Update on Primary Cicatricial Alopecias

Overview

- Classification
- Epidemiology
- Pathophysiology
- General Approaches
- Subtypes
  - Lymphocytic, Neutrophilic, Mixed

Hair Loss Categories

- Non-scarring
  - Androgenetic
    - Male
    - Female
  - Effluviums
    - Telogen
    - Anagen
  - Alopecia Areata
  - Traumatic
    - Trichotillomania
    - Traction alopecia
  - Drug/toxin induced

- **Scarring**
  - **Pseudopelade of Brocq**
  - **Central centrifugal**
    - **Follicular Degeneration**
    - **Folliculitis Decalvans**
    - **Tufted Folliculitis**
  - **Alopecia Mucinosa**
  - **Lichen Planopilaris**
    - **Graham Little Syndrome**
    - **Frontal Fibrosing Alopecia**
  - **Acne Keloidalis**
  - **Dissecting Cellulitis**
  - **DLE**

Overview

- Cicatricial Alopecia: Definition
  - Must include both visible loss of the follicular ostia and destruction of the hair follicle on histopathology
- Can be Primary or Secondary
  - Primary: Hair follicle is main target of destruction
  - Secondary: Non-follicular disease indirectly causes follicular destruction

Classification

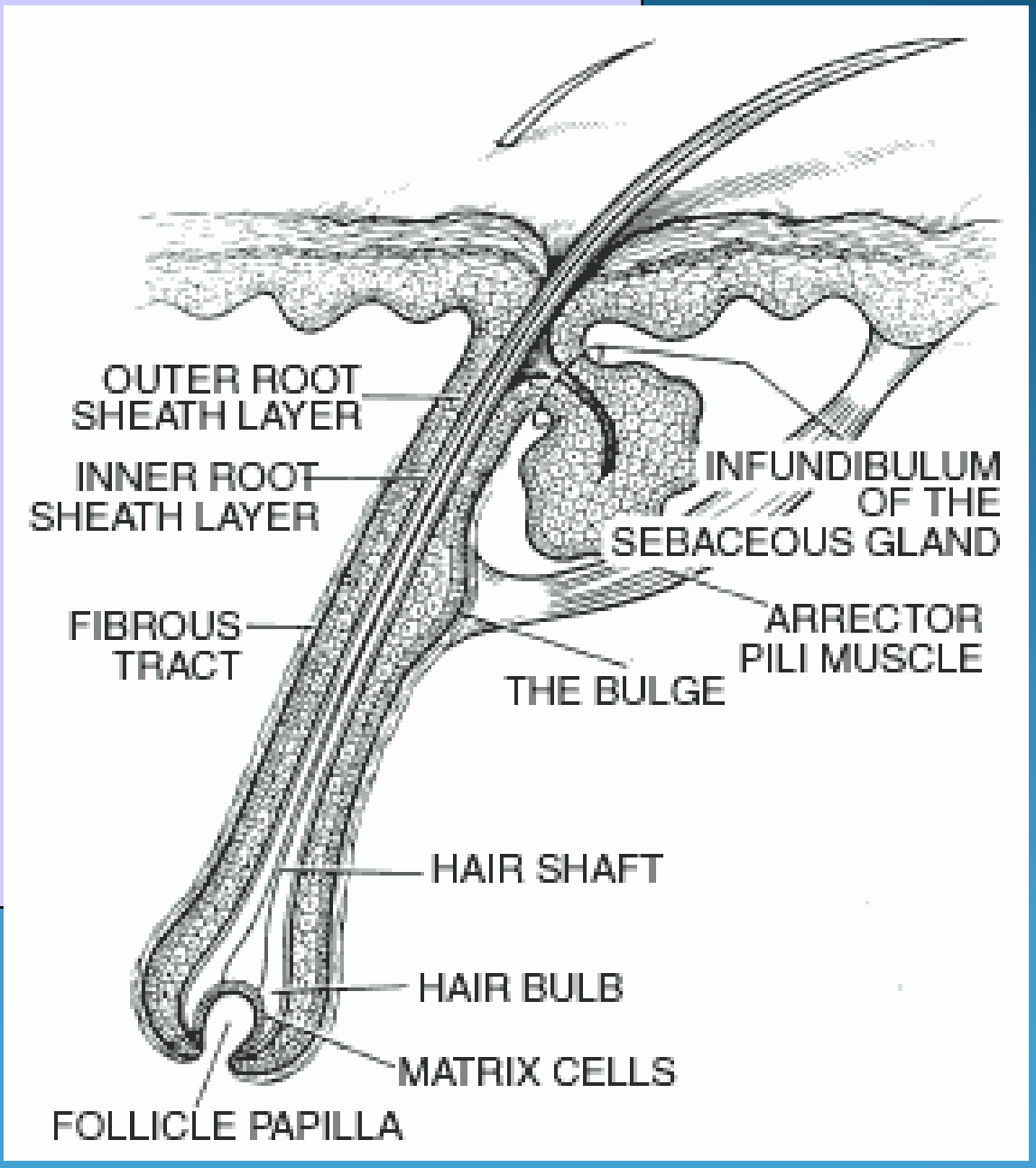
- NAHRS System (2001) based on primary inflammatory cell type
  - **Lymphocytic**: DLE, Lichen Planopilaris, Pseudopelade of Brocq, Central Centrifugal, Alopecia Mucinosa, Keratosis Fol. Spin. Decal.
  - **Neuts:** Folliculitis Decalvans, Dissecting Cellulitis
  - **Mixed**: Acne Keloidalis, Acne Necrotica

Epidemiology

- Retrospective Studies:
  - Cicatricial alopecia in 7.3% (n=427)
  - Primary cicatricial in 3.2% (n=112)
- F:M 2.6:1
- Average age
  - Women: 43
  - Men: 36

Pathophysiology

- Inflammation damages the upper/mid portion of the hair follicle (bulge) required for hair regeneration (insert McGinness basic science lecture here……)
- Non-cicatricial alopecias (ie.AA)—inflam affects the lower (non-critical) portion of the hair follicle


OUTER ROOT SHEATH LAYER
INNER ROOT SHEATH LAYER
FIBROUS TRACT
INFUNDIBULUM OF THE SEBACEOUS GLAND
ARRECTOR PILI MUSCLE
THE BULGE
HAIR SHAFT
HAIR BULB
MATRIX CELLS
FOLLICLE PAPILLA

General Approach

- Physical Exam:
  - Follicular and interfollicular erythema, hyperkeratosis, pigmentary alteration, atrophy
  - Pattern: patchy, reticulate, central, etc.
  - Other general skin lesions or systemic symptoms

General Approach

- Scalp Biopsy
  - Biopsy clinically active areas, not “burnt-out”
  - Obtain 2, 4mm punch biopsies
    - Send one for horizontal sectioning
    - Send the other for vertical only or cut in half and send half for H&E and the other for DIF

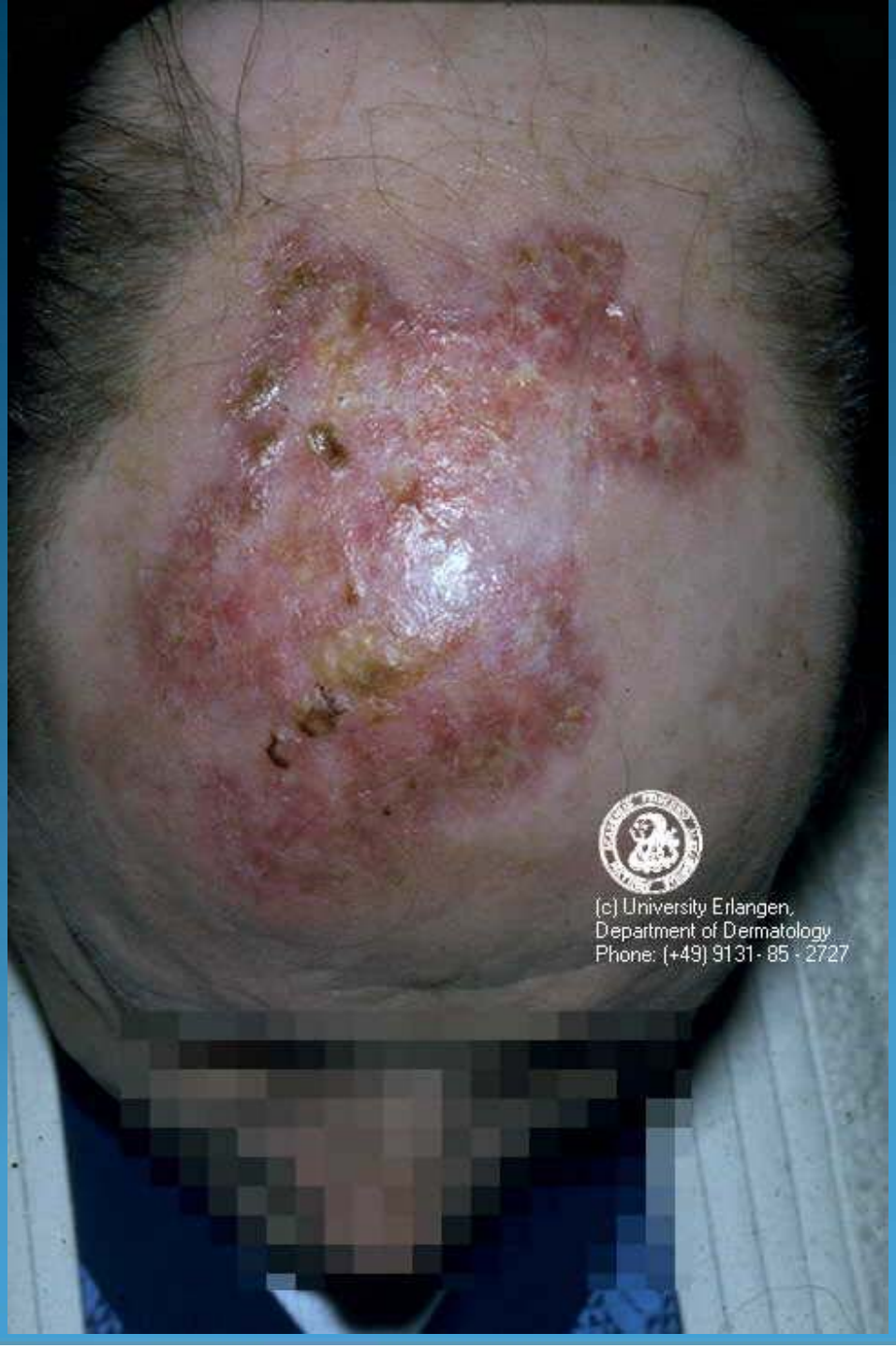
Lymphocytic Cicatricial Alopecias

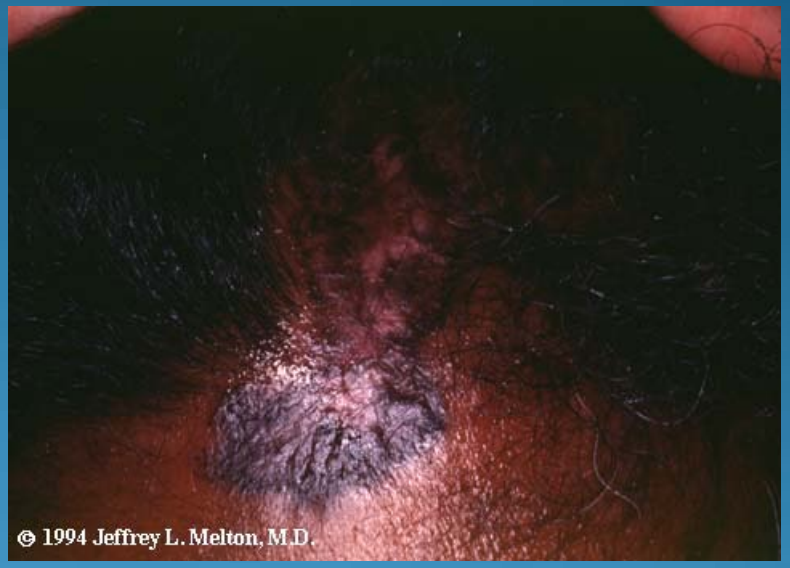
- DLE
- LPP
- Frontal Fibrosing
- Graham-Little Syndrome
- Lupus/LPP overlap
- Pseudopelade of Brocq
- Central Centrifugal
- Alopecia Mucinosa
- Keratosis Follicularis Spinulosa Decalvans

Discoid Lupus Erythematosus

- F>M, onset age 20-40
- 5-10% with DLE will progress to SLE
- 34-56% scalp involvement with DLE
- classic discoid erythematous plaques with follicular plugging and ‘carpet-tack’ sign
- later, hypo/hyperpigmentation, atrophy and telangectasias
- Complications: cosmetic disfigurement, ulceration, SCC’s

Discoid Lupus





Discoid Lupus

DLE

- Histopath
  - Vacuolar interface change of the follicular epithelium
  - Scattered dyskeratotic keratinocytes
  - Periadnexal, perifollicular and interstitial lymphocytic infiltrate with dermal mucin
  - Follicular plugging
  - DIF often positive

Treatment of DLE alopecia

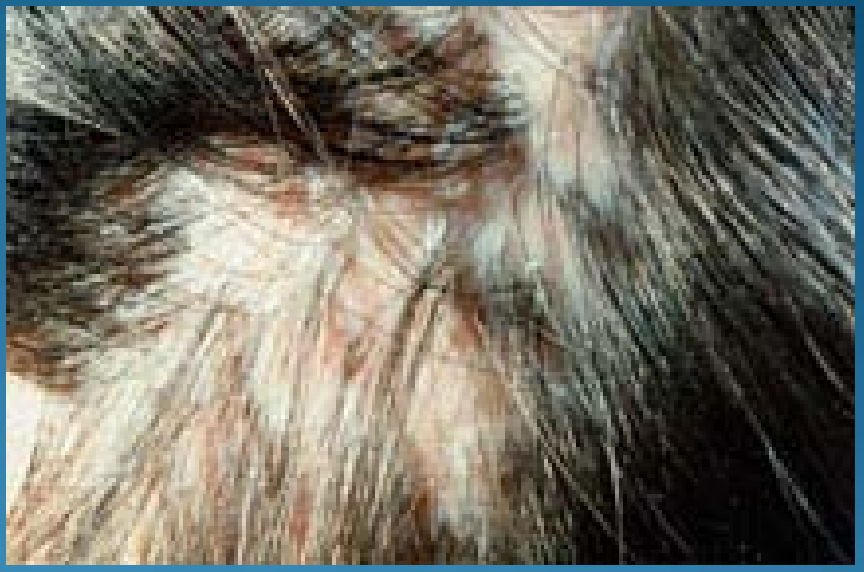
- ROS focused on sx of SLE
- ANA and U/A
- Limited, active disease
  - Class I or II topical steroids BID
  - Intra-lesional Kenalog 3-10mg/cc Q4-6 weeks
- Rapidly progressive or extensive disease
  - Plaquenil 200 mg BID +/- oral prednisone for the first 8 weeks
  - Accutane (2^{nd} line) 1mg/kg/day
- Sun protection and avoidance of trauma

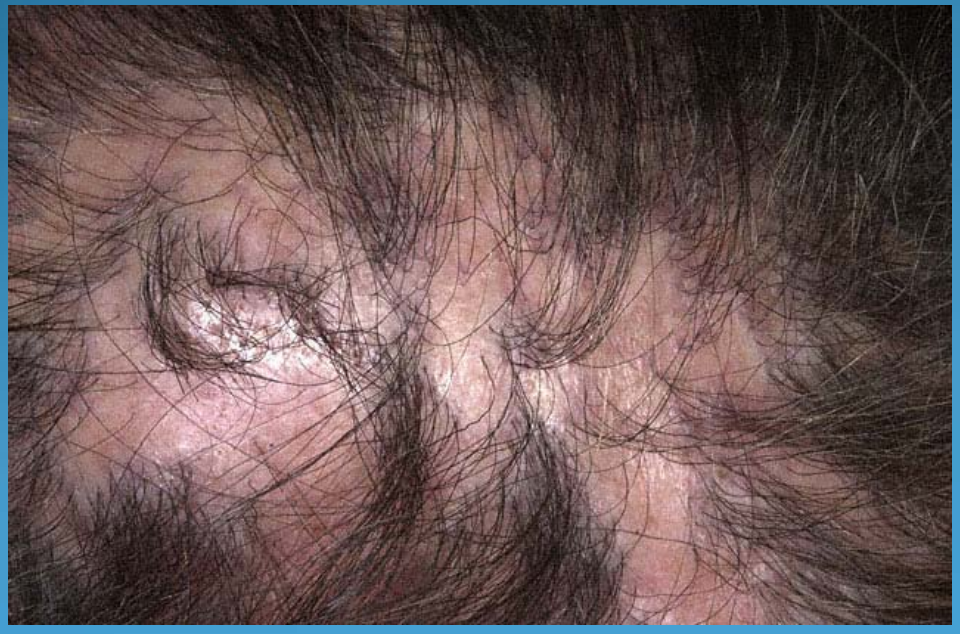
Lichen Planopilaris

- A follicular variant of lichen planus
- 3 forms
  - Classic, Graham-Little, frontal fibrosing
- Thought to be secondary to an antigenic trigger or related to certain medications (similar to classic LP)
  - Gold, quinacrine, atabrine, hep B vaccination, hepatitis C infection

LPP

- Seen in adults, usually arising in middle-aged females
- Extracranial LP present in 17-50% of pts.
- Sx at presentation: shedding, hair loss, pruritus
- PE: perifollicular erythematous papules and spinous follicular hyperkeratosis. Can see unaffected hairs in scarred areas

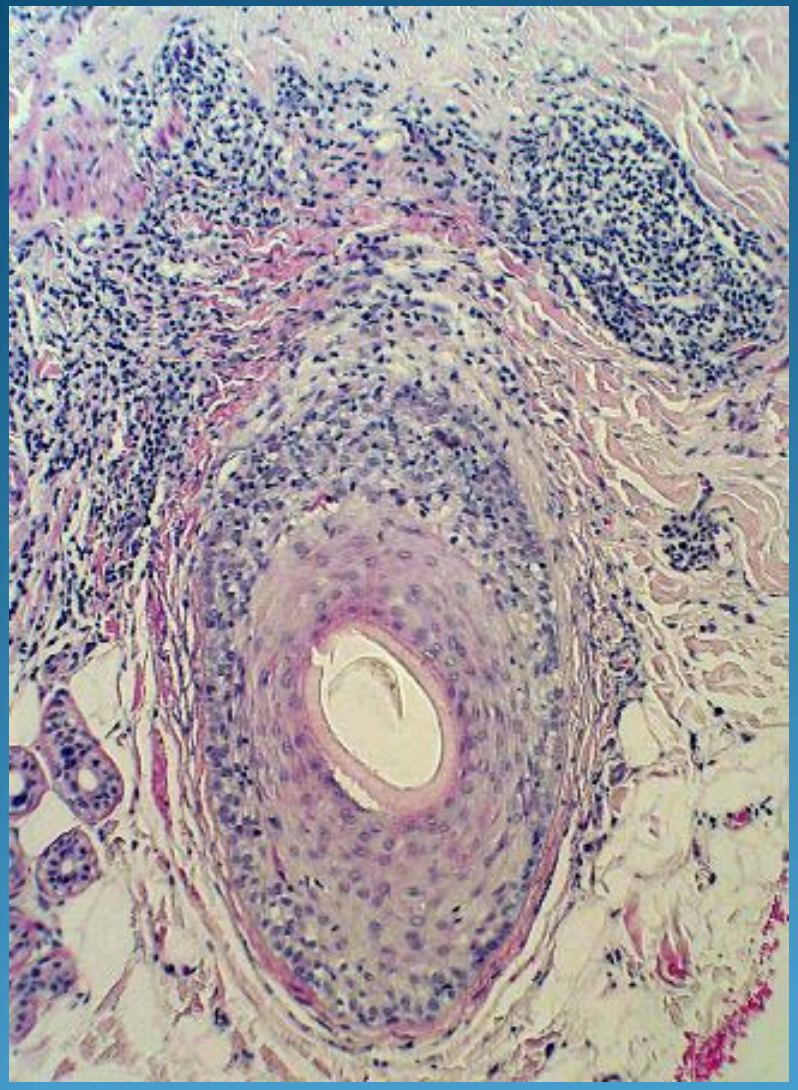


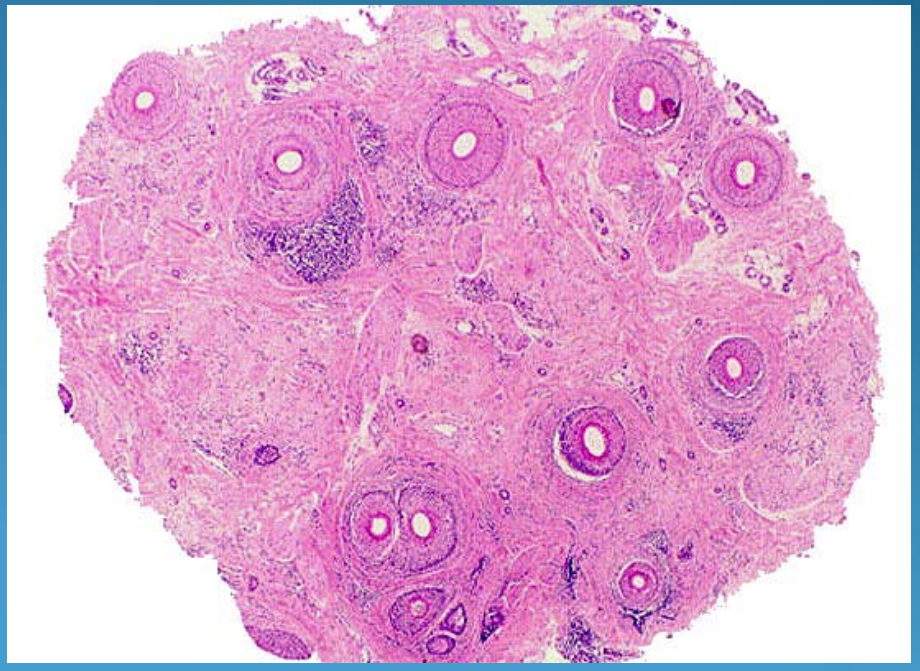


Lichen Planopilaris

LPP

- DDx includes other cicatricial alopecias
  - Activity limited to hair-bearing periphery (unlike DLE and alopecia mucinosa)
  - No pustules (unlike folliculitis decalvans)
- Histopath
  - Lichenoid infiltrate
  - Lymphs at upper portion of follicle
  - DIF may reveal patchy deposition of fibrinogen and IgM along the BMZ





Lymphocytic Infiltrate at upper/mid portion of follicle

LPP Management

- Explore possible drug related cause
- Test for Hep C (esp. if eroded or ulcerated scalp disease)
- Treatment
  - High potency topical steroids BID or intra-lesional 3-10 mg/cc
  - Prednisone 1 mg/kg tapered over 2-4 months
  - Low dose accutane 10 mg/d over months
  - Plaquenil 200 mg BID
  - Griseo 250 mg BID x 7-10 months

Frontal Fibrosing Alopecia

- Also called postmenopausal FFA
- Frontotemporal hairline scarring pattern mainly affecting postmenopausal women
- Shiny, pale, bandlike zone; active areas reveal hairs with perifollicular erythema and hyperkeratosis
- Absent or thinned eyebrows
- May have axillary and extremity hair loss
- May also have classic LPP or LP

Frontal Fibrosing Alopecia

- DDx: ophiasis, female pattern recession, traction alopecia
- Histopath:
  - Features indistinguishable from LPP
  - Lichenoid infiltrate
  - Upper follicular inflamm

Management

- Attempt to stabilize disease with topical mid-potency steroids BID
- Po prednisone or plaquenil may slow progression
- Other case reports: intralesional steroids, accutane, soriatane, griseo, minoxidil, etc. are mostly ineffective

Graham-Little Syndrome

- Aka Graham-Little-Piccardi-Lassueur
- Considered a variant of LPP
- Uncommon alopecia seen in adults
- Patchy cicatricial scalp alopecia, nonscarring alopecia of the axillary and pubic areas, and grouped follicular papules on the trunk/extremities resembling lichen spinulosus or KP

Graham-Little Syndrome

- Histopath:
  - similar to LPP
- Treatment:
  - High potency topical steroids alone or in combo with intralesional steroids (10mg/cc)
  - PO steroids
  - Cyclosporine 4mg/kg/d x 3 months

Pseudopelade of Brocq

- Distinct entity or common endpoint?
- Onset in adulthood
- Atrophic, oval to round, white to ivory scarred plaques of alopecia, vertex almost always involved
- No clinical evidence of inflammation
- Slowly progressive

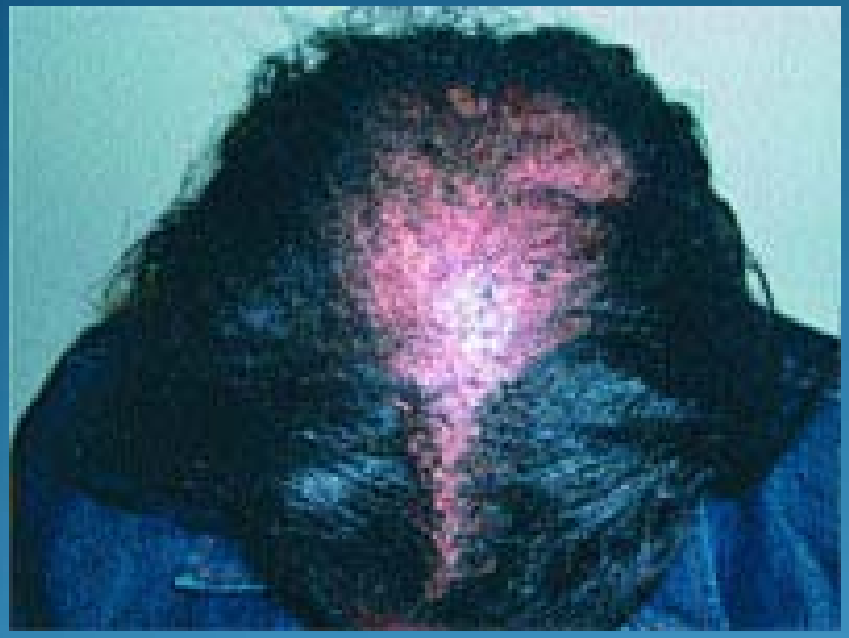
Pseudopelade of Brocq

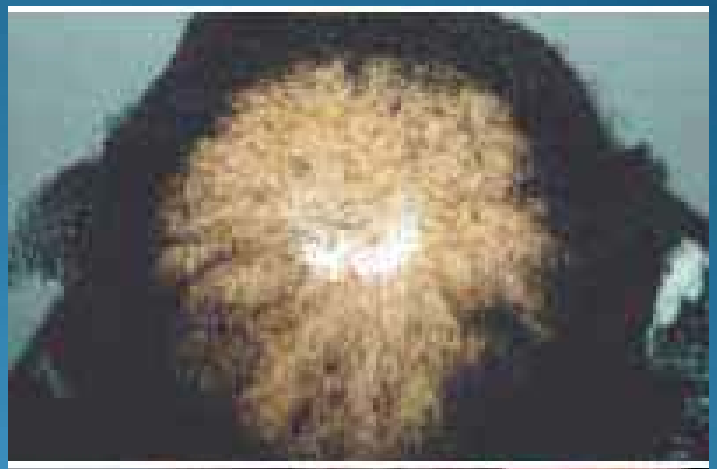
Pseudopelade of Brocq

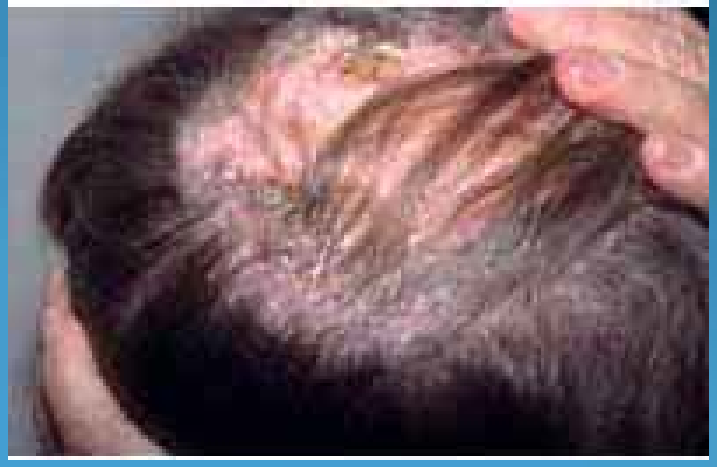
- Histopath: (none classic)
  - Early- perifollicular lymphocytic infiltrate
  - Late-concentric lamellar fibrosis of the hair follicle
- Treatment:
  - Mainly none
  - Many topical and oral therapies have been tried with little success

Central Centrifugal Cicatricial Alopecia

- New term coined to encompass hot comb alopecia and follicular degeneration syndrome
- Most commonly in African-American women
- Presents with flesh-colored, non-inflammatory cicatricial alopecia of the central scalp that enlarges over time
- inherited follicular defect vs. exogenous trauma?







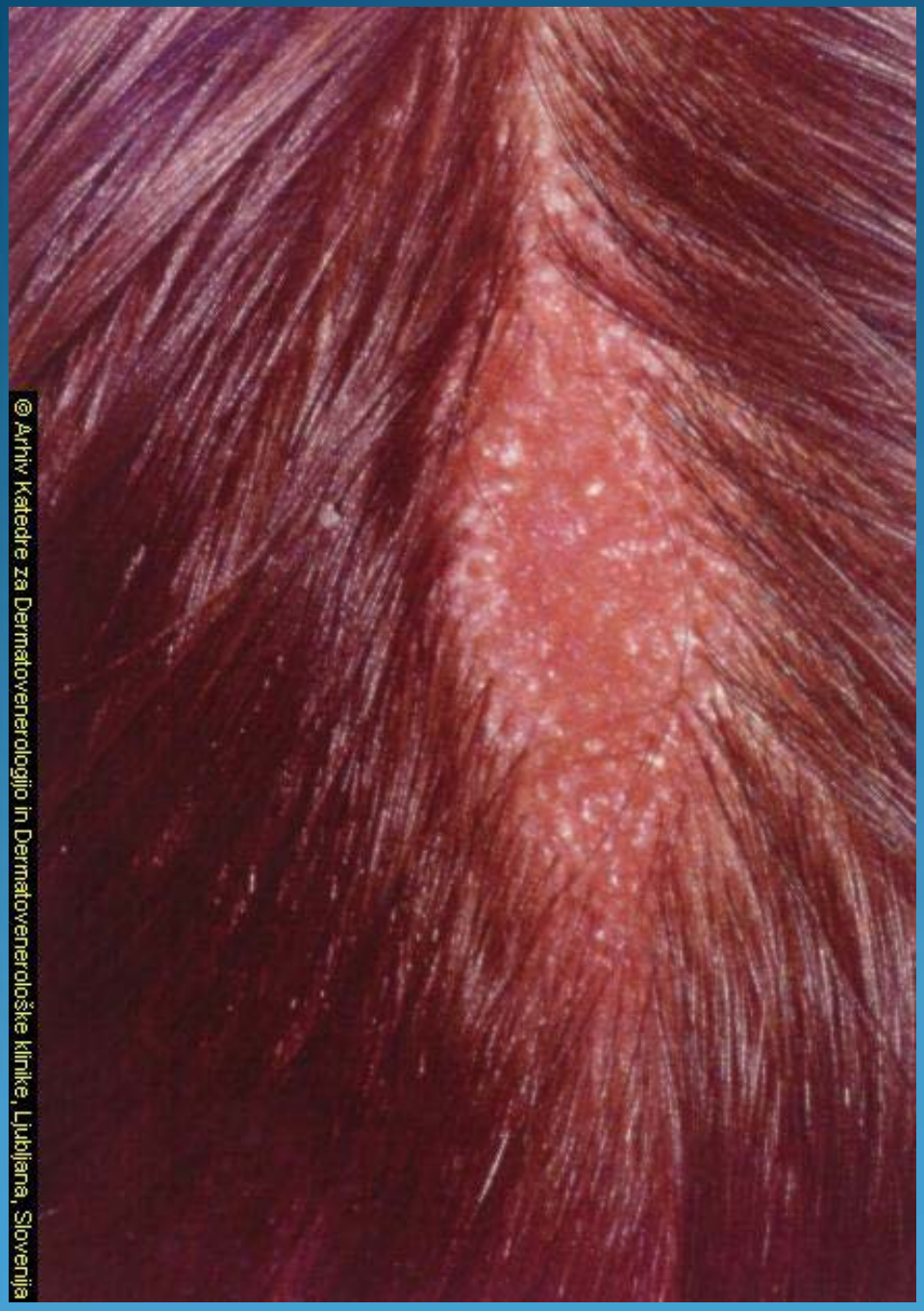
Central Centrifugal Cicatricial Alopecia

CCC Alopecia

- Histopath:
  - Characteristic premature desquamation of the inner root sheath
  - Perifollicular lymph infiltrate surrounding the upper portion of the follicle
- Management
  - Cessation of traumatic hair practices
  - Topical steroids
  - Tetracycline 500 mg BID

Alopecia Mucinosa

- Characterized by intrafollicular deposition of mucin (follicular mucinosus)
- Idiopathic and lymphoma related types
- All ages affected
- Lesions are often pruritic, dysesthetic, and/or anhidrotic
- Other body locations besides the scalp can be involved



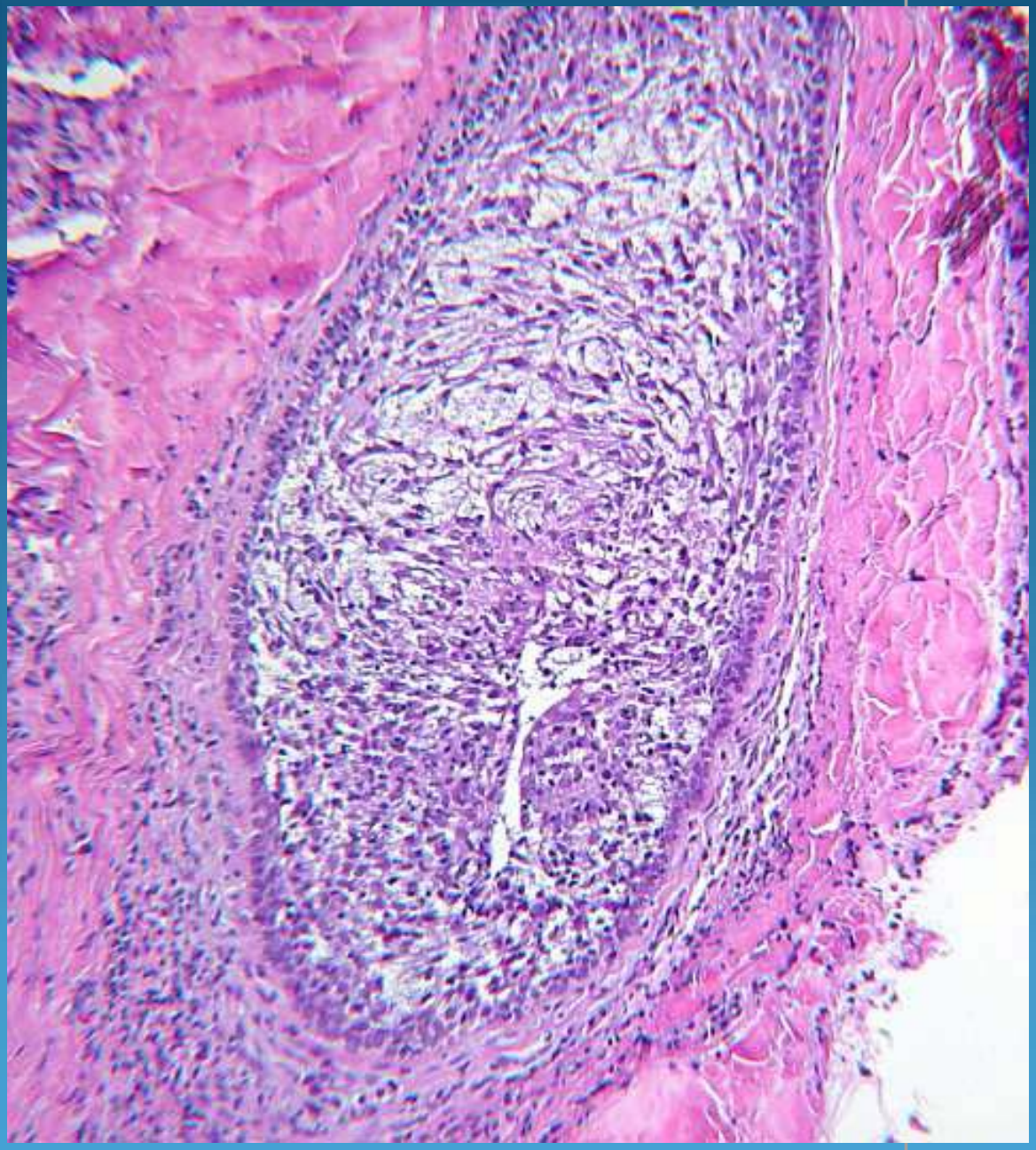
Alopecia Mucinosa

Alopecia Mucinosa

- Presentation of scalp disease is highly variable
- In adults, MF is associated 9-60% of the time
- In children, Hodgkin's lymphoma is the most commonly associated malignancy
- Alopecia mucinosa can present as a paraneoplastic phenomenon

Alopecia Mucinosa

- There are no reliable clinical or histo criteria to distinguish benign from malignant cases
- Histopath:
  - Intrafollicular mucin
  - Perifollicular lymphocytic infiltrate
  - No lamellar fibrosis



Management

- Close follow up and serial biopsies if progression of disease
- Lymph node examination at all visits
- Topical or intra-lesional steroids
- Minocycline 100 BID for 5-8 weeks
- Accutane 0.5-1.0 mg/kg for 4-5 months
- If colonized with Staph, treating with Abx may clear disease

Keratosis Follicularis Spinulosa Decalvans

- Aka keratosis pilaris decalvans
- related to KP atrophicans faciei and atrophoderma vermiculata
- X-linked or sporadic
- Follicular hyperkeratosis beginning on the face and spreading to involve other body areas, eventually leading to punctate atrophy
- Begins in infancy or childhood
- Also have photophobia and scarring alopecia

Management

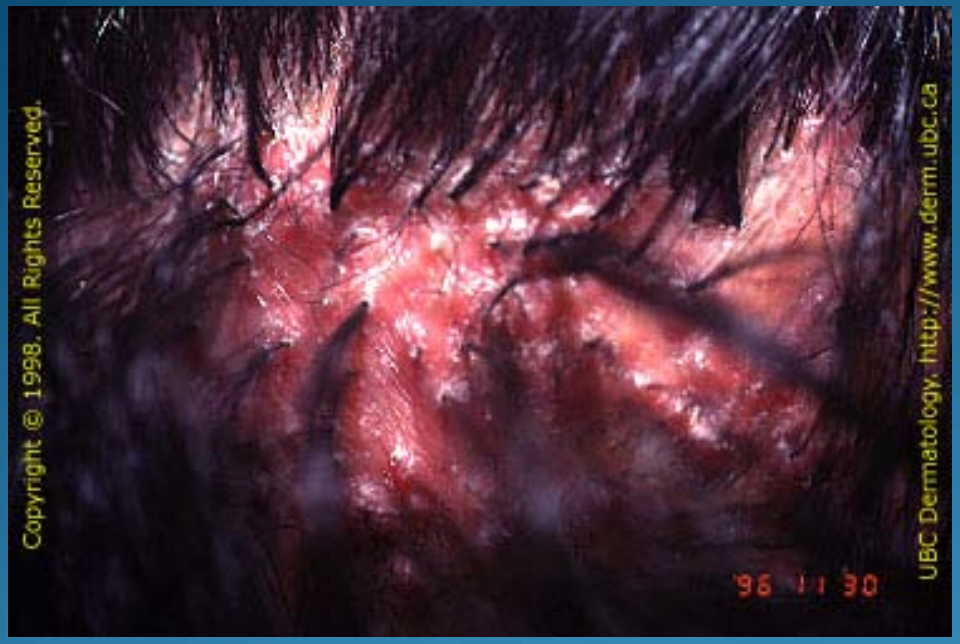
- Intervene early in childhood when the disease is active
- Treatment studies are limited
- High potency topical or intralesional steroids
- Accutane 0.5 mg/kg for 3 months
- Baseline and routine ophtho exams recommended

Neutrophilic Cicatricial Alopecias

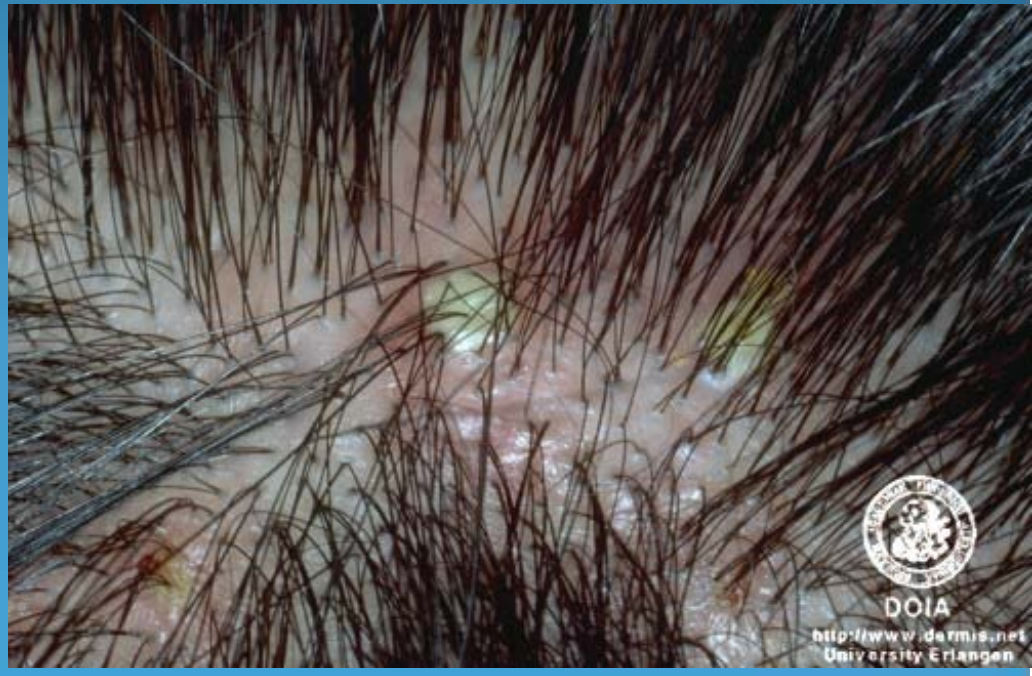
- Folliculitis Decalvans
- Dissecting Cellulitis

Folliculitis Decalvans

- Common form of primary cicatricial alopecia
- Is a destructive suppurative folliculitis seen in young and middle aged adults
- Staph aureus is thought the be the inciting factor
- Begin as grouped follicular pustules which evolve into abscesses and eventually scarring
- Often see tufted folliculitis



Folliculitis Decalvans



Folliculitis Decalvans

- Histopath:
  - Upper and mid follicular neutrophilic infiltrate
  - Late disease: granulomatous inflamm and perifollicular fibrosis

Management

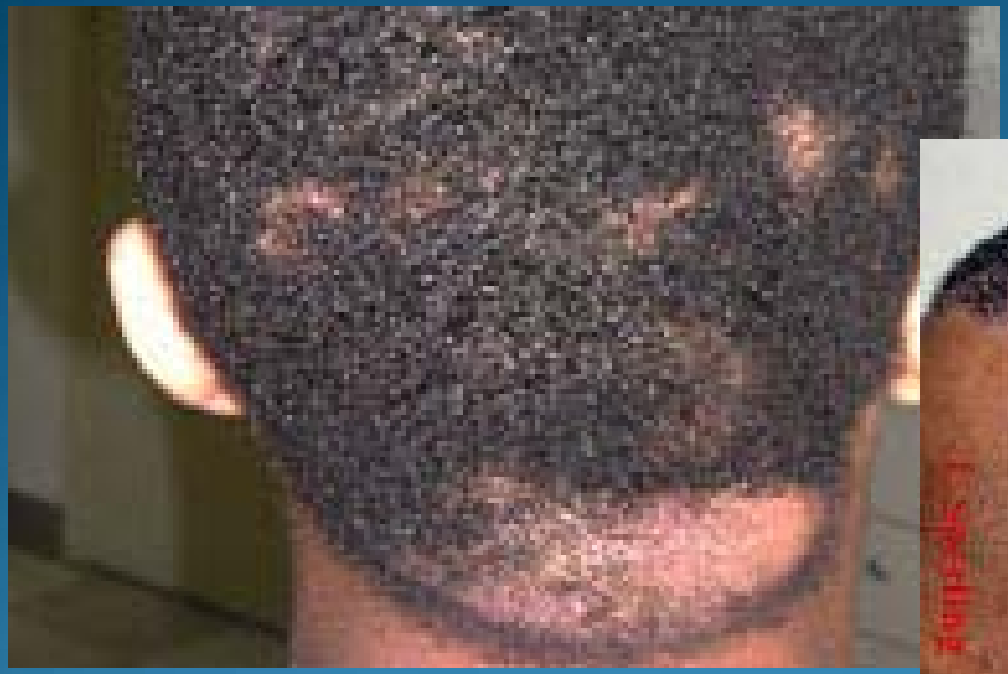
- Culture pustules
- Abx with anti-Staph coverage
- Rifampin 300 mg BID in combination with Clindamycin 300 mg BID for 10 weeks
- New combo of Rifampin, fusidic acid (not available in US) and zinc has shown good success
- Eliminate Staph carrier state with mupirocin

Dissecting Cellulitis

- Aka perifolliculitis capitis abscedens et suffodiens
- Part of the follicular occlusion triad (tetrad)
- Abnormal follicular keratinization leads to obstruction, secondary infection, and follicular destruction
- >80% of patients are black men ages 18-40

Dissecting Cellulitis

- Initial lesions are pustules often beginning on occipital or vertex scalp
- Later large,fluctuant nodules that coalesce and form tracts
- Coexisting acne conglobata or HS is a risk factor for development of spondyloarthropathy.
- Peripheral and axial joints may be involved



Dissecting Cellulitis

Dissecting Cellulitis

- Histopath:
  - Intra and perifollicular neuts
  - Abscesses in mid to deep dermis
  - Late: sinus tracts lined with squamous cells and fibrosis

Management

- Accutane now considered first line
  - 1 mg/kg/d for at least 4 months followed by 0.75-1 mg/kg/d for another 5-7 mo. if needed
- Other opitions
  - Topical clindamycin gel
  - Oral TCN
  - Dapsone or colchicine
  - CO_2 laser and surgical excision

Mixed Cicatrical Alopecias

- Acne Keloidalis
- Acne Necrotica
- Erosive pustular dermatosis

Acne Keloidalis

- Mostly seen in AA males
- Thought to be secondary to mechanical traumas, seborrhea, or infections
- Present as follicular papules or keloidal plaques on the occiput and nape of neck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Acne Keloidalis

Management

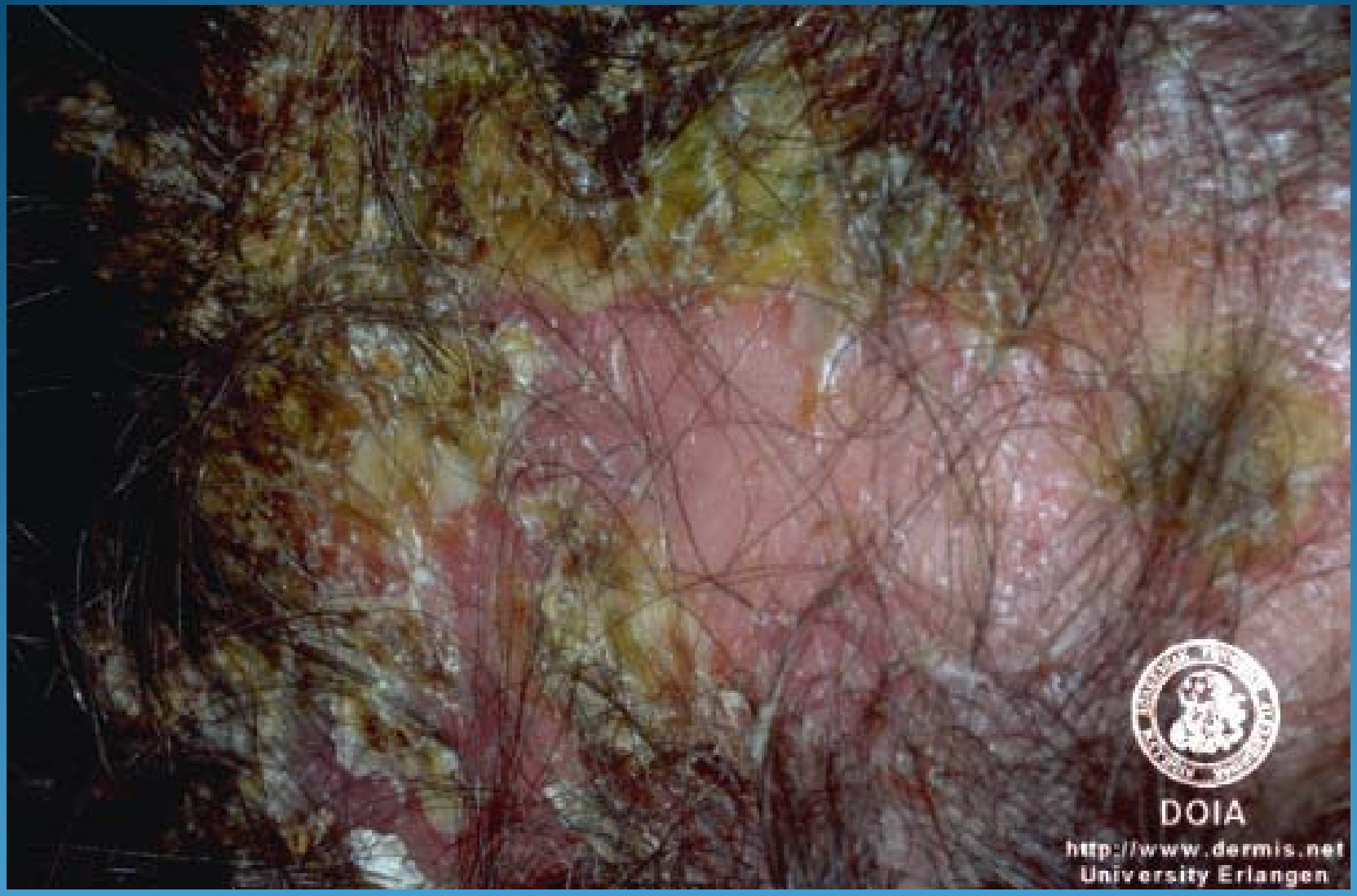
- Early, limited disease
  - Avoid mechanical traumas
  - Topical high potency steroids in combo with topical clinda
  - Intralesional kenalog (10mg/cc)
  - Oral TCN
- More extensive disease
  - Surgical excision

Acne Necrotica

- Aka Greer's disease
- Chronic, relapsing d/o of crops of pruritic, small pustules that undergo central necrosis and crusting
- Management
  - Cx pustules and tx accordingly
  - Antibacterial shampoos
  - Topical antibiotics
  - Topical steroids

Erosive Pustular Dermatosis

- Uncommon d/o that affects the elderly
- Most cases have a preceding trauma
- Characteristic lesion is large, asx, boggy crusted plaque that when unroofed reveals a beefy, red erosion with pustules
- Long standing lesions can develop BCC or SCC's



Erosive Pustular Dermatosis

Management

- Topical high potency steroids BID
- Dovonex cream BID
- Oral or topical abx prn

Adjunctive Therapies for all Types

- Hair pieces/wigs
- Hair color matched powders (Toppik, Spencer Forrest, Westport)
- Hair transplantation
- Scalp reduction