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 Keloidalisis
  - 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
  - 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 hyperkeratosi
- 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 inflammation
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, non-scarring 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 ophthalmic 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

- Histopath:
  - Upper and mid follicular neutrophilic infiltrate
  - Late disease: granulomatous inflammatory 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 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
Acne Keloidalis
Management

- Early, limited disease
  - Avoid mechanical traumas
  - Topical high potency steroids in combo with topical clinda
  - Intralional 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