

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 Mucinososa
 - 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

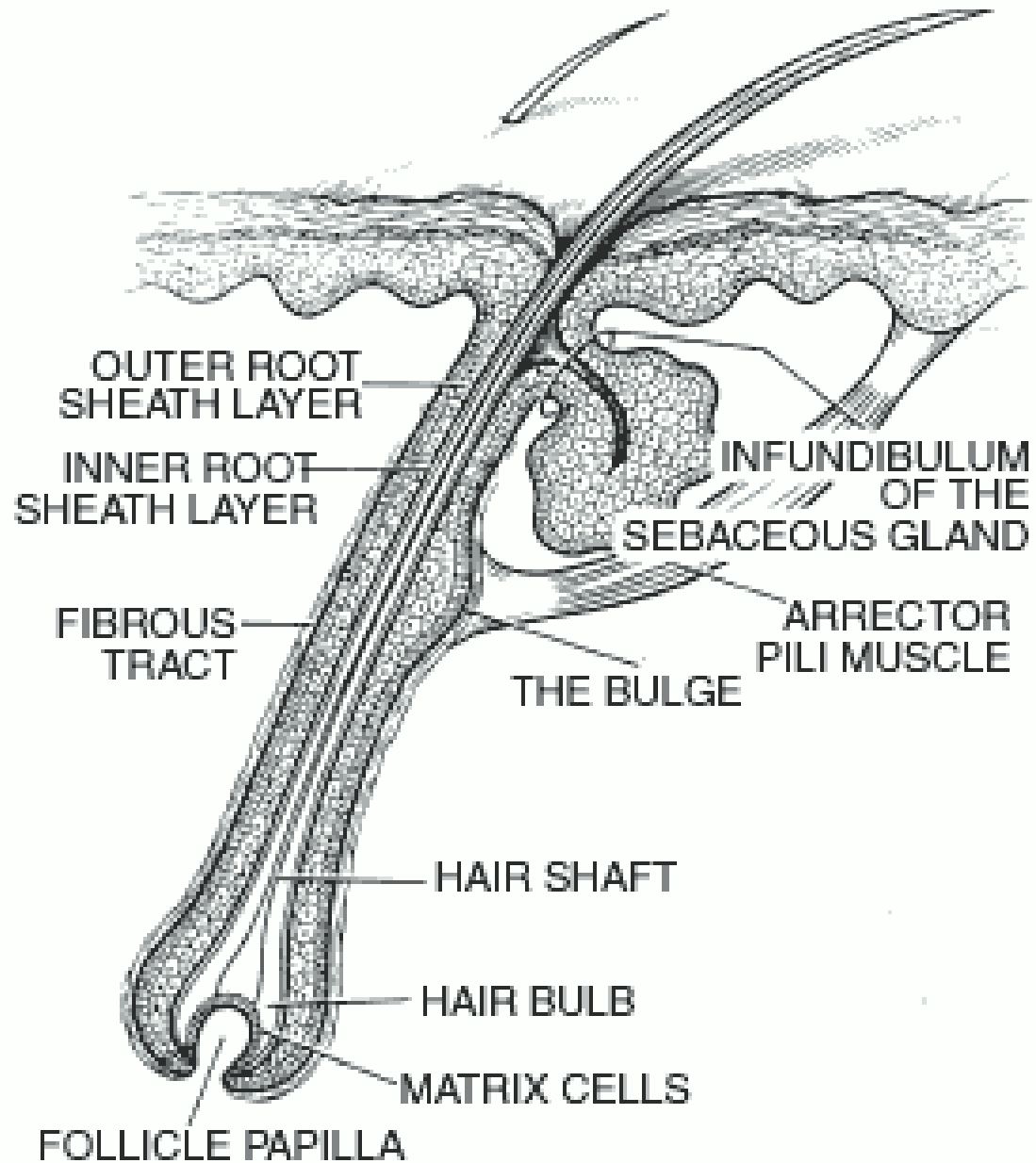
- NAHRS System (2001) based on primary inflammatory cell type
 - **Lymphocytic:** DLE, Lichen Planopilaris, Pseudopelade of Brocq, Central Centrifugal, Alopecia Mucinososa, Keratosis Fol. Spin. Decal.
 - **Neuts:** Folliculitis Decalvans, Dissecting Cellulitis
 - **Mixed:** Acne Keloidalis, Acne Necrotica

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



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 (2nd 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

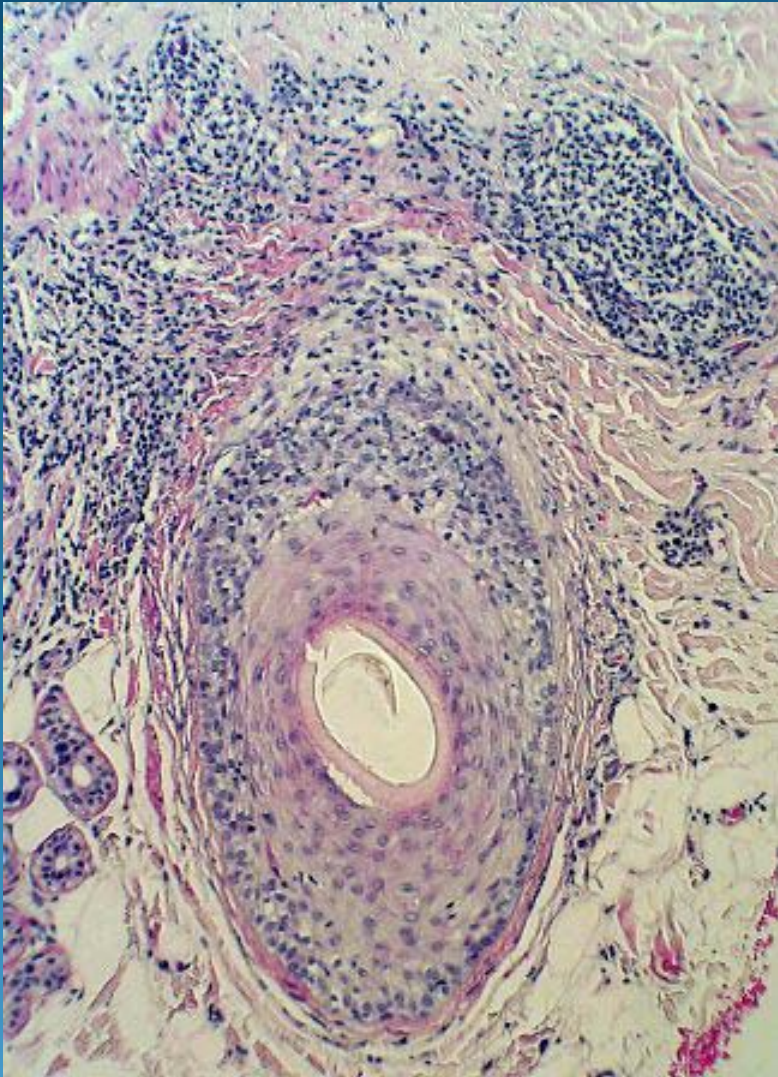
- Seen in adults, usually arising in middle-aged females
- Extracranial LP present in 17-50% of pts.
- Sx at presentation: shedding, hair loss, pruritus
- PE: perifollicular erythematous papules and spinous follicular hyperkeratosis. Can see unaffected hairs in scarred areas



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



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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?



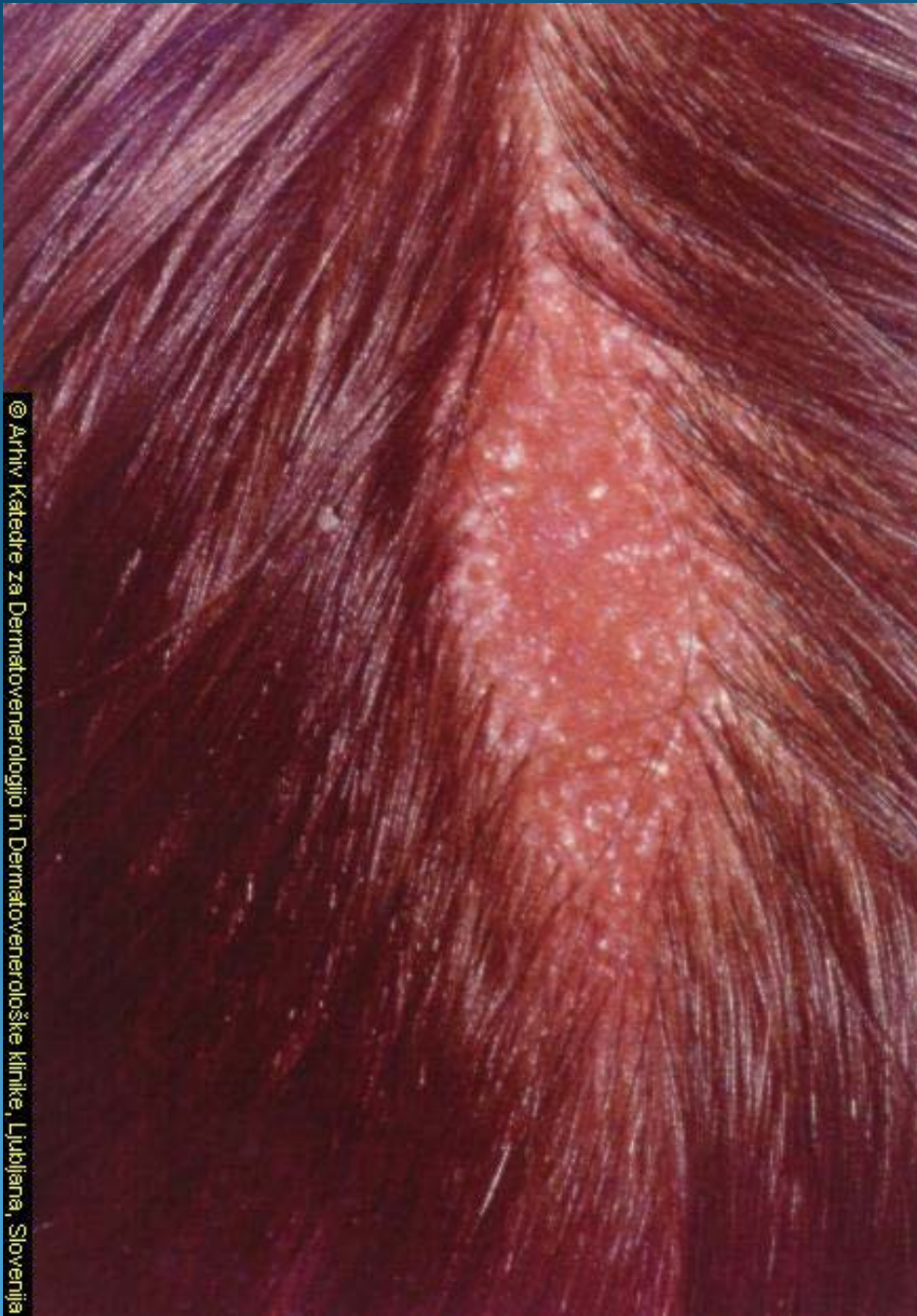
Central Centrifugal Cicatricial Alopecia

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 Mucinososa

- 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



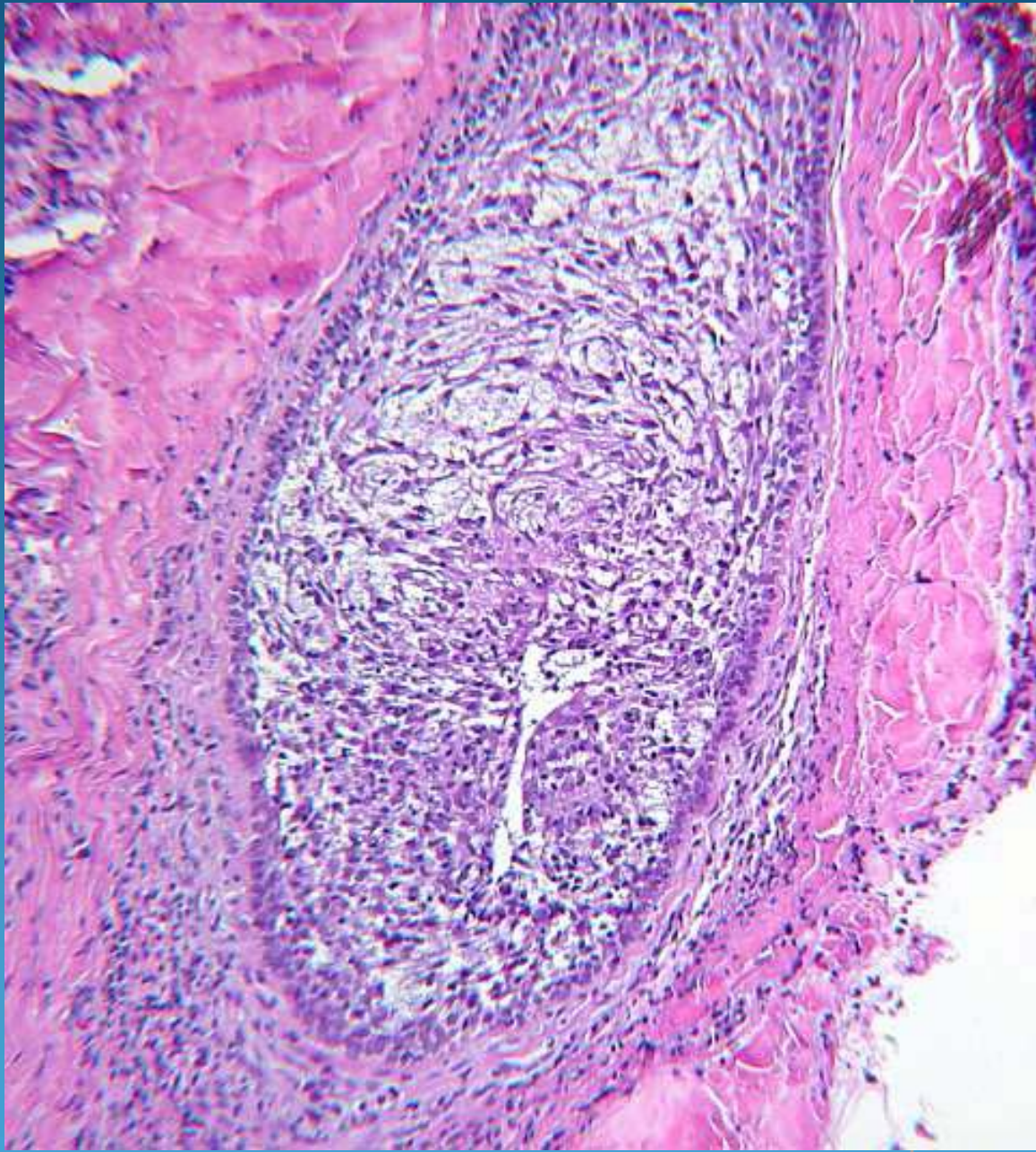
Alopecia Mucinoso

Alopecia Mucinososa

- 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 to 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 inflammation 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 options
 - Topical clindamycin gel
 - Oral TCN
 - Dapsone or colchicine
 - CO₂ laser and surgical excision

Mixed Cicatricial 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 Dermatitis

- 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 Dermatitis

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