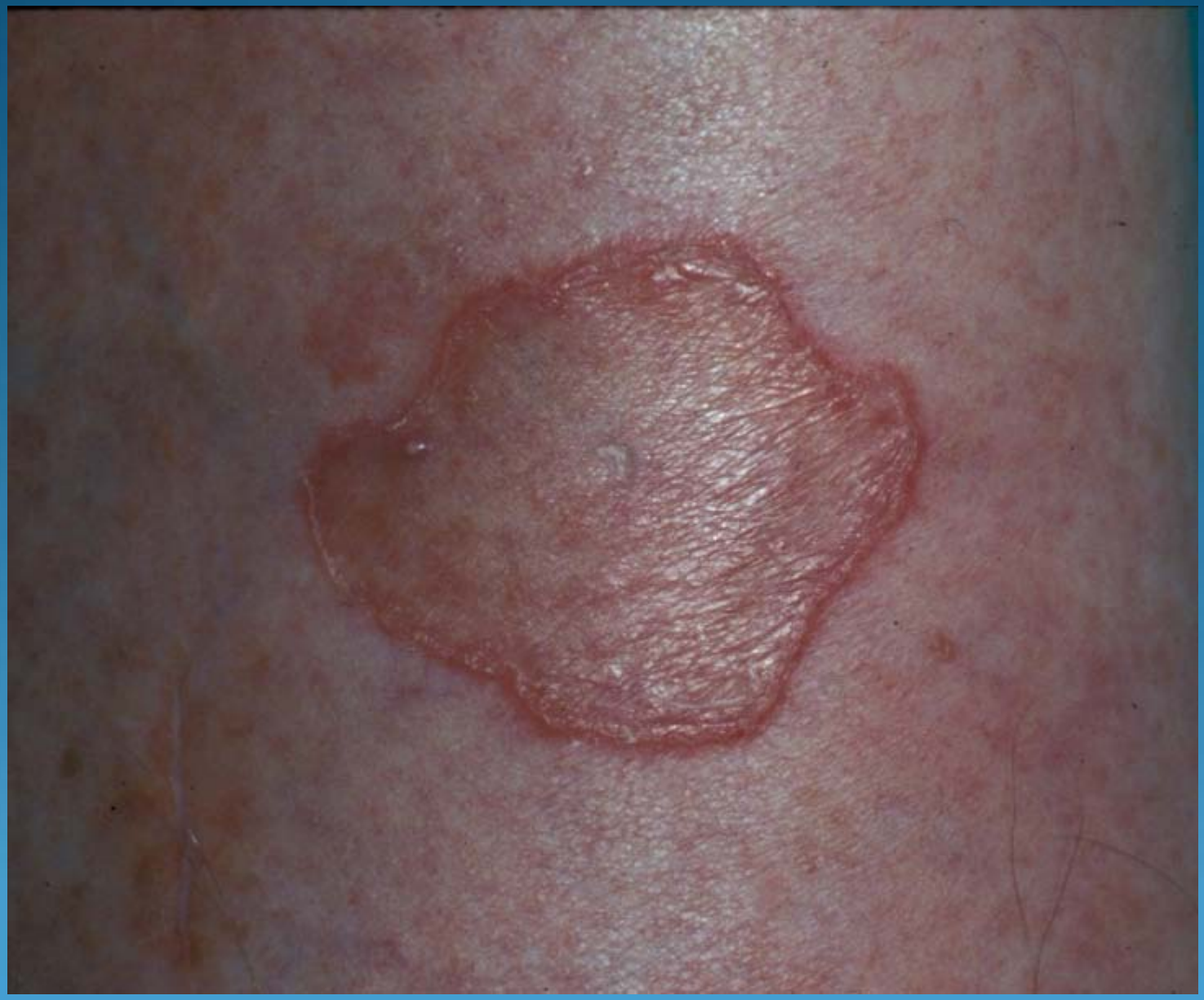
POROKERATOSIS



Porokeratosis: Introduction

- Benign epidermal proliferation
- Distinct clinical & histologic features
- 5 clinical subtypes
- Erroneously named *poro*keratosis

CLINICAL SUBTYPES

1. Porokeratosis of Mibelli
2. Disseminated Superficial (Actinic) Porokeratosis
3. Porokeratosis Palmaris Plantaris et Disseminata
4. Linear Porokeratosis
5. Punctate Porokeratosis

Pathogenesis

- Definitive pathogenesis unclear
- Disorder of keratinization (?)
- Hypotheses:
  1. *Mutant clone of keratinocytes* with an inflammatory response
     - Abnormal DNA ploidy in keratinocytes
     - Immunocompromised hosts

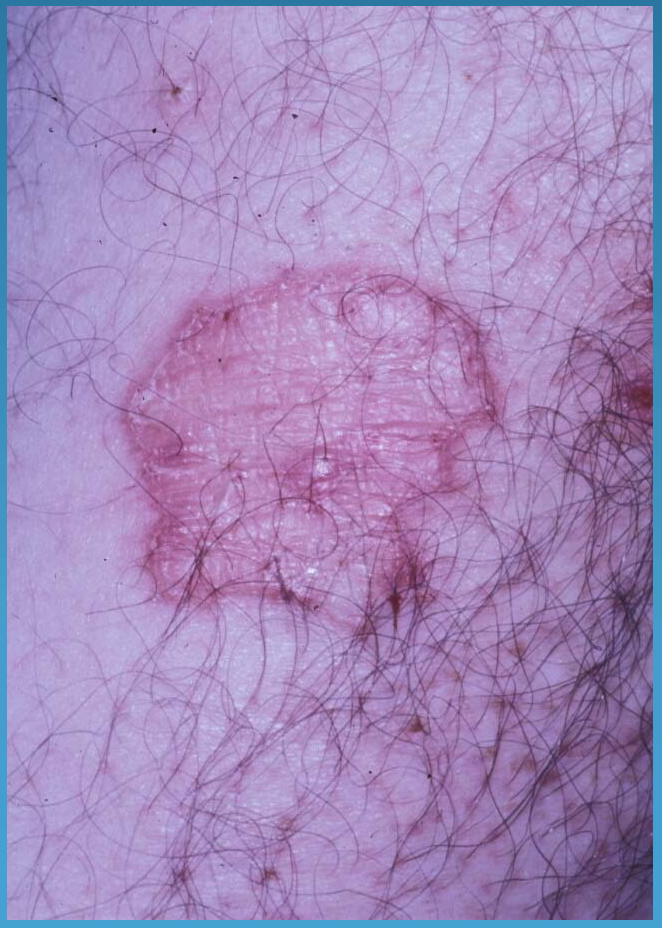
Pathogenesis

2. *Unidentified epidermal antigen* with an inflammatory infiltrate directed against it
   - Mediators released by the infiltrate provide mitotic stimulus for epidermal cells

POROKERATOSIS

- Exacerbating factors:
  - Chemotherapy
  - Ultraviolet radiation/PUVA
  - Immunosuppression (AIDS, transplant)
  - Chemical exposures/drugs
    - benzylhydrochlorothiazide

POROKERATOSIS OF MIBELLI

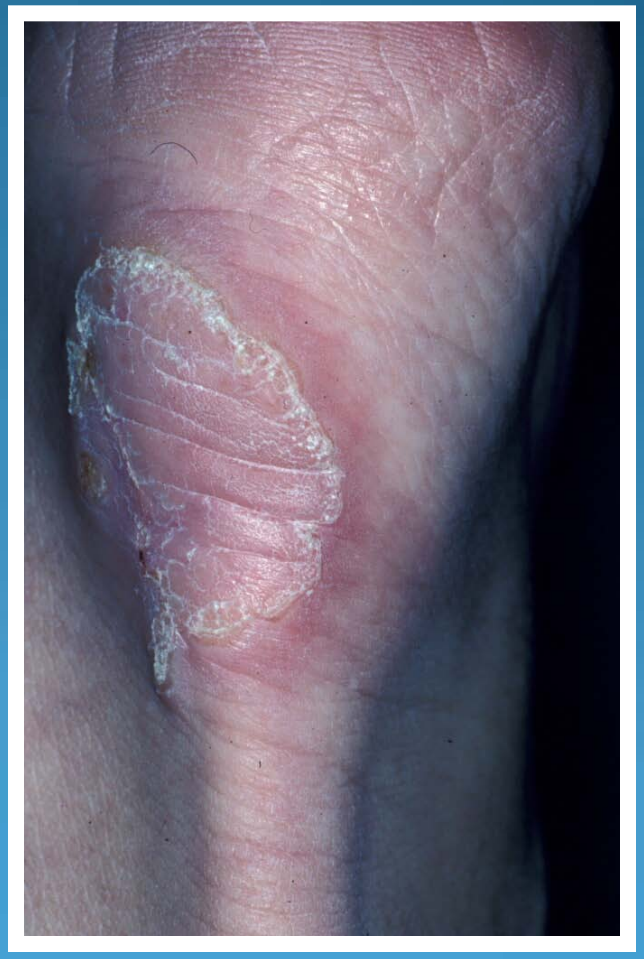


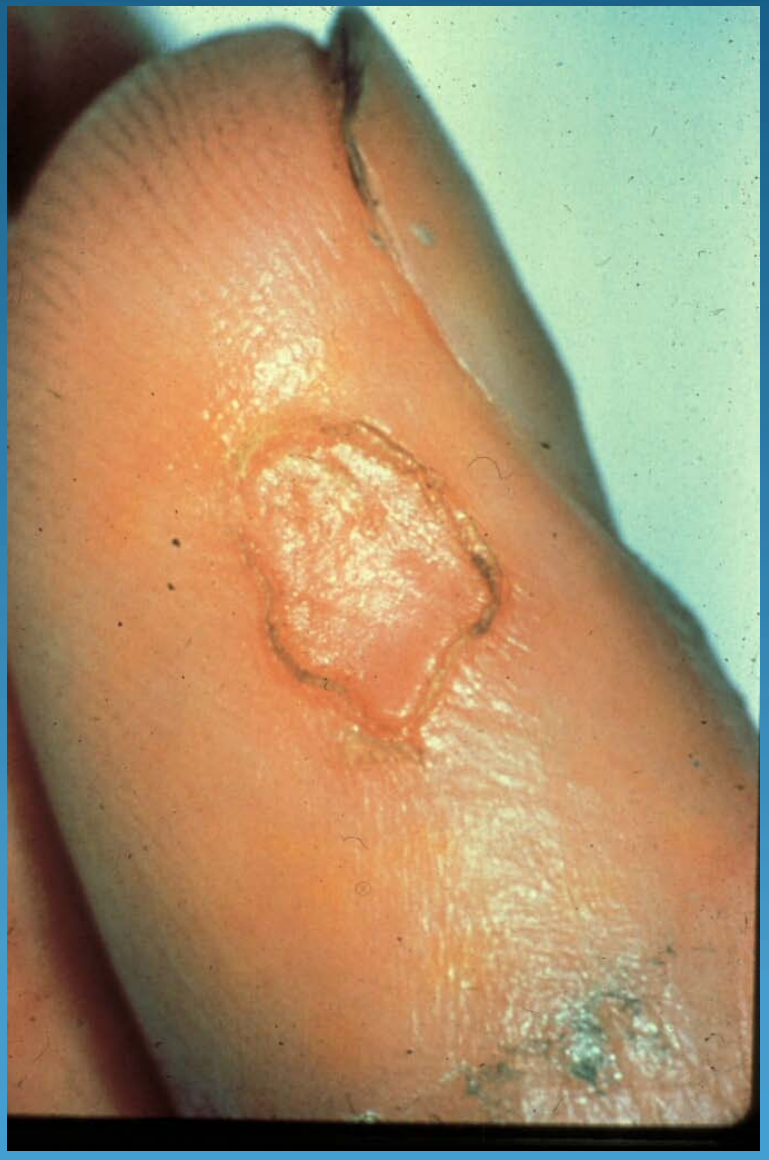
Epidemiology

- Mibelli
  - Begins in infancy/childhood
  - Autosomal Dominant or sporadic
  - Males > females

Porokeratosis of Mibelli

- Asymptomatic
- Localized & unilateral
- Grow slowly- up to 20cm
- Persists indefinitely

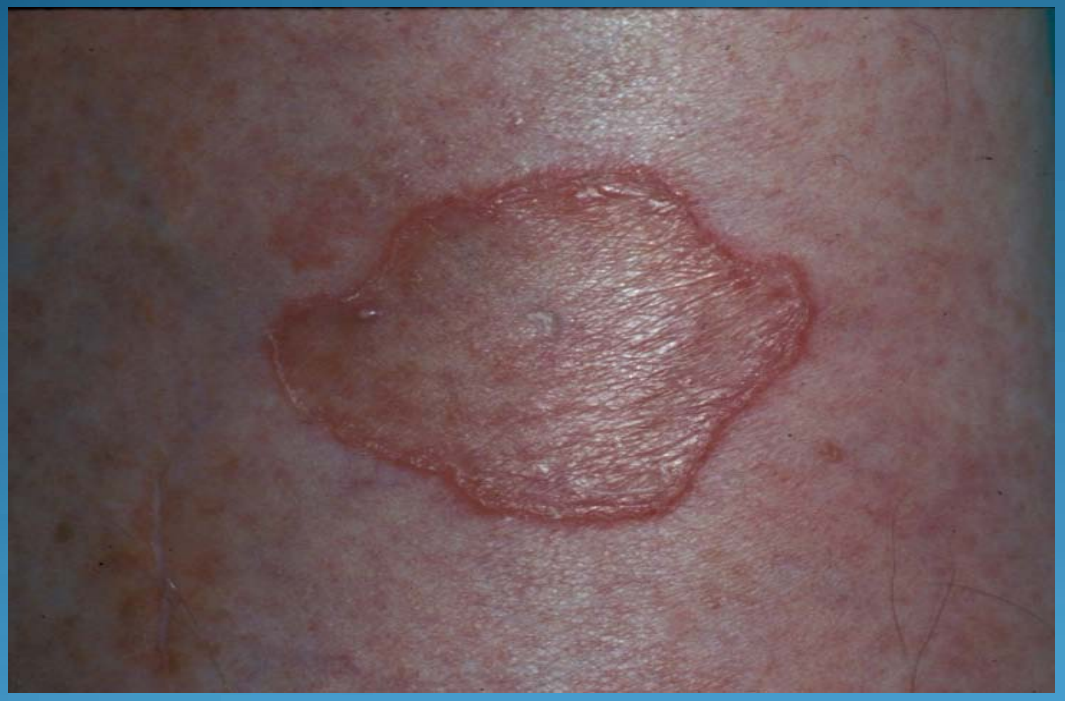




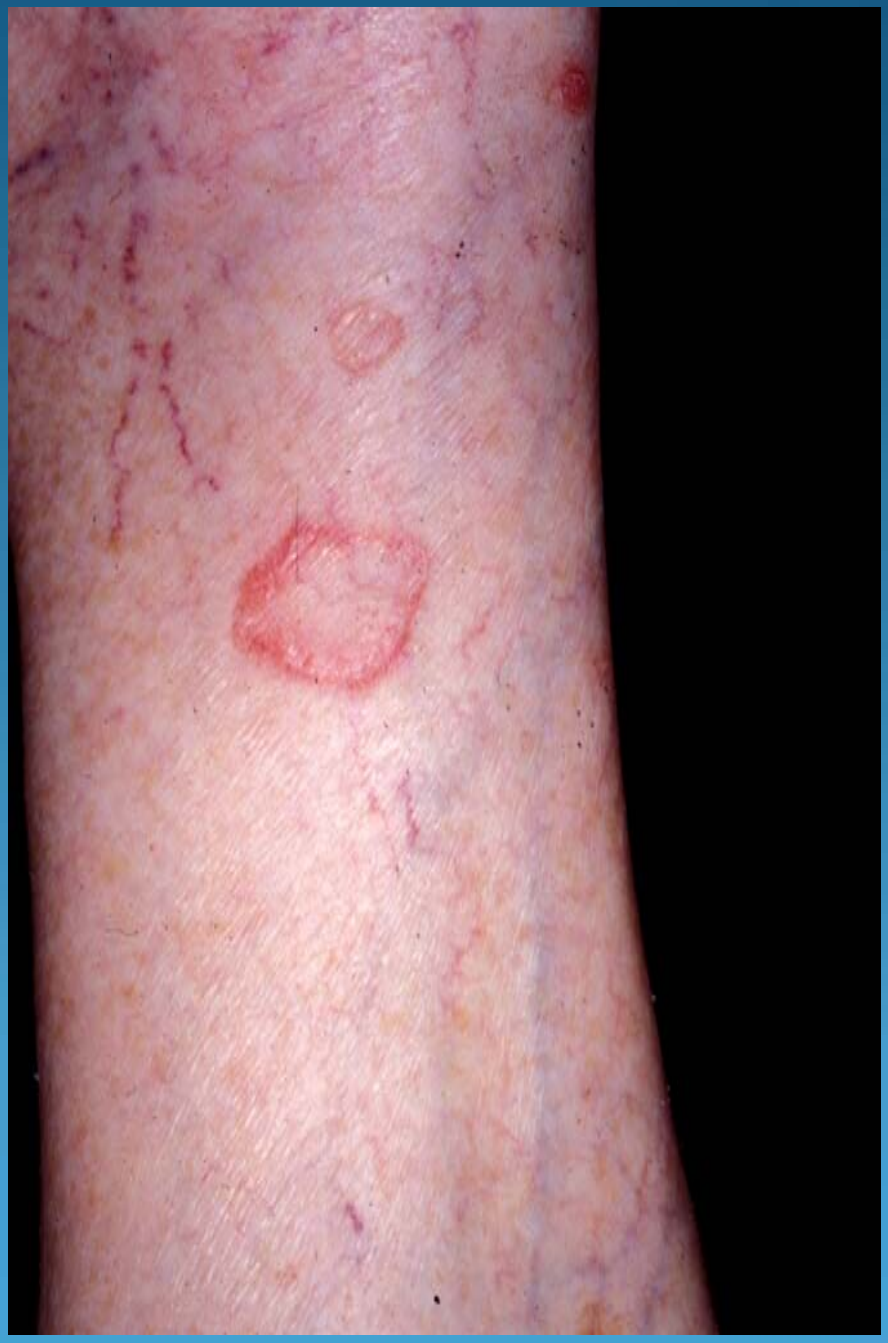
Porokeratosis of Mibelli

- Lesions may occur anywhere
  - *acral locations are most common*
- Nail Matrix: nail dystrophy
- Scalp lesions: alopecia
- Glans penis: erosive balanitis
- Buccal mucosa: macerated; scale appears as a milky white cord

Porokeratosis of Mibelli

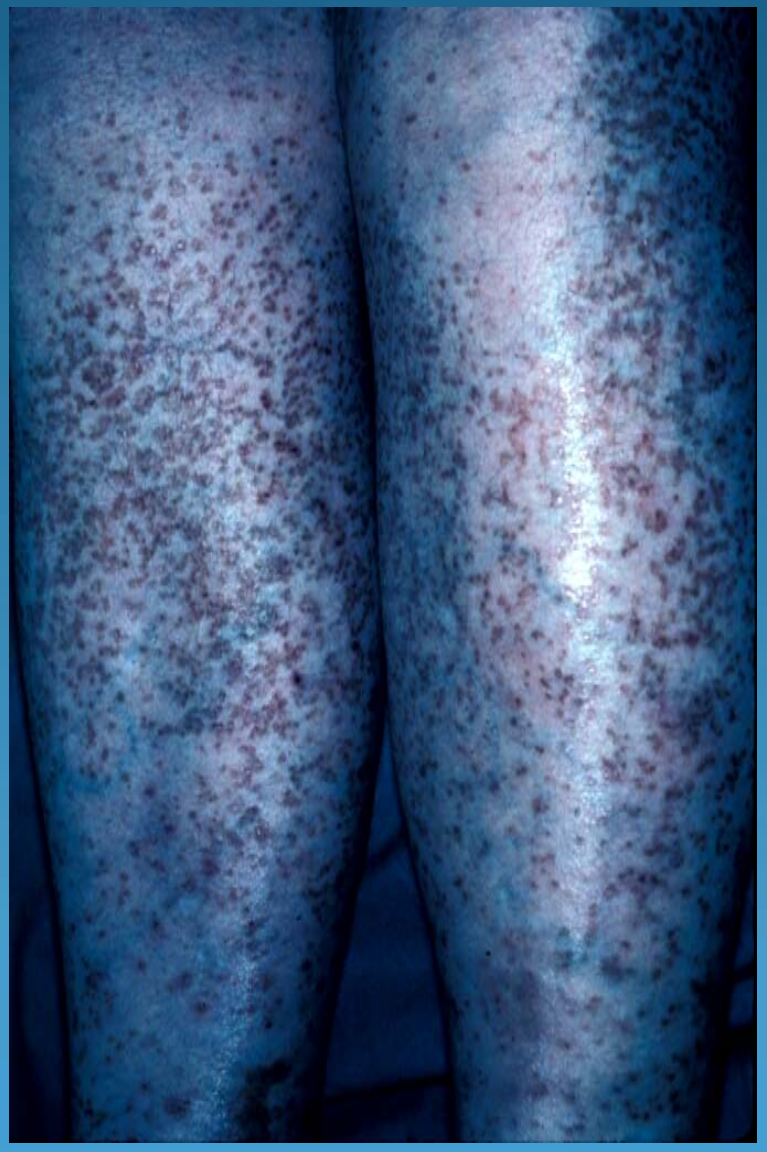


- Bowen's Disease, SCC, and BCC all reported to develop from the lesions of porokeratosis
- Removal is recommended



Disseminated Superficial Porokeratosis

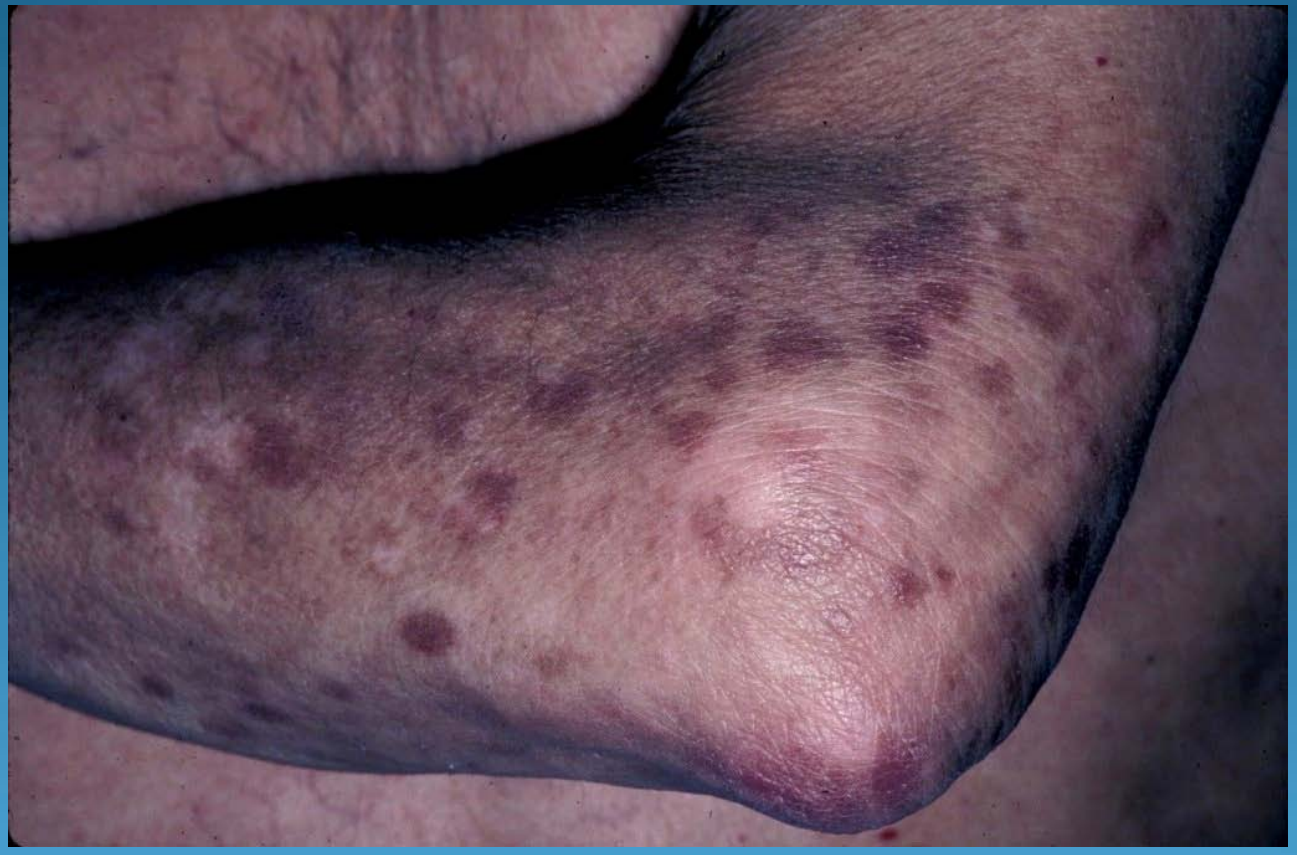
- Multiple lesions with classic clinical and histopathologic appearance
- Bilateral and symmetric- loves the extremities
- *50%* of cases are on sun exposed areas (i.e. the actinic variant)

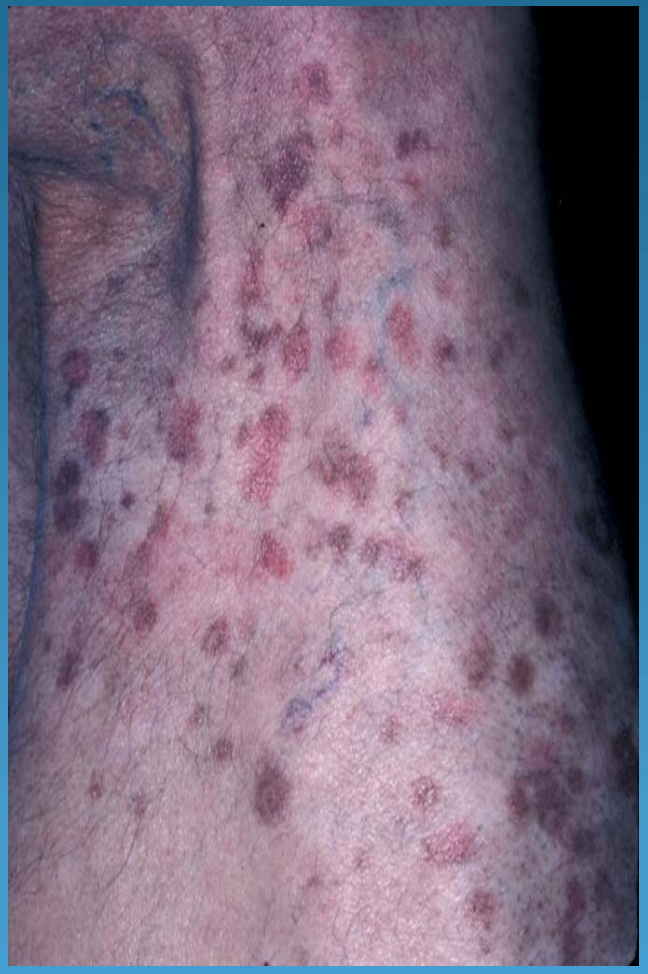


Disseminated Superficial Porokeratosis

- Immunosuppression = exacerbating factor

- Associations:
  - AIDS
  - Cirrhosis
  - Crohn's Disease



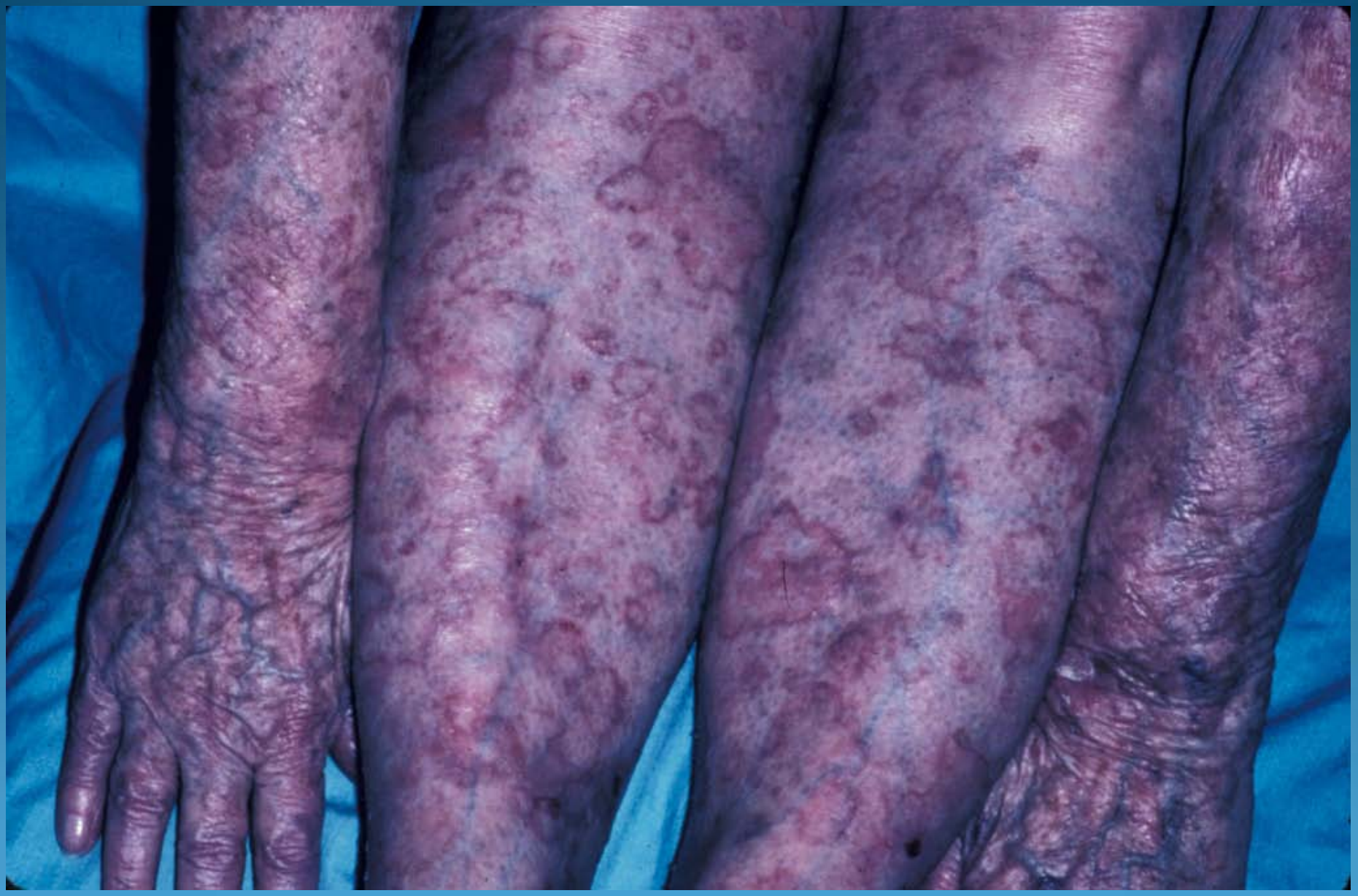


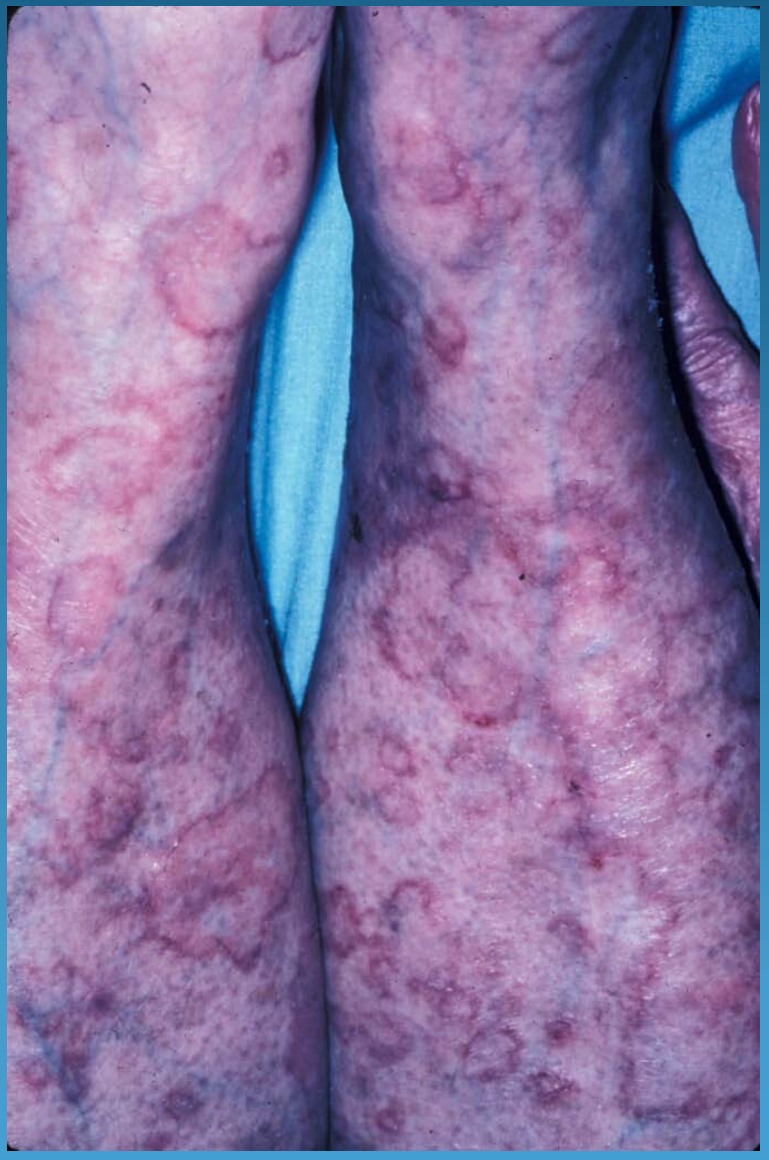
Epidemiology

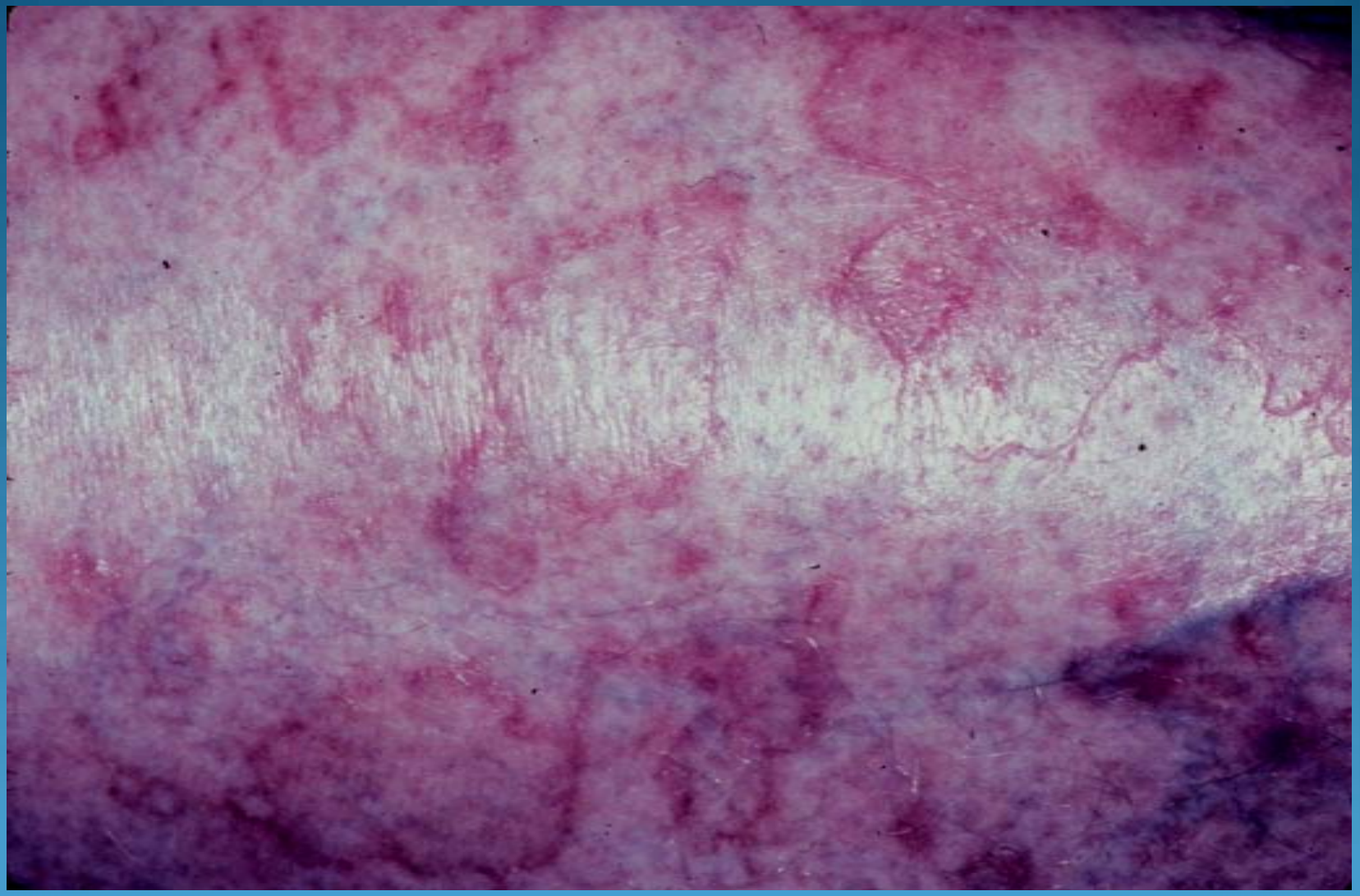
- Disseminated Superficial (Actinic) Porokeratosis
  - Most common variants
  - Autosomal dominant
  - Present in 3rd - 4th decades
  - Female predilection

DSAP

- Asymptomatic or pruritic
- Distribution:
  - DSP: trunk, genitals, palms, soles, mm
  - DSAP: sun exposed sites, spares face!
- Slow growth
- Malignant degeneration may occur





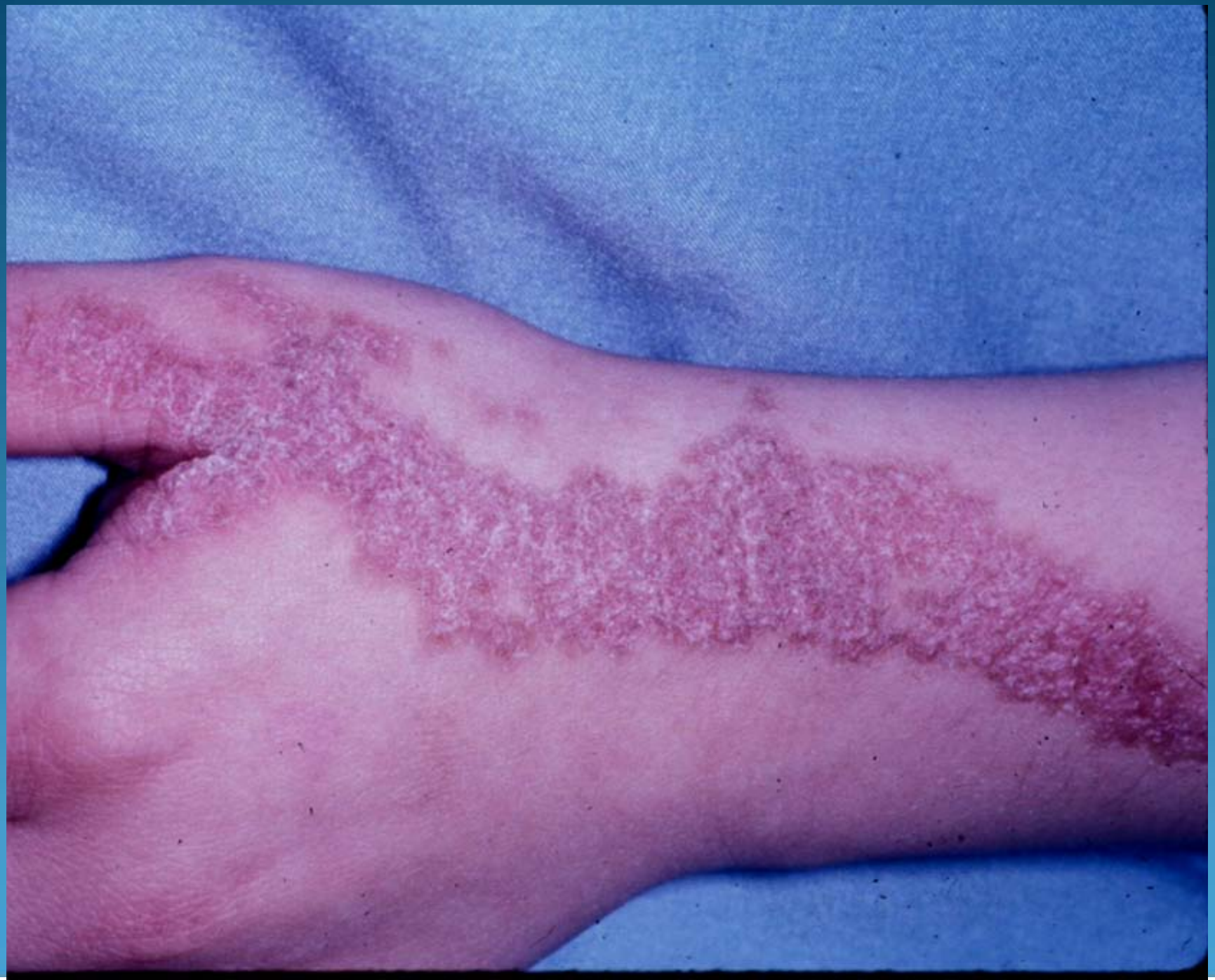


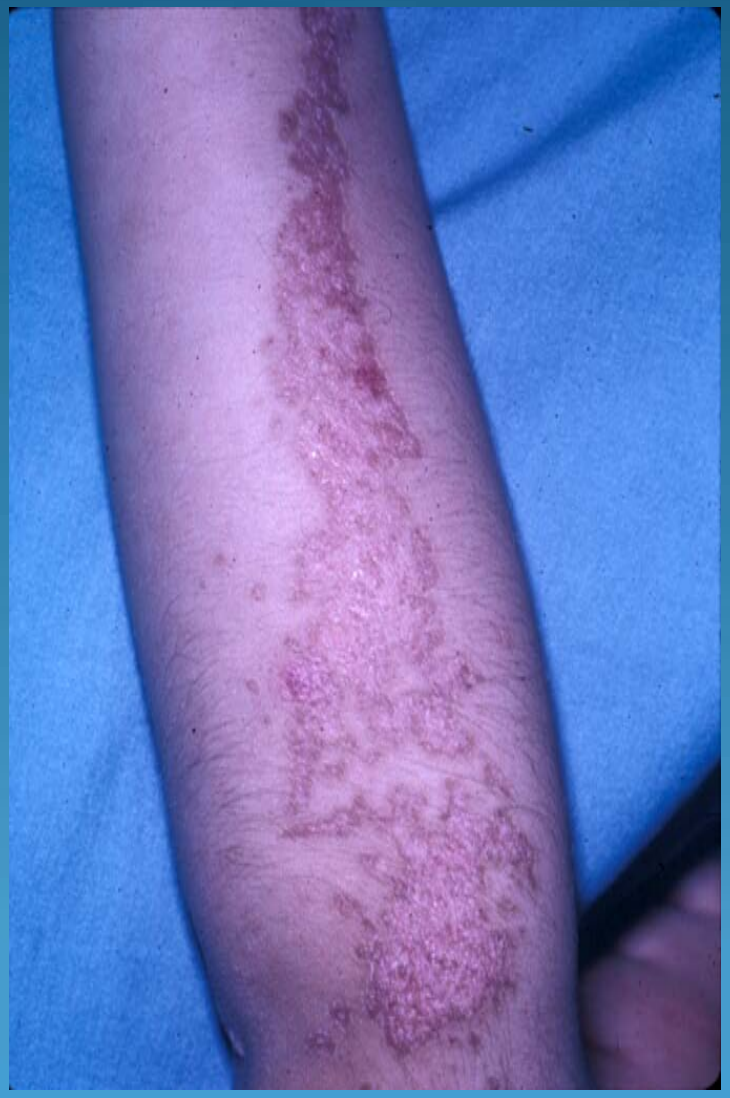
Linear Porokeratosis

- Rare
- Onset usually in infancy/childhood
- Inheritance pattern unclear
- Highest risk for malignant degeneration

Linear Porokeratosis

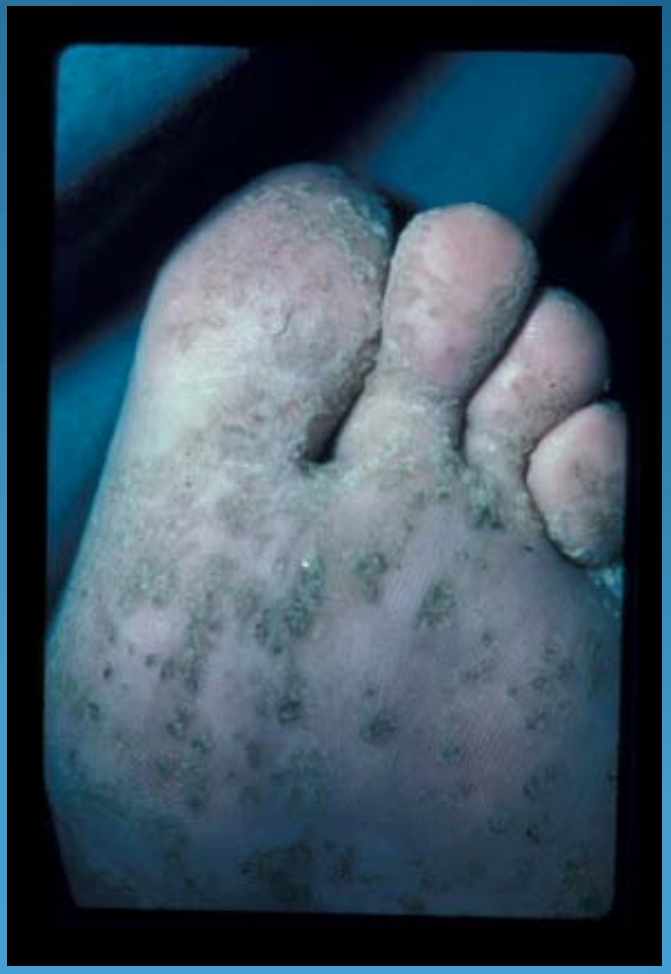
- *Unilateral*
- Segmental or generalized
- May follow Blaschko's lines
- Extremities, trunk, face
- *May mimic linear epidermal nevus*





Porokeratosis Palmaris, Plantaris et Disseminata

- Starts on palms/soles
- May extend over entire body- can burn/sting
- Mucous membranes- asymptomatic

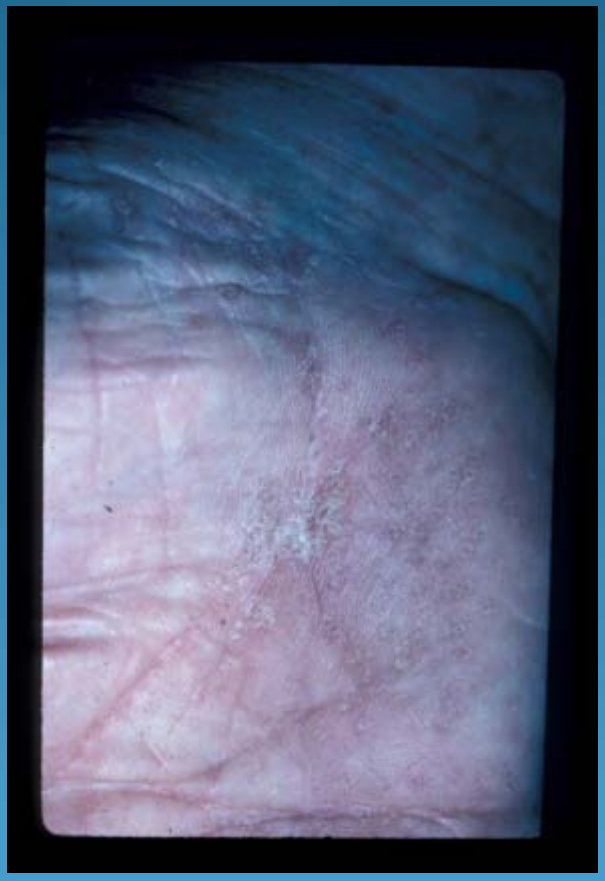


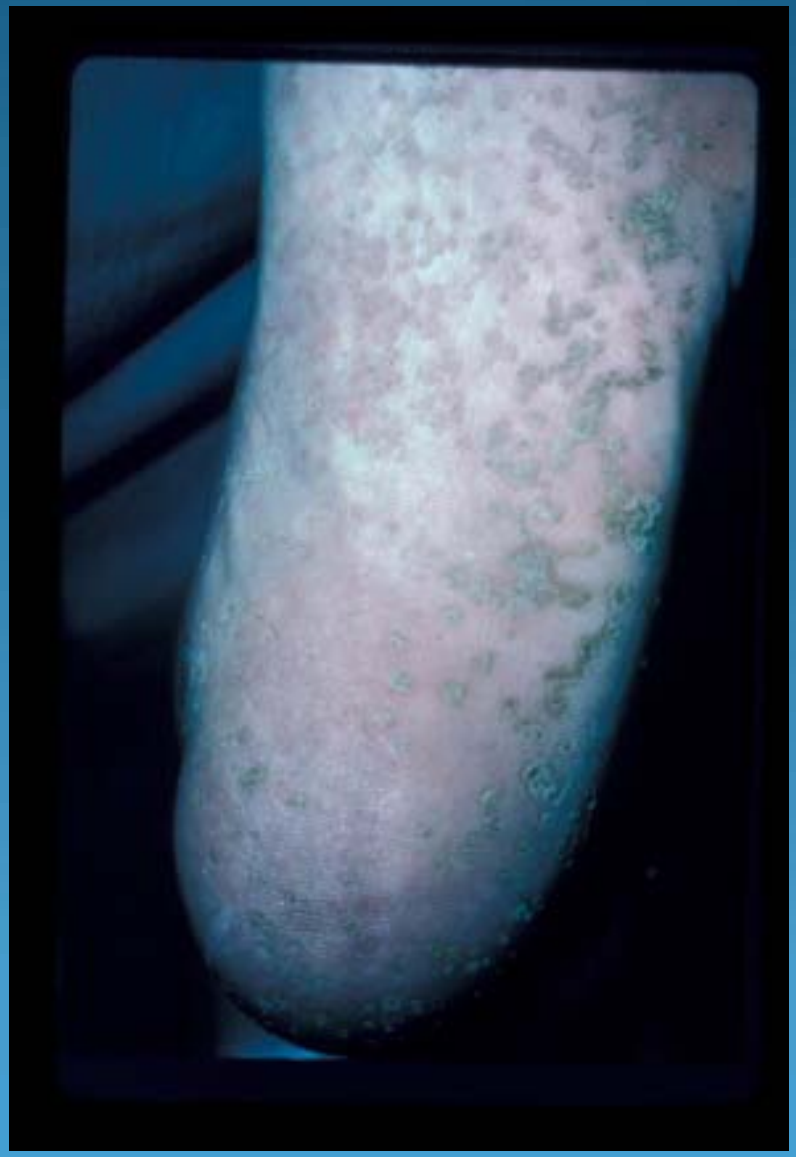
Epidemiology

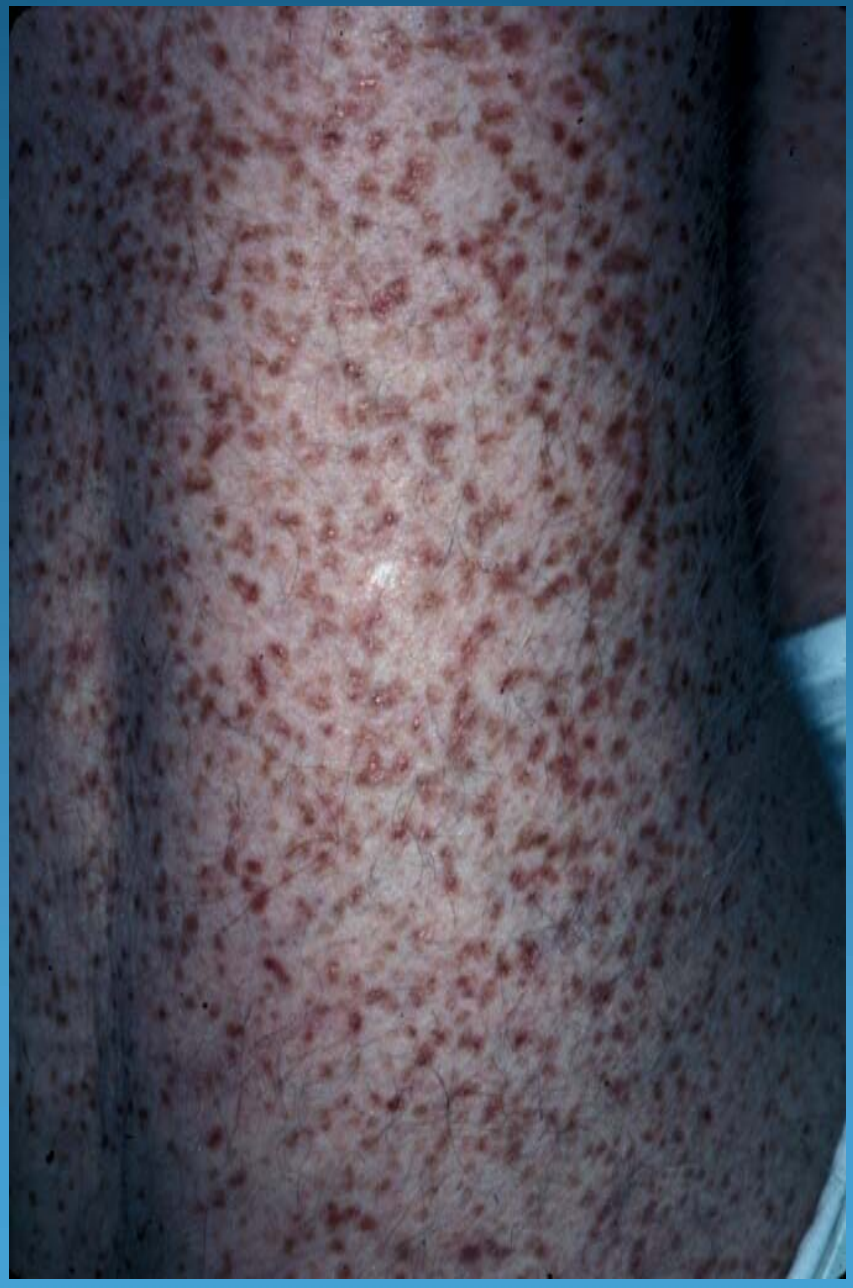
- Porokeratosis Palmaris Plantaris et Disseminata
  - Rare
  - Autosomal dominant
  - Adolescence/early adulthood
  - Men:women = 2:1

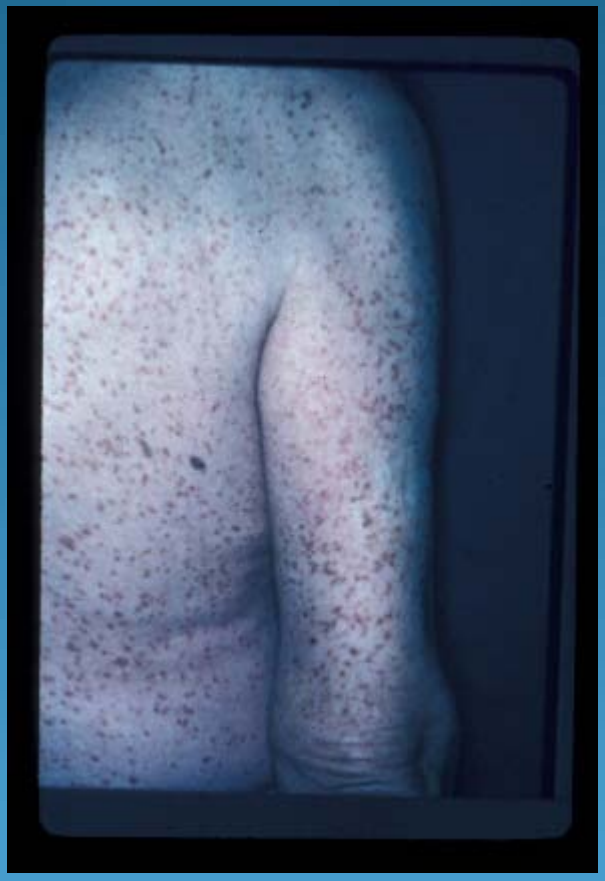
PPPD

- May exacerbate in summer months
- Malignant degeneration









Punctate Porokeratosis

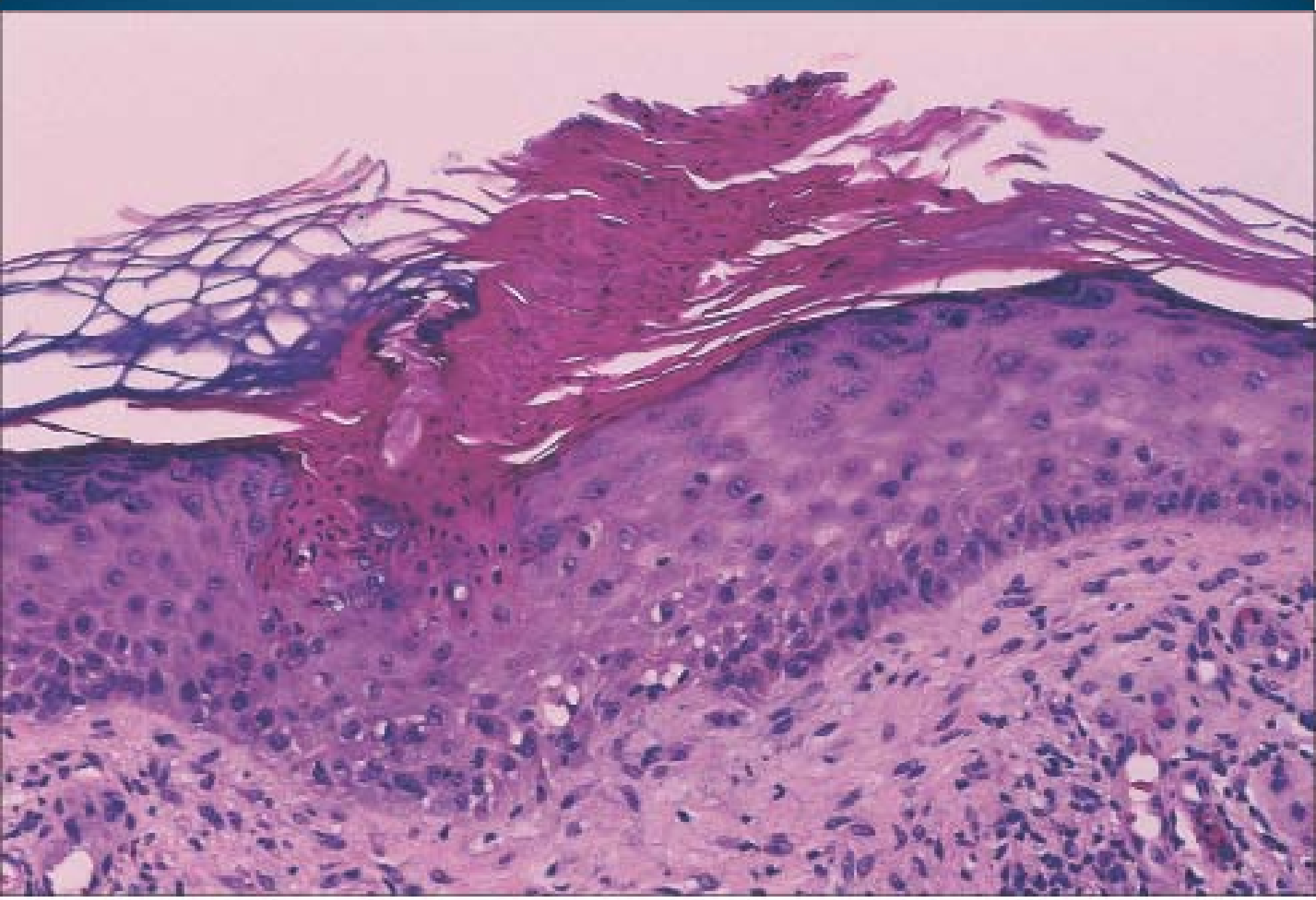
- Multiple minute, discreet, punctate, hyperkeratotic lesions surrounded by a thin, raised margin
- Palms & soles

Epidemiology

- Punctate Porokeratosis
  - Adolescence/adulthood
  - Concomitant involvement with other types
    - Mibelli
    - Linear

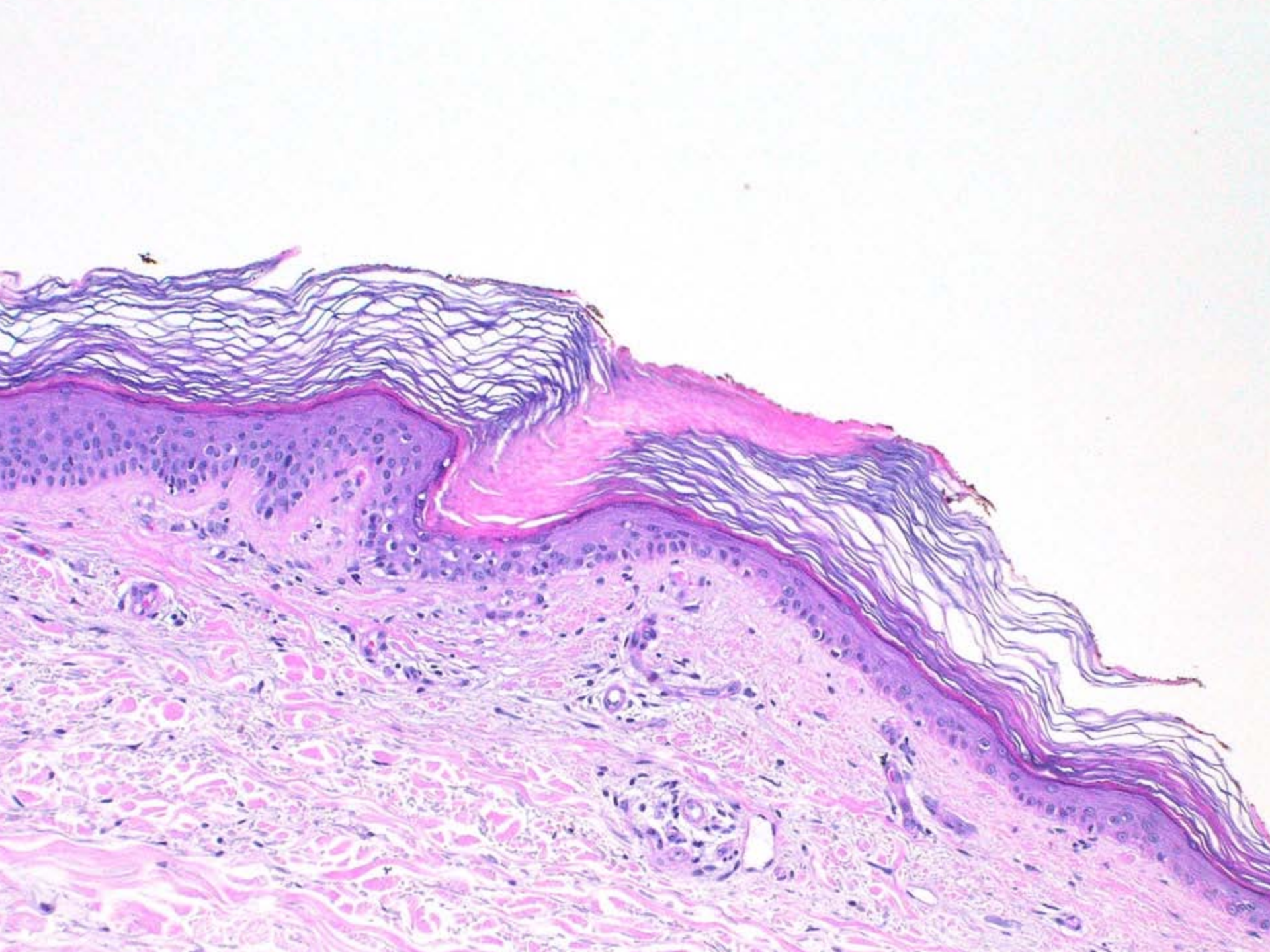
Differential Diagnosis

- Actinic Keratosis
- Stucco Keratoses
- Acrokeratosis Verruciformis
- Verruca plana
- Elastosis Perforans Serpiginosa
- CTCL



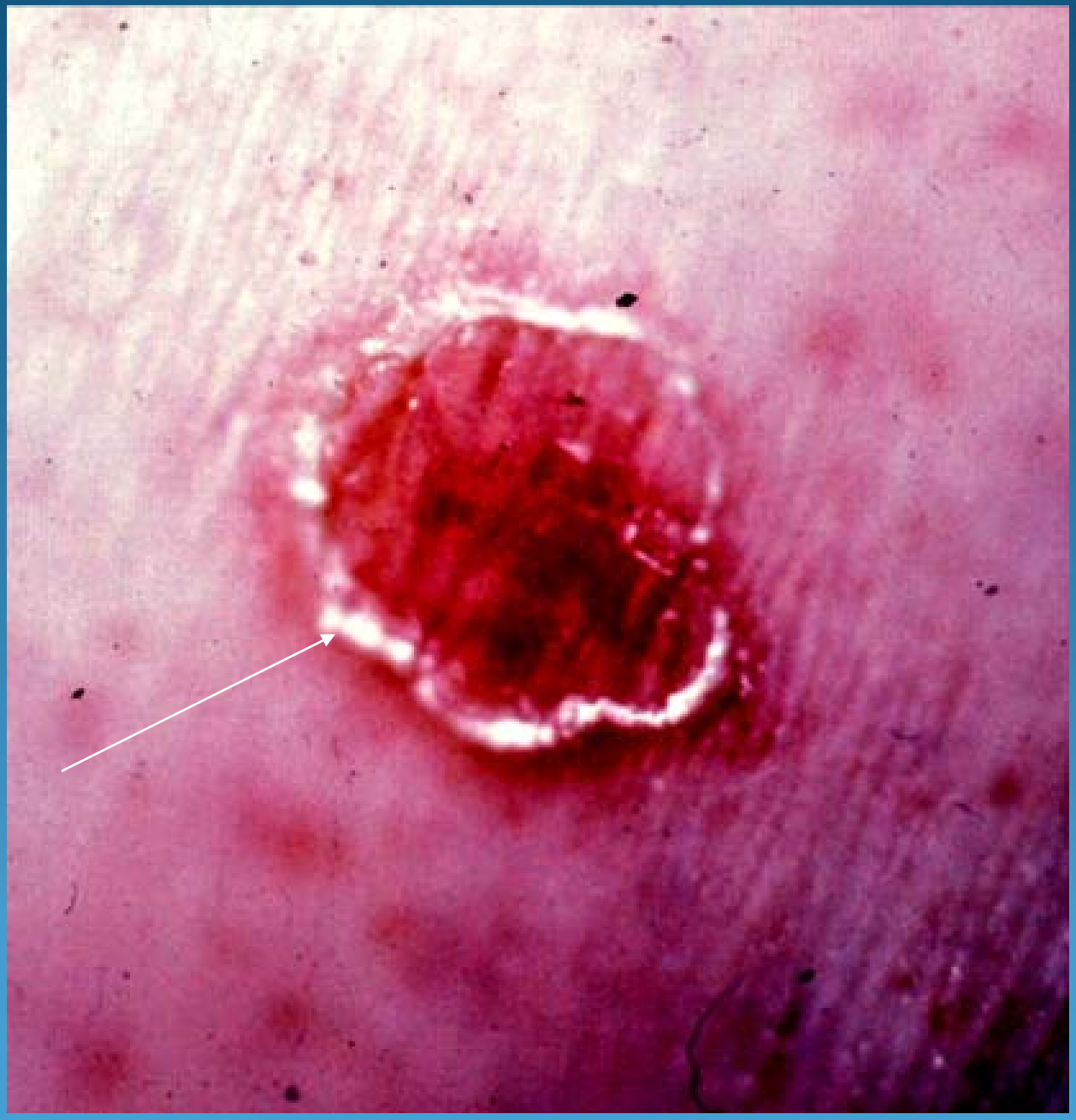
Histology: Porokeratosis

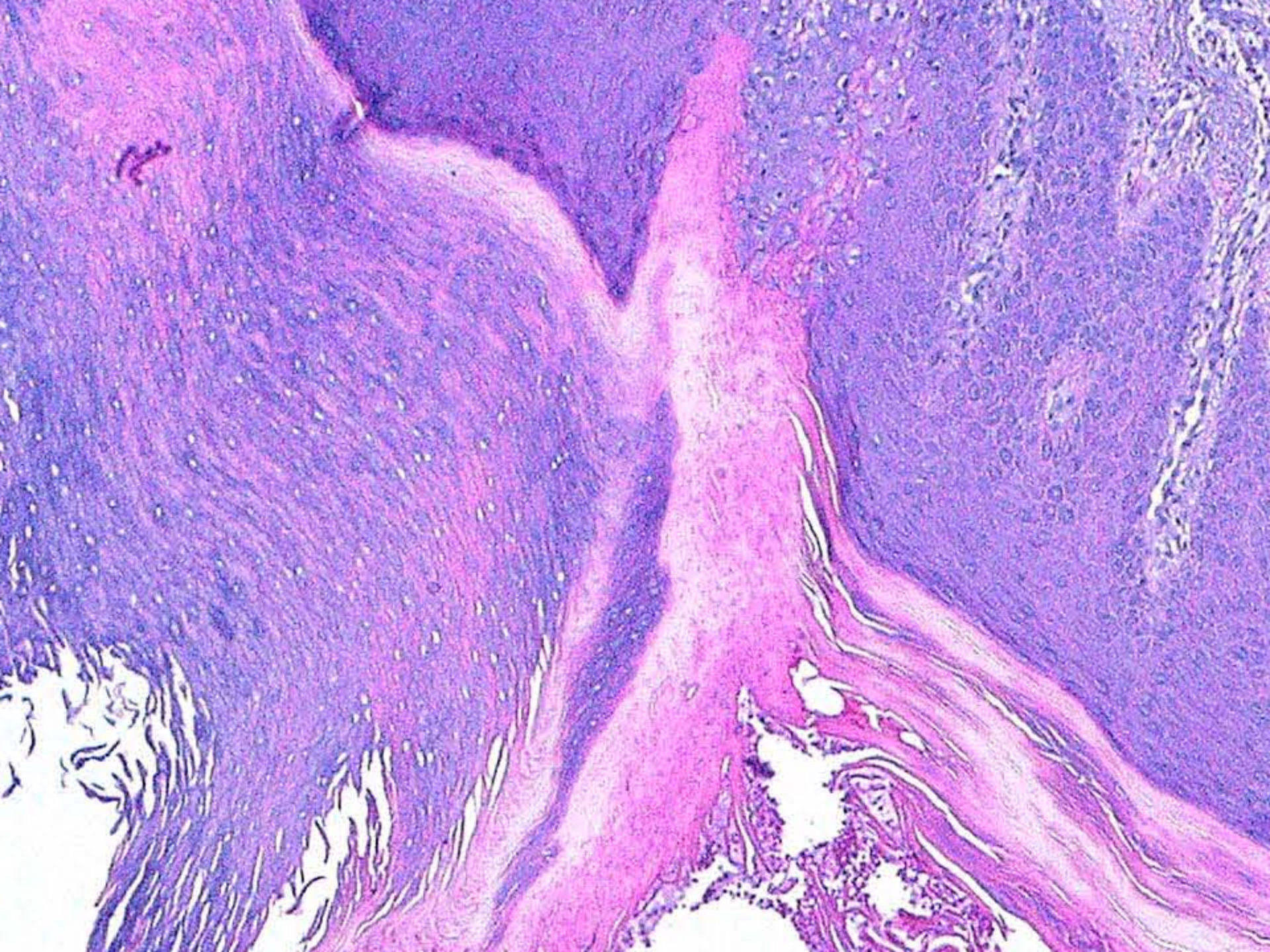
- *Cornoid lamella*
- Dyskeratotic, pyknotic nuclei in kc
- Absent or decreased GL under CL
- Dermal inflammatory infiltrate
- Central epidermal atrophy variable
- +/- Malignant transformation



Cornoid Lamella

- Localized area of faulty keratinization/disordered epithelial metabolism
- Arises in epidermis and may involve ostia of hair follicles or sweat ducts
- Also found incidentally in inflammatory, hyperplastic, and neoplastic disorders of the skin





Treatment Options

- Cryotherapy
- Keratolytics
- Topical/Oral Retinoids
- Topical 5-FU
- Electrodessication
- Excision/Grafting
- *Observation*

Choice of Treatment Depends On:

- Size
- Location
- Function
- Aesthetics
- General health
- *Clinical change*

Natural History / Prognosis

- Lesions usually increase in size/number
- Progression pronounced in DSP and DSAP with UV exposure
- Variable course in immunocompromised

Natural History / Prognosis

- Sudden erratic change- look for immunosuppression
- Malignant transformation
  - Widespread mets and fatality has been reported (Sawai, JAAD 1996)
- Giant PM in acral locations w/ underlying soft tissue and bone destruction