Scarring Alopecia

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Disclosures

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Off-label use of medication will be discussed
Challenges of scarring alopecia

- Rare diseases
- Making the correct diagnosis
- Monitoring treatment
- Dirth of effective treatment
  - Defining "effective"
- Time management
Objectives

- Define scarring alopecia
  - Primary vs. secondary
- Discuss an approach to accurate diagnosis of scarring alopecia
- Presentation of cases/diagnoses
- Review management of scarring alopecia
  - Early intervention is key
  - Goals to reduce symptoms, slow disease progression, and improve hair density

Scarring alopecia

• Permanent loss of hair (a “trichologic emergency”)
  • Most are indolent, but rapid progression can occur unexpectedly

• Primary scarring alopecia is due to a neutrophilic, lymphocytic, or mixed inflammatory infiltrate

• Secondary scarring alopecia may be due to:
  • Trauma or tumors (benign or malignant).
  • Morphea, late tinea capitis, and late traction or trichotillomania are considered secondary scarring alopecias as well
Scarring alopecia - primary

• **Lymphocytic**
  - *Central centrifugal cicatricial alopecia*
  - *Chronic cutaneous lupus erythematosus*
  - *Lichen planopilaris (frontal fibrosing alopecia, fibrosing alopecia in a pattern distribution, Graham-Little syndrome)*
  - Alopecia mucinosa
  - Keratosis follicularis spinulosa decalvans

• **Neutrophilic**
  - *Folliculitis decalvans*
  - *Dissecting cellulitis of the scalp*

• **Mixed**
  - Acne keloidalis
  - Erosive pustular dermatosis of the scalp

Diagnosing scarring alopecia

• History and physical
  • Timing/duration, presence of increased shedding, pattern of loss, associated symptoms
  • Trichoscopy – erythema, pili torti like hairs, scale
  • Erythema, scale – diffuse or perifollicular, greasy or adherent, presence or absence of vellus hairs along hairline especially
  • Hair pull test – may not be markedly positive, but **anagen hairs on gentle pull**

• Histopathology
  • Two **4mm** punch biopsies, one for horizontal sectioning and one for vertical sectioning. Consider direct immunofluorescence or tissue culture
  • Site of biopsy matters – try for an active edge, perhaps more of a subacute location than a very active location
  • Type of inflammation, location of inflammation, perifollicular fibrosis, loss of sebaceous glands
Central centrifugal cicatricial alopecia

• Thought to affect at least 50% of African-American women by the age of 50
  • Some familial cases reported
  • Can happen in men and non-black individuals
• Unclear association with hair care practices
  • Genetic association has been found
  • Possible association with fibroproliferative diseases including uterine leiomyomas
• Lymphocyte-predominant on path
  • May be CD4 predominant vs. CD8 predominance in LPP

Central centrifugal cicatricial alopecia – clinical features

• May be symptomatic, especially in acute phase
  • Itch, tenderness, erythematous papules or pustules
• Broken off hairs/breakage can be an early sign
• Often with a marginal component or traction alopecia
• Trichoscopy -
  • Peripilar halos, possible erythema, pinpoint white macules

Lichen planopilaris

- Inflammatory, lymphocyte-predominant primary cicatricial alopecia
- Up to 50% of patients may have lichen planus on body or in mouth
- Pruritus and tenderness are common
- May have allergic contact dermatitis
  - 76% of pts with FFA or LPP relevant positive patch test results
- Can be refractory to treatment

Lichen planopilaris – clinical features

- Irregular patchy alopecia with loss of follicular ostia
- Perifollicular erythema and scale
- Diffuse presentation possible
- Biopsy shows interface lichenoid dermatitis of the follicular epithelium
  - Variably dense lymphocytic infiltrate surrounding the infundibulum, isthmus, and bulge region
  - Loss of sebaceous glands and perifollicular lamellar fibrosis

Chantal Bolduc, Leonard C. Sperling, Jerry Shapiro, Primary cicatricial alopecia: Lymphocytic primary cicatricial alopecias, including chronic cutaneous lupus erythematosus, lichen planopilaris, frontal fibrosing alopecia, and Graham-Little syndrome, Journal of the American Academy of Dermatology, Volume 75, Issue 6, 2016, Pages 1081-1099,
Frontal fibrosing alopecia

• Variant of lichen planopilaris
• Typically in postmenopausal women, though cases of younger women and men do occur
• Increasing in incidence
• Unlike LPP, often without symptoms
• May lose eyebrows and body hair
• Different clinical patterns – linear, zig zag, pseudo-fringe
• Loss of vellus hairs, facial papules
• Very slowly progressive

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Chronic cutaneous lupus

- Erythema, atrophy, follicular plugging, mottled hyper and/or hypopigmentation
- May see classic discoid lesions, tenderness, itch
- Predominantly in women between ages 20 and 40 years
- 5-10% may progress to SLE over 5 or more years
- DIF may be helpful – needs to be an active lesion, ~2-3 months old, no treatment for 3 weeks.
- ANA positive in 15-45% of patients
- Strict sun protection!!

  - Consider *polypodium leucotomos* in addition to UPF hats and SPF

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Folliculitis decalvans

- Neutrophilic primary cicatricial alopecia
- Male predominance, middle aged
- Typically vertex and occipital scalp
- Painful or pruritic papules, pustules, crusts
- Indurated or thickening of scalp with tufted hairs
- May represent an abnormal immune response to *Staphylococcus aureus*
- Polytrichia, intra and perifollicular neutrophils, mixed inflammation upper half of dermis

Perifolliculitis capitis abscedens et suffodiens (dissecting cellulitis of the scalp)

- Boggy, VERY TENDER, suppurative nodules/sinus tracts
- Young men, often of African descent
- Part of follicular occlusion tetrad, though often seen alone
- Uncommonly associated with a seronegative arthritis
- Consider tissue culture to rule out infectious etiology, especially tinea capitis
- Histopathology:
  - Early – deep peribulbar and subfollicular lymphocytic infiltrate, intact sebaceous glands
  - Established fluctuant nodules – deep perifollicular and lower dermal abscesses with mixed inflammation, increased catagen/telogen hairs
  - Late stage – granulation tissue, epithelial-lined true sinus tracts with loss of seb glands

Treatment of scarring alopecia

• Goals of treatment are to:
  1. Reduce symptoms of pain and itch (if present)
  2. Slow the progression of hair loss
  3. Maintain the hair that is present as much as possible (treat underlying androgenetic alopecia)

• Regrowth sometimes, but uncommon and should not be the primary goal
• Multiple treatments/medications are often necessary
• It may take 6 months or more to determine any benefit
Treatments

- Scalp directed
- Inflammation directed
- Hormone directed
- Hair directed
Scalp directed treatments

• Treat underlying seborrheic dermatitis and/or folliculitis
  • Zinc pyrithione, selenium sulfide, ketoconazole, salicylic acid shampoos
• Consider patch testing
  • Special consideration regarding possible association between personal care products and FFA
  • Consider avoiding chemical sunscreens and those containing titanium dioxide
• Ensure adequate scalp health – frequency of hair washing especially
Treatments targeting inflammation - topicals

Lymphocytic

• Topical and/or intralesional corticosteroids – 10 mg/cc
  • Caution along frontal hairline
• Topical calcineurin inhibitors – can be compounded into solution, but ointment or cream may be used
  • Alternate with corticosteroids
• Topical metformin solution or cream 10%

Neutrophilic

• Topical antimicrobials
  • OTC benzoyl peroxide, chlorhexidine, dilute sodium hypochlorite, zinc pyrithione
  • Clindamycin, minocycline, dapsone
• Topical and/or intralesional corticosteroids – up to 40 mg/cc
Treatments targeting inflammation - systemic

**Lymphocytic**
- Doxycycline or minocycline 100 mg one to two times daily
- Hydroxychloroquine 5 mg/kg/day
- Pioglitazone 15 mg daily
- Mycophenolate mofetil, methotrexate, cyclosporine, azathioprine
- Janus kinase inhibitors
- Cetirizine, omalizumab, apremilast, low dose naltrexone (1.5-4.5 mg), isotretinoin

**Neutrophilic**
- Doxycycline or minocycline 100 mg twice daily
- Clindamycin and rifampin 300 mg each twice daily
- Other antibiotics (ciprofloxacin, trimethoprim-sulfamethoxazole)
- Dapsone (50-150 mg daily)
- Isotretinoin (lower dose, longer course)
- TNF inhibitors or other biologics
Hair directed and hormone targeted therapies

• Hair growth promoters
  • Topical minoxidil 2% (women's) or 5% solution (men's) or foam (men's or women's))
  • Low dose oral minoxidil (0.25 mg to 5 mg daily)
  • Topical bimatoprost 0.03% or latanoprost 0.005%-0.5%
  • Low level laser technology - Laser comb, band, or cap
  • Platelet rich plasma or fibrin therapy

• Antiandrogen therapy
  • Spironolactone (young women, or women with h/o breast cancer)
  • Finasteride 5 mg, dutasteride 0.5 mg (postmenopausal) - more for FFA

• Hair restoration surgery – when medically controlled, no further signs of inflammation
Treatment algorithms - CCCA

• Topical corticosteroid and/or topical calcineurin inhibitor
• Topical minoxidil (oil available OTC or compounded) or oral minoxidil
• If/when symptomatic with active inflammation
  • Intralesional corticosteroids
  • Doxycycline or minocycline 100 mg twice daily x 3 months
Treatment algorithms - LPP

• Topical corticosteroid shampoo +/- solution or oil prn symptoms and alternating with topical calcineurin inhibitor
• Intralesional corticosteroids
• Hydroxychloroquine
• Topical or oral minoxidil
Treatment algorithms – frontal fibrosing alopecia

- Finasteride 5 mg daily in women; dutasteride 0.5 mg daily in men
- Pimecrolimus cream or tacrolimus ointment alternating with clobetasol shampoo or topical corticosteroid
- Consider intralesional corticosteroid injections if not too much baseline atrophy or if notable perifollicular scale or symptoms
- Minoxidil topical foam or oral (0.625-1.25 mg) depending on patient preference
- Escalate to dutasteride 0.5 mg daily in women and add hydroxychloroquine if needed
- Other anti-inflammatory treatments as needed
Summary

• Cicatricial (scarring) alopecias are a trichologic emergency
• Accurate diagnosis is important for treatment, though treatment options overlap and are similar across diagnoses
• Goals of treatment are to slow progression of disease and manage patient’s symptoms
Thank you!

Feel free to email with questions: cgoh@mednet.ucla.edu

www.scarringalopecia.org