Elastin staining patterns in primary cicatricial alopecia

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Background: Most biopsy specimens of cicatricial (scarring) alopecia can be readily subclassified as lymphocytic versus neutrophilic, but specific diagnosis remains difficult, particularly when a late stage of the disease is sampled.

Objective: We sought to document patterns of scarring highlighted by elastic tissue staining in primary cicatricial alopecia.

Methods: We documented Verhoeff elastic van Gieson staining patterns in 58 routinely embedded (vertical) biopsy specimens of cicatricial alopecia. Patterns of fibrosis included perifollicular (wedge-shaped vs broad tree trunk–shaped) and diffuse. The patterns were compared against the diagnosis obtained by independent expert clinical review, including central centrifugal cicatricial alopecia (CCCA), lichen planopilaris, traction alopecia, frontal fibrosing alopecia, discoid lupus erythematosus, and tufted folliculitis.

Results: Wedge-shaped perifollicular fibrosis was seen in lichen planopilaris but also in CCCA. Broad tree trunk–shaped perifollicular fibrosis was most commonly encountered in CCCA.

Limitations: The retrospective nature of the study precluded temporal staging of the disease process.

Conclusions: Patterns of fibrosis highlighted by elastin staining in primary cicatricial alopecia appear to be disease specific. Superficial wedge-shaped perifollicular fibrosis is associated with but may not be specific for lichen planopilaris. Broad tree trunk–like perifollicular fibrosis is specific for CCCA but not present in many cases. Elastin staining represents a useful ancillary study for the evaluation of late-stage scarring alopecia in routinely oriented punch biopsy specimens. (J Am Acad Dermatol 2013;69:776-82.)

Key words: cicatricial alopecia; dermatopathology; elastin; lichen planopilaris; scarring alopecia; traction alopecia.

Alopecia is among the most challenging areas of medical dermatology. The substantial morbidity associated with alopecia is reflected in the perception that a typical dermatology office visit for alopecia entails a significantly greater amount of time devoted to patient counseling compared with most dermatology office visits (if the patient is to deem the encounter satisfactory).[1] When a biopsy is warranted, the distinction between nonscarring and scarring (cicatricial) alopecia is usually straightforward. However, specific histologic diagnosis of cicatricial alopecia is often elusive. Most

Abbreviations used:

CCCA:	central centrifugal cicatricial alopecia
DLE:	discoid lupus erythematosus
EVG:	Verhoeff elastic van Gieson
FFA:	frontal fibrosing alopecia
H&E:	hematoxylin-eosin
LPP:	lichen planopilaris

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biopsy specimens can be readily classified as lymphocytic or neutrophilic based on the North American Alopecia Working Group classification for primary cicatricial alopecia.[2] Mirmirani et al[3] reported that histopathologic features fail to achieve specific histologic diagnosis of primary cicatricial alopecia (excluding discoid lupus). Whiting and Olsen[4] wrote: "The end-stage changes [in central centrifugal cicatricial alopecia] are indistinguishable from those in other primary scarring alopecias, such as lichen planopilaris, discoid lupus erythematosus, and folliculitis decalvans." However, prior studies have used elastin staining to characterize cicatricial alopecia. In 1978, Pinkus[5] described patterns of elastic fibers in alopecia, noting that discoid lupus erythematosus (DLE) was associated with diffuse (perifollicular and interfollicular) loss of elastin. Elston et al[6] described patterns of scarring in established lesions of cicatricial alopecia, including wedge-shaped perifollicular fibrosis in lichen planopilaris (LPP). Wedge-shaped perifollicular fibrosis was also noted in the series of LPP reported by Bergfeld and coworkers.[7] These patterns may be appreciated using fluorescence microscopy of routine (ie, vertically oriented) hematoxylin-eosin (H&E)-stained sections.[8] Although these patterns continue to be reported in recent reviews,[9,10] the conclusions are based on relatively few cases. Moreover, most studies describing histopathologic features of cicatricial alopecia do not document independent diagnostic confirmation. We investigated the diagnostic use of elastin staining in different forms of primary cicatricial alopecia. To our knowledge, this study is the largest to date and the only study to date documenting the correlation of patterns of fibrosis with independent, blinded diagnosis by expert clinical case review.

CAPSULE SUMMARY

- Specific diagnosis of primary cicatricial alopecia based on histopathology alone is frequently impossible.
- Elastin staining highlights distinctive patterns of fibrosis in primary cicatricial alopecia.
- Elastin staining represents a useful ancillary study for the evaluation of late-stage scarring alopecia in routinely oriented punch biopsy specimens.

METHODS

Cases were identified from 2003 through 2010, a period when elastin stains were not used in our practice for the assessment of alopecia. Searches for the following phrases were performed: "scarring alopecia," "lymphocyte-mediated scarring alopecia," "lymphocytic scarring alopecia," "neutrophil-mediated scarring alopecia," "neutrophilic scarring alopecia," "cicatricial alopecia," "central centrifugal scarring alopecia," "follicular degeneration syndrome," "lupus erythematosus," "discoid lupus erythematosus," "lichen planopilaris," "frontal fibrosing alopecia" (FFA), "pseudopelade," "traction alopecia," "folliculitis decalvans," "dissecting cellulitis," and "acne keloidalis." Ten patients with scar were identified through a search for "scar" and "hypertrophic scar." The study was approved by institutional review boards (University of California, Davis, Health System, Kaiser Permanente Northern California).

Inclusion criteria included: (1) routinely oriented punch biopsy specimen with a diagnosis of cicatricial alopecia listed in the line diagnosis or note of the pathology report from a board-certified dermatopathologist (M. A. F., K. L. B., T. H. K.); (2) diagnosis obtained by independent expert clinical review by a board-certified dermatologist with fellowship training in disorders of the hair (P. M.) who reviewed electronic medical records and in some cases personally treated the patient; and (3) availability of tissue to perform elastin staining. Exclusion criteria included: (1) availability of only transversely sectioned biopsy specimens; (2) absence of specific diagnosis on expert clinical review; and (3) insufficient tissue or nonrepresentative (ie, nonlesional) sections. Diagnoses were based on criteria outlined by the North American Alopecia Working Group.[2]

Verhoeff elastic van Gieson (EVG) stain and a concurrent H&E-stained recut was performed at the time of study. Slides were interpreted at a multiheaded microscope in groups of 2 to 5 observers, in all cases including the senior investigator (M. A. F.) and at least 1 other board-certified dermatopathologist (T. H. K., K. L. B.). Interpretation of the slides and clinical records occurred independently. EVG-stained sections were viewed at scanning magnification. Ten surgical scars of known duration (range, 2 months to >10 years) were also stained with EVG and H&E.

Patterns of fibrosis demonstrated by loss of elastin on EVG-stained sections were classified into 2 groups: perifollicular and diffuse (perifollicular and interfollicular). The perifollicular group was subclassified into superficial wedge-shaped versus broad tree trunk–like patterns of fibrosis, as previously described.[6] Discrepancies in perceived patterns of scarring were resolved by additional concurrent review and/or consensus. Cases where