Central Centrifugal Cicatricial Alopecia (CCCA): From Fashion to Family History

Yolanda M. Lenzy, MD, MPH
Clinical Associate
University of Connecticut
Lenzy Dermatology & Hair Loss Center
ylenzy@lenzyderm.com

American Academy of Dermatology
Washington, DC
March 2, 2019
Disclosures

Board of Directors, CARF
Ethnic Hair Disorders

- Still new area (Pubmed):
  - CCCA: 97 (in 50 years)
  - Traction Alopecia: 143
  - Dissecting Cellulitis: 139
  - Pseudofolliculitis Barbae: 110
  - Acne Keloidalis Nuchae: 79

- Compared to:
  - Psoriasis: 47,528
  - Acne: 17,216
  - AA: 4,106
  - FFA: 328 (+103)
Central Centrifugal Cicatricial Alopecia (CCCA)

-Most common cause in AAF

Initially called “hot comb alopecia” in 1968¹
- Caused by scalp burns
- Diagnosed in people who never used heat

Coined as “CCCA” by North American Hair Research Society²

Likely multi-factorial:
- Genetics
- Curly nature of follicle
- Hair grooming practices
  - Traction-sew-in weaves, braids
  - Chemicals

CCCA: Hereditary

Family 1

A) 63 y/o daughter: loose braids on natural hair most of the time
B) 92 y/o mother: no history of chemical or mechanical grooming
C) 65 y/o daughter: occasional use of relaxers four times a year
D) 55 y/o daughter: braids on natural hair most of the time
E) 45 y/o daughter: chemical relaxers every 2–3 months with occasional loose braiding

Family 2

A) 35 y/o mother: h/o chemical relaxers, tight braids w/ extensions, and sewn-in weaves
B) 11 y/o daughter: no history of any chemical or mechanical trauma

16 y/o (4 year duration)
- + Family Hx of CCCA:
  - Mother & MGM
  - Asymptomatic

15 y/o (2 year duration)
- Family Hx of CCCA
  - Mother
  - Tender Papules
Fibroproliferative genes are preferentially expressed in central centrifugal cicatricial alopecia

Crystal Aguhi, MD,a Ycmisi Dina, BS,b C. Conover Talbot, Jr, BS, and Luis Garza, MD, PhD,a Baltimore, Maryland, and Nashville, Tennessee

- 5 patients bx-proven CCCA
- 2 specimens: vertex & unaffected occipital scalp
- Microarray analysis: gene expression patterns
- Upregulation of genes implicated in Fibroproliferative Disorders in the CCCA affected vs unaffected scalp: PDGF, COL1/3, MMPs
- Significant overlap in pathways for CCCA & atherosclerosis hepatic fibrosis
Variant PADI3 in Central Centrifugal Cicatricial Alopecia

Liron Malik, M.Sc., Ofer Sarig, Ph.D., Maria-Teresa Romano, M.Sc., Marie-Claire Méchin, Ph.D., Alon Peled, B.Med.Sci., Mor Pavlovsky, M.D., Emily Warshauer, M.D., Liat Samuelov, M.D., Laura Uwakwe, M.D., Valeria Briskin, Ph.D., Janan Mohamad, B.Med.Sci., Andrea Gat, M.D., Ofer Isakov, M.D., Ph.D., Tom Rabinowitz, B.Med.Sci., Noam Shomron, Ph.D., Noam Adir, Ph.D., Michel Simon, Ph.D., Arvy McMichael, M.D., Ncoza C. Dlova, M.B., Ch.B., F.C.Derm., Ph.D., Regina C. Betz, M.D., and Eli Sprecher, M.D., Ph.D.

- **Discovery Set**: 16 CCCA bx proven patients exome sequencing
  - 1 splice site & 3 heterozygous missense mutations in PADI3 in 5 patients (31%)
    - ↓PADI3 expression → protein misfolding
- **Replication Set**: 42 patients direct sequencing
  - PADI3 in 9 patients (21%)
- Prevalence of PADI3 mutation higher in CCCA patients than in control cohort of women of African ancestry (p=0.002)
PADI3 (Peptidyl Arginine Deiminase, type 3): enzyme responsible for post-translational modification of proteins essential for hair shaft formation

- PADI also implicated in Uncombable Hair Syndrome
  - Altered post-translational modification of Trichohyalin
Breakage: A forme fruste of CCCA

- 9 cases of hair breakage with or without scalp symptoms presented as sign of early CCCA
- 50 y/o 6 mo intense scalp burning

Diagnostic Approach

1. Detailed history
2. Scalp examination
3. Photography
4. Dermoscopy
5. +/- Biopsy
   - DIAGNOSIS & Staging
6. Treatment & Monitoring Q2-3 mos

A Retrospective Review of Treatment Results for Patients With Central Centrifugal Cicatrical Alopecia

Ariana Eginli BA, Emily Dothard MD, Courtney W. Bagayoko MD, Karen Huang MS, Alyssa Daniel MD, and Amy J. McMichael MD
15 subjects with CCCA
Before & After Treatment
Blinded Comparisons
- 7-8 Intralesional Steroid Injections
- Class I/II Topical Steroid
- +/- Minoxidil
- +/- Anti-Dandruff Shampoo

Median change in severity score:
- 5/15 Decreased severity score
- 2/15 No change in severity
- 8/15 Increased severity score
CCCA Treatment

Treatment Goals:
- Decrease inflammation & symptoms
- Stop expansion
- Enhance growth

Counseling
- Set realistic expectations

My approach:
1. Doxycycline x3 months (Probiotic)
   - Plaquinil
   - Low Dose Naltrexone
   - Kenalog injections 7.5mg/cc Q 4-6 wks x3-6 treatments
2. Class 1 topical steroid QOD
3. Minoxidil 5% foam daily
   - Compound: Clobetasol & 12% Minoxidil in Argon cream base
   - Nutraceutical
     - Viviscal Pro
     - Nutrafol
     - Platelet Rich Plasma

Maintenance Plan
- Potent topical steroid 1-2 times weekly
- Camouflage techniques

Pre-Treatment: 7/2012
Topicals only: 11/2012
Stage 2B
Regrowth!!!

Off/On topicals: 1/2014
Stage 3B

After 6 injections & Topicals:
11/2015
Stage 2B
Importance of Maintenance Treatments

12/2014  
No Treatment  

21 months  

9/2016  

Stage 2B  
Stage 3B
“Going Natural”

- Natural Hair Revolution
- Learn the language
- 2 methods:
  - Transitioning
  - Big Chop → TWA
- Low maintenance
- Exercise Friendly
- Professional acceptability

Natural Hair Revolution

Learn the language

2 methods:

- Transitioning
- Big Chop → TWA

Low maintenance

Exercise Friendly

Professional acceptability

CARF Communiciqué

A Dermatologist’s Experience of Going Natural

By Yolanda M. Lenzy, MD, Medical Director, Lenzy Dermatology & Hair Loss Center

After a recent Facebook post about my decision to stop straightening my hair and “go natural” this summer, I was asked to share with the CARF community about my decision and my journey. Yes, a bit back story. Three years ago, we relocated from Boston to small town in western Massachusetts. While all situations come with their own set of stressors, it was the change in environment and the obvious increase in humidity that most affected my hair. The weather in our new home was quite different from the climate I had grown accustomed to in the Northeast. My hair seemed to have a mind of its own, becoming more frizzy and unmanageable. I began researching different techniques to manage my natural hair, and I discovered the low-maintenance style of TWA (Very Short Haircut). This style, which involves cutting the hair very short and allowing it to grow out naturally, appealed to me because it was manageable and didn’t require frequent visits to the hairdresser. I decided to give it a try and cut my hair very short, leaving only a few inches in length. Initially, I was nervous about the change, but I was excited to see how my hair would look and feel.

As a dermatologist, I also wanted to share my personal experience with going natural. I believe in the power of self-care and the importance of embracing one’s natural beauty. I find that it’s crucial to make time for yourself and take care of your body. I strive to live a healthy lifestyle, both physically and mentally. I believe in the importance of exercise, healthy eating, and stress management. I also believe in the power of positive thinking and the importance of setting realistic goals. I strive to lead a balanced and fulfilling life.

As you can see, I am committed to living a natural lifestyle and I am a strong advocate for embracing one’s natural beauty. I hope that my personal experience can inspire others to embrace their natural hair and live a healthy, fulfilling life.

CARF International

www.carfintl.org
Need larger studies

- 80% of Black women have relaxed

**BWHS: Ongoing US prospective cohort 59,000 black women since 1995**
- Every 2 yrs participants new questionnaire

- Asked questions about relaxer use in 1997
  - BWHS published Increased risk of **uterine fibroids** with relaxer use (2012)
  - Approached in 2011 to make the case
  - Hair Loss in 2013 & 2015 questionnaires

- **Prevalence/Risk Factor study**

Women’s Dermatological Society Career Development Award
CARF (Cicatricial Alopecia Research Foundation)
- Support groups
- Patient Education Materials

www.carfintl.org
Questions?

LenzyDerm.com/Book
ylenzy@lenzyderm.com

Connect with me:
@dryolandalenzy