An Atlas of HAIR PATHOLOGY WITH CLINICAL CORRELATIONS
THE ENCYCLOPEDIA OF VISUAL MEDICINE SERIES

An Atlas of HAIR PATHOLOGY WITH CLINICAL CORRELATIONS

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This textbook is the first comprehensive presentation of the microscopic pathology of hair disease. The subject is an area of great difficulty for pathologists and dermatopathologists who must evaluate specimens submitted with the clinical diagnosis of hair disease. The pathology of hair disease is rarely or very superficially taught during residency training. Many pathologists are inexpert at evaluating ‘hair biopsy specimens’, and often the submitting dermatologist must attempt to act as pathologist as well as clinician.

The book is intended to serve as a primer, an atlas and a reference text. As a primer, the book reviews basic information, including hair anatomy and the ‘nuts and bolts’ of processing and evaluating specimens. The author assumes that the reader knows very little about hair disease or hair pathology, and so a step-by-step approach is utilized. As an atlas, the book is rich in photographs demonstrating both basic and advanced histological features of hair disease. As a reference, the book includes the most up-to-date information about the pathology of hair disease presented in a synopsis format. Basic clinical features are also reviewed to provide clinical—pathological correlation.

I expect that dermatopathologists will be the most enthusiastic audience for the book. However, general pathologists and dermatologists who care for patients with hair loss will also find the book to be useful.

The ‘data source’ for the book is largely my collection of histological specimens and clinical photographs, although several dermatopathologists were kind enough to lend me some useful slides. The majority of my specimens were obtained from my own patients, many of whom I have known for years. This allows for very good clinical-pathological correlation. A few figures are based on material borrowed from other specialists in the field.

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Acknowledgements

My interest in the pathology of hair disease was stimulated by Dr John T. Headington’s seminal article on the subject (Headington J. Transverse microscopic anatomy of the human scalp. Arch Dermatol 1984; 120:449–56). Dr Headington was neither the first to utilize transverse sectioning nor the first to describe the histological features of hair disease. However, his teachings on the subject have had such a profound influence on enthusiasts of hair disease that Dr Headington can be regarded as the ‘father of hair disease pathology’.

I have been fortunate to follow in the footsteps of many pioneers in the field. Many of the concepts and observations described in the text are not my own, but have been passed down to us by such ‘giants’ as David Whiting, Vera Price, Wilma Bergfeld, Al Solomon, Elise Olsen, Rodney Sinclair and Rodney Dawber. David Whiting, in particular, has been a mentor and ‘cheerleader’ for much of my work. I have also received support and encouragement from many other leaders in the field of hair disease such as Jerry Shapiro, George Cotsarelis, Kurt Stenn and Jeffrey Miller. The magnificent textbooks of A. Bernard Ackerman have been an inspiration, and his love for teaching dermatopathology is infectious.

I still have much to learn about the pathology of hair disease. If I were to postpone publishing this work until I felt truly expert, it might never happen. The confidence to move forward is due, in part, to the encouragement I have received from my academic idols and mentors: William D. James, Stephen I. Katz, John Stanley, Kim Yancey, Jeffrey Callen, Kenneth Arndt, George Lupton and Val Hemming, among others.

And of course Joanne and Laura are always there for me. They make the effort worthwhile.
CHAPTER 1
Normal hair anatomy and architecture

The histological findings in many forms of hair loss are subtle, and an accurate diagnosis depends on distinguishing abnormal from normal follicular architecture. The word ‘architecture’ refers to the anatomy of individual hair follicles as well as the number, size and distribution of follicles within a biopsy specimen.

SIZE OF HAIRS: TERMINAL, INDETERMINATE, VELLUS

Long, thick hairs with bulbs in the fat are called terminal hairs. Vellus hairs are thin, short and often hypopigmented, with bulbs located in the upper portion of the dermis. Indeterminate hairs are intermediate in size between terminal and vellus hairs. Hair shaft diameters can be readily measured in transverse section; terminal hairs are thicker than 0.06 mm and vellus hairs are less than 0.03 mm. A vellus hair can be readily identified by simple inspection, since its inner root sheath will be as thick as or thicker than the shaft (Figures 1.1 and 1.2).

PHASE OF HAIR CYCLE: ANAGEN, CATAGEN, TELOGEN

Every follicle, regardless of size, can be found in one of three phases of the ‘hair cycle’. The anagen phase is the active growing period, and lasts weeks to years, depending on the size and site of the hair. For human terminal scalp hair, anagen lasts between 2 and 7 years. Catagen is a brief transitional phase between anagen and telogen, and lasts about 2–3 weeks. The telogen phase lasts about 100 days, at the end of which the shaft is shed. Depending on the individual, between 85 and 100% of terminal scalp hair is in the anagen phase at any given time; 0–15% is in the telogen phase and only about 1% is in the catagen phase. The percentage of terminal telogen hairs present (usually based on a lock of hairs forcibly plucked from the scalp) is called the ‘telogen count’. ‘Average’ telogen counts are in the range of 6–13%, and a count greater than 20% is abnormal.
Anagen hair anatomy

Anagen, catagen and telogen hairs differ considerably in their appearance. Anagen hairs can be described as having four zones. From deep to superficial, these zones are the hair bulb, the suprabulbar zone, the isthmus and the infundibulum.

The hair bulb, usually located in the fat, is composed of the hair matrix, the basophilic germinative layer, which surrounds the hair papilla. The hair papilla is a mesenchymal structure derived from the dermis. Inferiorly, the stalk of the papilla merges with the fibrous root sheath, which surrounds the entire follicle and blends into the overlying dermis (Figures 1.3 and 1.4).

Just above the hair bulb is the suprabulbar zone, at which point the various layers of the anagen follicle begin to differentiate. Transverse sectioning allows all the layers of the anagen follicle to be easily identified at this level. Starting
from the center of the follicle and moving outward, the layers are the hair shaft medulla, the hair shaft cortex, the cuticular layer, Huxley’s layer of the inner root sheath, Henle’s layer of the inner root sheath, the glassy (vitreous) layer and the fibrous root sheath. The cuticular layer is composed of the inter-locking flattened cells of the hair shaft cuticle and the inner root sheath cuticle. These cells are so tightly inter-locked that they appear to form a single anatomical layer (Figures 1.5 and 1.6).

Moving toward the skin surface, the next zone is the isthmus. The inferior landmark for the isthmus is the insertion of the arrector pili muscle into the fibrous root sheath of the follicle. The superior landmark for the isthmus is the entrance of the sebaceous duct into the follicular canal. The isthmus is an important transitional zone of follicular keratinization. In the mid-portion of the isthmus, the inner root sheath desquamates, resulting in a separation between the hair shaft and the follicular wall. At this point, the cells of the outer root sheath
begin to cornify without the formation of a granular cell layer. This is called trichilemmal keratinization (Figures 1.7 and 1.8).

The uppermost zone of the follicle is the infundibulum, bounded inferiorly by the entry of the sebaceous duct. Cornification of the outer root sheath occurs with the formation of a granular cell layer, similar to the epidermal surface (Figures 1.9, 1.10 and 1.11).

The epithelium adjacent to the insertion of the arrector pili muscle (the bulge) is the putative site of the follicular stem cells. Although an actual anatomical bulge is evident in rodent follicles, human follicles do not usually have an obvious protuberance of epithelium at this site. Special histochemical stains directed against antigens such as cytokeratin 15 are required to identify the stem cells (Figures 1.12 and 1.13).

Hair shaft size and shape differs between racial groups. The hair shafts of African Americans tend to be elliptical or kidney-bean-shaped (Figure 1.14), and are situated eccentrically within the epithelium of the follicle. Caucasian hair shafts tend to be circular or slightly oval, and are usually located directly in the center of the follicle.

**Catagen hair anatomy**

Each terminal anagen hair on the scalp grows for about 2–7 years, depending on the individual. At the end of that time, the hair shaft ceases active growth and enters a brief catagen phase. Within just a week or two, the entire anatomy of the follicle changes. At the beginning of catagen, the hair matrix disappears and is replaced by a thin rim of epithelial cells surrounding the hair papilla. These epithelial cells along with an overlying homogeneous column of epithelial cells demonstrate nuclear pyknosis. The epithelium of the lower follicle is undergoing disintegration by way of apoptosis (programmed cell death). As these epithelial changes occur, the vitreous (or glassy) layer markedly thickens, so that it becomes a prominent structure. The fibrous root sheath also thickens. As the catagen phase progresses over a 2–3-week period, the hair papilla follows the
disintegrating epithelial column upwards into the dermis, and the papilla eventually comes to rest just below the bulge zone (attachment of arrector pili muscle).

As the epithelial column moves upward, a collapsed fibrous root sheath is left behind. This collapsed structure is called the *stela* (or stele; plural, stelae), derived from the Greek word for ‘pillar’. The stela is also referred to as the follicular ‘streamer’. In this text, the terms ‘stela’ and ‘streamer’ are considered to be synonymous.

Just above the epithelial column of the catagen hair, an expanded mass of epithelium forms the presumptive club hair. Early in catagen the cells of the presumptive club are still nucleated, but the nuclei disappear as the club begins to cornify from the center outward (Figures 1.15–1.22).

*Figure 1.5* Terminal anagen hair, suprabulbar zone, vertical section. Just above the bulb, the various layers of the anagen hair can be identified: medulla (M) if present, cortex (C), cuticular layer (Cu), inner root sheath (IRS), outer root sheath (ORS), vitreous/glassy layer (V) and fibrous root sheath (F). Original magnification ×400
Telogen hair anatomy

At the end of catagen and the beginning of telogen, the hair papilla is a condensed ball of spindle-shaped nuclei within a scanty stroma. The papilla lies just below a nipple of epithelium called the secondary hair germ (Figure 1.23). When sectioned transversely, the secondary hair germ has an asterisk-like appearance (Figure 1.24). After the club hair has been shed from the follicle, the secondary hair germ is sometimes referred to as the telogen germ unit.

The secondary hair germ (or telogen germ unit) is adjacent to the bulge zone and the cornifying presumptive club. This places the hair papilla cells in close proximity to the stem cells residing in the bulge zone. Below the hair papilla lies the stela (collapsed fibrous root sheath; Figures 1.20–1.22).

Just above the secondary hair germ the telogen club is progressively cornifying and expanding in a centrifugal fashion. Several layers of non-cornified epithelium surround and tightly adhere to the cornifying club. As telogen progresses, the cornified club expands to occupy the full width of the follicle. This progressive cornification occurs over about a 3-month period, at the end of which the hair shaft with its clubbed root is shed from the follicle (Figures 1.25 and 1.26). Like anagen hairs, telogen hairs may be large (terminal) or small (vellus or miniaturized; Figure 1.27).

At the end of the telogen phase, shortly before or after the club hair is shed, the stem cells of the follicular bulge are activated and form a population of rapidly proliferating epithelium (‘transient amplifying cells’). These cells descend into the vacant and collapsed fibrous root sheath. Thus begins the formation of a new anagen hair (Figure 1.28). A hair matrix is re-formed and the various layers of the hair differentiate. The new anagen hair will continue to produce a hair shaft for the next 2–7 years.
PULLED, PLUCKED AND SPONTANEOUSLY SHED HAIRS

Hairs that are shed or extracted from the scalp can be examined directly under the microscope without any special processing. Covering the hairs with a few drops of immersion oil and a second glass slide facilitates examination. Normally, the only hairs that can be gently and easily pulled from the scalp are telogen hairs. Any other type of hair that is easily removed from the scalp can be regarded as abnormal. Examples of abnormal findings are the tapered, ‘pencil point’ hairs seen in chemotherapy, radiation therapy and alopecia areata (see Chapter 28). Fragile hair shafts will fracture when gently pulled, and the fracture can be identified at the proximal end of the shaft. In some highly inflammatory scalp diseases, entire anagen hairs including their root sheaths can sometimes be extracted. For example, this can occasionally be seen in lichen planopilaris.

Hairs can also be obtained from the scalp by forcibly plucking the shafts. In this case a mixture of anagen and telogen hairs will usually be found. The
process of plucking normal anagen hairs often leads to considerable artifactual distortion. Often anagen hairs are extracted with one or both of the root sheaths missing from the bulb. This sort of artifactual distortion should not be confused with a pathological abnormality. A forcible hair pluck is usually performed to determine the percentage of terminal telogen hairs (the trichogram).

**Anagen pull/pluck findings**

Normally, anagen hairs cannot be gently pulled from the scalp. They must be forcibly plucked. It is pathological for anagen hairs to be easily and painlessly extracted. Examples of this abnormality occur in loose anagen hair syndrome and lichen planopilaris.

A plucked anagen hair is characterized by a pigmented bulb, which is often expanded into a triangular, ‘delta’ shape as it tears free from the hair papilla. The hair papilla is usually (but not always) left behind in the dermis. Because the hair

![Image](image-url)
bulb is not cornified, it is soft and can take on a bent or ‘hockey stick’ configuration. Just above the bulb, portions of both an inner and an outer root sheath can be identified. Unfortunately, the outer root sheath or both root sheaths are often left behind in the dermis, an artifact of plucking. This gives the hair a ‘dysmorphic’ (not dystrophic) appearance (Figures 1.29–1.31).

**Catagen pull/pluck findings**

Like anagen hairs, catagen hairs are seldom gently pulled from the scalp. They are sometimes found when a lock of hair is forcibly plucked from the scalp. Plucked catagen hairs closely resemble telogen hairs, except that a clear, non-cornified sac surrounds the ‘club’, and a ‘tail’ of soft, clear tissue lies below the bulb. This ‘tail’ represents the degenerating epithelial column that lies between the hair papilla and cornifying ‘club’ (Figure 1.32).
Telogen pull/pluck findings

Telogen hair shafts are easily pulled or plucked from the normal scalp. It is estimated that the average person loses about 50 such hairs during the course of the day. Pulled or plucked telogen hairs have an expanded, depigmented or hypopigmented, club-shaped bulb. Because the entire telogen hair is cornified, the bulb is not ‘bent’ relative to the long axis of the shaft (Figures 1.33 and 1.34).

Trichogram findings

The forcible hair pluck, or trichogram, is a useful tool employed by some clinicians who evaluate hair loss. A small lock of approximately 50 hairs is grasped in a rubber-tipped surgical clamp, and the lock is quickly jerked out of the scalp. The shafts are taped down to a glass slide, with the bulbs placed in the
center, and the hemostat is released. A few drops of immersion oil are dripped onto the bulbs and a second glass slide or cover slip is placed on top. If the hair is kinky or curly, the slides may need to be taped together so that the hairs lie flat.

In theory, interpretation of the trichogram should be simple. The goal is to obtain a ‘telogen count’, the percentage of telogen hairs represented in the sample. However, considerable artifact may be introduced by the act of plucking, and the anatomy of anagen hair bulbs may be badly distorted, making some of them ‘dysmorphic’ and difficult to identify. Hairs with bulbs that have cleanly snapped off (as if cut) are counted as anagen hairs, as are bulbs that show remnants of an outer or inner root sheath. A pigmented and ‘bent’ bulb in the absence of root sheaths can also be counted as an anagen hair. In this case, the cuticle above the bulb will be ruffled (Figures 1.35–1.37).

**BIBLIOGRAPHY**


Figure 1.13 The bulge zone, as seen with immunohistochemical markers for cytokeratin 15 and smooth muscle actin. The dark red staining of the outer root sheath, found in the vicinity of the attachment of the arrector pili muscle, indicates the presence of cytokeratin 15. Cytokeratin 15 is a marker of the bulge zone. The brown staining of the arrector pili muscle indicates the presence of smooth muscle actin. Photomicrograph courtesy of George Cotsarelis, MD. Original magnification ×100
Figure 1.14 Transverse section of a terminal anagen hair from an African-American. The hair shaft is elliptical or kidney-bean-shaped, and is often eccentrically placed within the surrounding epithelium. Original magnification ×400

Figure 1.15 Terminal catagen hair, bulb, transverse section. A thin rim of pale-staining epithelial cells, many with pyknotic nuclei, surround the hair papilla (P). The vitreous layer (V) is markedly thickened. Original magnification ×400
Figure 1.16 Terminal catagen hair, suprabulbar, transverse section. Numerous pyknotic nuclei are present, and the markedly thickened vitreous layer (V) is clearly seen. Original magnification ×400
Figure 1.17 Terminal catagen hair, suprabulbar, transverse section, periodic acid-Schiff (PAS) stain. PAS staining highlights the thickened vitreous layer. The fibrous root sheath (F) is also much thicker than in an anagen hair. Original magnification ×400
Figure 1.18 Terminal catagen hairs, suprabulbar, transverse sections. The follicle on the right is sectioned just below the presumptive club hair where the epithelial column is widest. The other is sectioned through the presumptive club, which is just beginning to cornify centrally Original magnification ×200
Figure 1.19 Lower half of a terminal catagen hair, vertical section. The bulb (B; corresponding to Figure 1.15), disintegrating epithelial column (EC; corresponding to Figure 1.16) and presumptive club (PC; corresponding to Figure 1.18, left side) can be seen. Original magnification ×200
Figure 1.20 Vertical section of the stela (or ‘streamer’) below the bulb of a catagen hair. The stela is the collapsed fibrous root sheath found below hairs in the catagen/telogen phase or hairs that have miniaturized. Original magnification ×200
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Figure 1.24 Secondary hair germ (SHG) or ‘telogen germ unit,’ transverse section. Just below this structure lies the hair papilla. Just above lies the cornifying ‘club’ of the telogen hair (if it has not yet been shed). A terminal anagen hair (A) is nearby for comparison. Original magnification ×400

Figure 1.25 Transverse section of an early telogen hair bulb. The cornifying club’s serrated rim interdigitates with the surrounding envelope of outer root sheath epithelium. Original magnification ×400
Figure 1.26 Bulb of a late telogen follicle, transverse section. The cornified club now fills most of the width of the follicle, with only a thin rim of non-cornified epithelium remaining. The club hair is now ready to be shed. Original magnification ×400

Figure 1.27 Section through the bulbs of a terminal telogen hair and a vellus telogen hair. Original magnification ×200
Figure 1.28 Newly growing anagen hair, extending down from the bulge zone. The telogen club hair has not yet been shed. Original magnification ×100
Figure 1.29 Bulb of forcibly plucked, terminal anagen hair. Original magnification ×200
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Figure 1.31 ‘Dysmorphic’, plucked terminal anagen hair. Here the term ‘dystrophic’ should not be applied, because the loss of the root sheaths is not a growth defect but merely an artifact of plucking. Original magnification ×100
Figure 1.32 Forcibly plucked catagen hair. The tail-like remnant of the degenerating epithelial column is found just below the presumptive club. Original magnification ×200
Figure 1.33 Forcibly plucked early telogen hair. A noncornified epithelial sac surrounds the cornifying club. Original magnification ×200

Figure 1.34 Forcibly plucked or gently pulled late telogen hairs. The ‘clubs’ are completely cornified and the epithelial sac is gone. Original magnification ×200
Figure 1.35 Forcibly plucked lock of hairs—the trichogram. In this field of 23 terminal hairs, 21 anagen hairs and two telogen hairs are present. The telogen count is therefore 2/23, or about 9%. Original magnification ×40

Figure 1.36 ‘Snapped-off’ shaft, an artifact of plucking. A telogen hair (upper hair) and a ‘snapped-off’ shaft are found in this field. When deeply rooted, terminal anagen hairs are plucked, a clean transverse break in the shaft, as seen here, is sometimes the result. Shafts such as these can be counted as anagen hairs. Original magnification ×200
Figure 1.37 ‘Dysmorphic’, terminal anagen hair, an artifact of plucking. Sometimes when a terminal anagen hair is plucked, the entire shaft is removed but the root sheaths are left behind in the scalp. The pigmented, bent root (often resembling a hockey stick) and the ruffling of the cuticle of the proximal shaft identify such a hair as an anagen hair. A similar hair is seen in Figure 1.31. Original magnification ×200
CHAPTER 2
Specimen acquisition, handling and processing

ACQUISITION
Selecting the biopsy site is the most difficult and important part of the process. The most fruitful site will vary depending on the disease, and often the clinician is uncertain of the diagnosis. Sampling the center of a lesion of alopecia areata or trichotillomania would be appropriate. Sampling the bald center of a lesion of scarring alopecia is seldom useful, and the ‘active’ peripheral margin would be a more suitable target. Compounding the difficulty of site selection is sampling error. Clinicians cannot see below the surface of the skin, and even the most experienced physician may choose an unrewarding spot. If a recently involved area of scalp showing early clinical changes is selected, the diagnostic yield will be higher. Highly inflamed sites (pustules or papules) are often very advanced lesions and are frequently non-diagnostic. Multiple separate specimens chosen from several sites in or around the lesion will increase the diagnostic yield. However, clinicians may not have the luxury of obtaining multiple specimens.

In some cases the patient’s ‘normal’ scalp can serve as a basis for comparison with the abnormal scalp. For example, a specimen from the mid-occiput can help establish a diagnosis of common balding in a patient with hair loss on the crown.

Once the site is selected, it should be anesthetized with lidocaine with epinephrine (adrenaline). A generous amount of anesthetic (1–3 ml) should be injected into the deep dermis and superficial fat, and allowed to act for 15–30 minutes before the biopsy is performed. This will minimize bleeding.

The blade of the punch biopsy tool should extend through the dermis down into the fat, so that intact bulbs of deeply rooted terminal hairs can be removed.

A 4-mm biopsy wound can be easily closed with 3–0 suture because the needle can traverse the wound in a single pass. A suture color that contrasts with the patient’s hair will assist in suture removal 1 week after the biopsy is performed.
HANDLING

Once obtained, the scalp biopsy specimen should be allowed to fix in formalin for at least 24 hours before sectioning. Biopsy specimens obtained for direct immunofluorescence testing should of course be placed in the appropriate transport solution.

PROCESSING

The required tools include a sharp blade, a blade holder, a pair of fine-toothed forceps, marking ink, a cotton-tipped applicator and the standard plastic specimen cassette. Sponges should be used inside the cassette to prevent the thin slices of tissue from escaping. The sharpest blade for the job is a disposable, flexible shaving blade (‘blue blade’). These can easily be snapped in half for economy and safety (Figure 2.1). The author prefers to use red marking ink, but
any color will do. A specially colored cassette will alert the histology technician that embedding must be performed in a particular way.

There are several ways to slice the tissue into transverse sections. The technique used by Headington and Whiting involves a single transverse slice about 1 mm below the epidermal surface. Both cut sides of the specimen are embedded down in the cassette. As the microtome cuts deeper into the tissue block, the sections become progressively more superficial in one half of the specimen and deeper in the other. Simply sectioning deeper into the block allows one to obtain sections as superficial or as deep as required.

An alternative method is that of Frishberg and Sperling. One treats the biopsy specimen like a cylindrical loaf of bread, and cuts it into three or four slices (Figures 2.2 and 2.3). The ‘slices’ are about 1 mm thick. The deep surface of each slice is inked (Figure 2.4) and the inked sides are placed down in the cassette. Once the ink has been removed by the microtome, a single section is taken. In this way, the specimen is sampled at several different depths (Figure 2.5). Because only a single section is needed to view multiple levels on a
Yet another but more tedious method involves embedding the epidermal and/or fat end down in the cassette, and taking multiple horizontal sections through the entire specimen. Dozens of sections can be required to section through the entire block, and the block is exhausted in the process. However, every possible level of every follicle in the specimen can be carefully studied, and this technique is useful for research purposes.

**BIBLIOGRAPHY**


Figure 2.5 The final product is a single slide containing disks of tissue from multiple levels (superficial to deep)


CHAPTER 3
Evaluating and describing transverse (horizontal) sections

THE RATIONALE OF TRANSVERSE/HORIZONTAL SECTIONING

Most specimens obtained for the diagnosis of hair loss should be sectioned transversely (horizontally). The technique of transverse sectioning is unfamiliar to many pathologists, and so it will be useful to discuss its rationale. Vertical sections are of limited value in the study of hair disease because the number and type of hairs found on a vertical section are subject to considerable sampling error. The information obtained from vertical sections is usually incomplete and often misleading, as demonstrated in Figure 3.1. A vertical section bisecting the specimen through plane ‘X’ would sample only four follicles out of a total of 28 contained in the specimen. A section through plane ‘Y’ would not sample any follicles. In many cases of alopecia, the diagnosis hinges on just a few follicles. The chance of ‘hitting’ these follicles on a routine vertical section is slim. Accurately counting follicles is nearly impossible using vertical sections, so quantitative data cannot be gleaned in this way.

Transverse/horizontal sectioning ensures that all follicles in the specimen are counted and examined, at multiple levels, with just a few sections. Pathologists and dermatologists who have been disappointed with the results of vertically sectioned hair biopsy specimens are truly gratified when transverse sectioning is employed.

THE PATHOLOGIST’S CHECKLIST FOR EVALUATING SCALP BIOPSY SPECIMENS

The pathologist can employ a checklist to avoid missing important information. The order of the checklist is a matter of personal preference, but the pertinent items that must be addressed before arriving at a diagnosis include:

(1) What is the specimen diameter (e.g., 4 mm)? This information is required to determine whether the number of follicles is normal or decreased.
(2) Are follicular units evenly spaced, or are there ‘blank spots’ with an absence of follicles, suggesting scarring or severe miniaturization (Figures 3.2 and 3.3)?

(3) Do most follicular units contain two to six follicles, with large follicles outnumbering small follicles (Figure 3.4)?

(4) Are a normal number (> 85%) of the terminal follicles in the anagen phase (Figure 3.5)?

(5) Do any follicles show incomplete or distorted anatomical features (Figure 3.6)?

(6) Is inflammation present, and at what level of the follicle (bulb, suprabulbar, isthmus, infundibulum)?

(7) Is perifollicular fibrosis present; have individual follicles been entirely replaced by connective tissue (true follicular scars; Figure 3.7)?

(8) What is the total number of viable follicles in the specimen? The total number of terminal anagen hairs? The total number of vellus hairs?

The information gleaned from this evaluation will be used to compose the microscopic description. Interpreting the data and arriving at a final diagnosis is more challenging, and the major purpose of this text is to assist in this process. With few exceptions, the diagnosis of most forms of alopecia hinges on a constellation of findings and not on a single finding. The group of findings typical of each disease is described in the chapters devoted to particular entities.

**THE NORMAL SCALP IN TRANSVERSE SECTION**

The ‘architecture’ of a transversely sectioned biopsy specimen is quite predictable, and numerical data from the study of normal scalp specimens are available. Most data are based on 4-mm punch biopsy specimens, which can be considered the ‘standard’. A specimen 4 mm in diameter samples a surface area...
This formula can be used to compare data from punch biopsies larger or smaller in diameter with those of a 4-mm punch. For example, a 3-mm punch biopsy specimen has a surface area of 7.07 mm$^2$.

In a 4-mm punch biopsy specimen from a Caucasian, there should be about 33 terminal hairs in a transverse section through the deep dermis (Figure 3.8). Sectioning through the upper dermis will reveal about five additional vellus hairs (Figure 3.9). The ‘average’ scalp biopsy specimen will therefore contain about 38 follicles. Considerable individual variation exists and the range of normal is quite broad; total hair counts as low as 19 and as high as 59 have been recorded. On average, two of the terminal hairs will be in the telogen phase, and 31 will be in the anagen phase. The *telogen count* (number of terminal telogen hairs divided by the total number of terminal hairs) will therefore be about $\frac{2}{33}=6\%$. Considerable individual variation exists for telogen counts among normal individuals, and figures within a range of 0–15% (based on histological sections) can be considered normal.
On the average, black patients of African descent have fewer, although larger, follicles than white patients (Figures 3.10 and 3.11). The average number of follicles for African Americans, based on 4-mm biopsy specimens, is 21 (18 terminal and three vellus follicles) with an average telogen count of 7%. The percentage of catagen and/or telogen hairs can prove to be an important diagnostic feature in many forms of hair loss. Catagen and telogen hairs have the same diagnostic significance, since all catagen hairs become telogen hairs within a few weeks. For the sake of simplicity, these two phases can be grouped together as the *catagen/telogen phase*. In this textbook, the term catagen/telogen phase will be used to refer to hairs in either the catagen or the telogen phase. For diagnostic purposes, it is not important to differentiate between catagen and telogen hairs, but just to recognize a follicle as being in either phase. Catagen

**Figure 3.4** Transverse section through the upper dermis of a specimen from a normal scalp. Two follicular units are shown, the one on the left containing one terminal and two vellus hairs, and the one on the right containing three terminal hairs. Although the first unit may not appear ‘normal’ because vellus outnumber terminal hairs, *all* the follicular units need to be assessed to make a conclusion about overall normality. Original magnification ×100

**Figure 3.5** In this field, only two of five terminal follicles (40%) are in the anagen phase. This would be clearly abnormal if it were representative of the entire specimen. Original magnification ×100
hairs can be included with the telogen hairs when determining the telogen count (Figure 3.12).

In the superficial fat and lower dermis, home to the bulbs and suprabulbar zones of terminal anagen hairs, the follicles are spaced apart fairly evenly. However, in the mid- and upper dermis, groups of follicles become segregated into follicular units. Each follicular unit contains about two to five terminal hairs and zero to two vellus hairs (Figure 3.13). A shortcut for assessing numerical ‘normality’ is by looking at individual follicular units (Figure 3.14). If each unit contains two to five terminal hairs (outnumbering vellus hairs), and most units are similar, then the number and size of hairs are probably normal. As would be expected, the number of terminal hairs per follicular unit is smaller in African Americans than in Caucasians.

The average density of follicular units is about 1/mm² surface area. Therefore, a horizontal section 4-mm in diameter (12.6 mm²) will contain about 12 follicular units.

**Figure 3.6** Distorted follicular anatomy, in this case due to forcible plucking of the hair shaft prior to taking the biopsy specimen. The shaft and a portion of the inner root sheath are missing, and the epithelium has collapsed inward. Original magnification ×400

**Figure 3.7** True follicular ‘scars’, diagnostic of a scarring alopecia. In transverse section, they appear as discrete, circular or oval structures composed of compact and concentric collagen bundles. Original magnification ×400
In the normal scalp, the terminal: vellus hair ratio should be 2:1 or greater. If biopsy specimens from the crown and occiput of the same person are compared, they should be similar in terms of follicular size, number and percentage of anagen/telogen hairs. The ‘architecture’ of scalp biopsy specimens should be uniform over

Table 3.1 Template for a ‘hair biopsy report’

<table>
<thead>
<tr>
<th>Accession number:</th>
</tr>
</thead>
<tbody>
<tr>
<td>Date:</td>
</tr>
<tr>
<td>Patient name/age/sex/race:</td>
</tr>
<tr>
<td>Submitting physician:</td>
</tr>
<tr>
<td>Clinical impression:</td>
</tr>
<tr>
<td>Macroscopic description: biopsy diameter/location on scalp (e.g. 4-mm punch biopsy, vertex of scalp):</td>
</tr>
<tr>
<td>Microscopic description of vertical sections:</td>
</tr>
<tr>
<td>Microscopic description of horizontal sections:</td>
</tr>
</tbody>
</table>

Figure 3.8 Transverse section from normal scalp skin (Caucasian) at the level of the superficial fat. Only terminal follicles can be found at this level, and they are not yet organized into follicular units. Original magnification ×40

Figure 3.9 Transverse section from normal scalp skin (Caucasian) at the level of the upper dermis. The bulbs of telogen hairs and vellus hairs can also be found at this level, organized into follicular units. Original magnification ×40

In the normal scalp, the terminal: vellus hair ratio should be 2:1 or greater. If biopsy specimens from the crown and occiput of the same person are compared, they should be similar in terms of follicular size, number and percentage of anagen/telogen hairs. The ‘architecture’ of scalp biopsy specimens should be uniform over
Figures 3.10 Transverse section from normal scalp skin (African American) at the level of the deep dermis. Normally, only terminal and indeterminate follicles can be found at this level, and they are not yet organized into follicular units. This specimen contained 16 total follicles. Original magnification ×40

Figure 3.11 Another transverse section from normal scalp skin of an African American, containing 22 follicles. These numbers would be regarded as low for a Caucasian but are normal in African Americans. Original magnification ×40

Terminal anagen hairs:
Terminal catagen hairs:
Terminal telogen hairs:
Telogen germinal units:
Vellus hairs:
Total hairs (terminal plus vellus, all phases):
Anagen : telogen percentages (e.g. 85% : 15%):
Terminal: vellus ratio (e.g. 3:1):
Follicular units:
Follicular stelae:
Lymphohistiocytic infiltrate:
Upper follicle:
Lower follicle/bulb:
Fibrosis:
Comments:
DIAGNOSIS:
Consultants:
Pathologist’s signature_________________

the entire scalp, with the possible exception of the fringes.
Mild *upper* dermal perifollicular inflammation may be present even in normal scalp specimens. This is especially true for African American patients.

**FIGURE 3.12** Transverse section through the level of the dermal-subcutaneous junction. In this particular field, there are 12 terminal anagen hairs and seven terminal telogen hairs, for a total of 19 terminal hairs. The telogen count for this field would therefore be 7/19=37%. Original magnification ×40

**FIGURE 3.13** Normal follicular unit. This unit contains four terminal hairs and one vellus hair. Original magnification ×100

**REPORTING HISTOLOGICAL FINDINGS - THE BIOPSY REPORT**

Transverse sectioning allows the pathologist to gather a considerable amount of data about the specimen. This can be summarized in a microscopic description (e.g. ‘A reduced number of hairs are present, and most are miniaturized’).
Alternatively, detailed data can be presented in tabular form. A template for a ‘complete’ report, adapted from that used by David A. Whiting, MD, is presented in Table 3.1. Such a detailed report not only serves as the basis for the final diagnosis, but also allows for efficient data collection for research purposes.

**BIBLIOGRAPHY**


CHAPTER 4
Classification of hair disease

Various classification schemes for alopecia exist, but all are imperfect. Most forms of alopecia demonstrate at least some overlapping clinical and histological features. This overlap blurs the distinction between diseases, making classification difficult. It would make the most sense to segregate diseases by etiology, but the causes of many forms of hair loss are unknown, making it difficult to group diseases with confidence.

CICATRICIAL (SCARRING) VS. NON-CICATRICIAL ALOPECIA

The most common classification system divides the diseases into cicatricial (scarring) and non-cicatricial (non-scarring) forms of alopecia. In this textbook, the terms ‘cicatricial’ and ‘scarring’ will be considered to be synonymous and will be used interchangeably. Cicatricial or scarring implies that follicular epithelium has been replaced by connective tissue. However, in some cases of alopecia, follicles seem simply to ‘disappear’ without noticeable alteration in tissue architecture. The broadest definition of scarring alopecia might include all forms of alopecia in which hair follicles are permanently lost. In contrast, non-scarring alopecia is potentially reversible.

However, certain hair diseases demonstrate a biphasic pattern, where non-scarring hair loss is seen early in the course of the disease, and permanent hair loss becomes apparent in the later stages of the disease (Figure 4.1). Examples of diseases demonstrating this biphasic pattern include androgenetic alopecia, alopecia areata and traction alopecia. These forms of alopecia are generally considered to be non-scarring. However, after many years or decades of continuous active disease, permanent dropout of follicles occurs.

CLASSIFICATION OF CICATRICIAL (SCARRING) ALOPECIA

This classification is especially confusing and controversial. There are no characteristic biological markers for most forms of scarring alopecia. We do not know whether the clinical and histological features found in a given patient are
manifestations of a distinct disease, or just individual host responses. Any classification of scarring alopecia should be considered provisional and subject to change as new information becomes available.

Scarring alopecia can be subdivided into two categories. The first is primary scarring alopecia, where the target of inflammation appears to be the follicle. In secondary scarring alopecia, the follicle is merely an ‘innocent bystander’ in the disease process, and is destroyed in a non-specific manner. Examples of secondary scarring alopecia include deep burns, radiation dermatitis, cutaneous

Figure 4.1 The biphasic form of alopecia. With the passage of time, follicles begin to disappear permanently and histological specimens take on the appearance of a cicatricial alopecia.
malignancies, cutaneous sarcoid, sclerosing dermatoses such as morphea and necrobiosis lipoidica, and certain chronic infections such as cutaneous tuberculosis. The various forms of secondary scarring alopecia should have distinctive histological features typical of the underlying disease. For example, alopecia caused by cutaneous sarcoidosis should reveal sarcoidal granulomata in the dermis. A discussion of the various forms of secondary scarring alopecia is beyond the scope of this text.

Primary scarring alopecia can be divided into six diagnostic groups (Figure 4.2). They are:

1. Central, centrifugal scarring alopecia;
2. Lichen planopilaris;
3. Chronic, cutaneous lupus erythematosus (discoid lupus erythematosus);
4. Acne keloidalis (folliculitis keloidalis; acne keloidalis nuchae);
5. Dissecting cellulitis (perifolliculitis abscedens et suffodiens);
6. Scarring alopecia, not otherwise classified.

This short list is a simplification of a longer list of confusing, vague and poorly defined diagnostic terms that have been coined and used by various authors during the past century. Notably absent from the above list are terms such as pseudopelade, pseudopelade of Brocq, folliculitis decalvans, tufted folliculitis and a variety of other more obscure terms. These entities are poorly defined, and
their nomenclature is used in various ways by different authors and clinicians. Almost all of these older terms can be incorporated into one of the six categories shown in Figure 4.2. Nevertheless, many of the older terms will be discussed in this text for the sake of clarity.

**BIBLIOGRAPHY**


Classical schemes are of little value to pathologists trying to extract the diagnosis from a glass slide. The histological features, in conjunction with a brief clinical description, must somehow guide the pathologist to the correct diagnosis. To assist in this process, some especially important histological features (listed below) will help to segregate the diagnostic entities. Identifying additional histological features will make it easier to establish the diagnosis. Frequently, different diseases may share two or more histological features. When this occurs, separating such diseases may rest on good clinical correlation or subtle histological clues.

Attempts have been made to create ‘alopecia algorithms’ for arriving at a diagnosis, but in the author’s experience, such algorithms provide little assistance. The starting point for arriving at a histological diagnosis is recognition of an obvious or dominant histological finding, such as miniaturization of hairs, ‘missing’ hairs, or perifollicular inflammation. Unfortunately, no single histological feature is sufficient to establish a definitive diagnosis in any form of hair loss. However, a differential diagnosis based on one or two histological features will help to create a ‘short list’ of possible diagnoses. Additional histological features characteristic of the entities on the ‘short’ list can then be used to narrow the field of possibilities. Clinical information will often help to decide the issue. Listed below are histological features, and those diseases that often demonstrate the feature.

**DECREASED HAIR DENSITY**

This is determined by counting the total viable follicles/mm² (Figures 5.1 and 5.2).

- Cicatricial alopecia (all forms)
- Congenital hypotrichosis (various syndromes)
- ‘End stage’ traction alopecia (an example of the biphasic pattern of hair loss)
- Androgenetic alopecia or hereditary balding (very long-standing disease; an example of the biphasic pattern of hair loss)
• Alopecia areata (very long-standing disease; an example of the biphasic pattern of hair loss)

DECREASED HAIR DENSITY AT THE LEVEL OF THE LOWER DERMIS

This is determined by counting only at this level.

• Telogen effluvium
• Androgenetic alopecia
• Temporal triangular alopecia
• Traction alopecia
• All forms of cicatricial alopecia
• Alopecia areata (long-standing disease)

Note: hair density in dark-skinned, kinky-haired individuals (e.g., many African Americans) is lower than in Caucasians.

NORMAL HAIR DENSITY DESPITE A CLINICAL REDUCTION IN THE AMOUNT OF HAIR

Here, there is a discrepancy between the clinical impression of marked hair loss and the histological finding (total viable follicles/mm²) of numerous hairs (Figures 5.3 and 5.4).

- Telogen effluvium (catagen/telogen increased)
Figure 5.4 As in the patient in Figure 5.3, this specimen was taken from a ‘bald’ spot of alopecia areata, but normal or nearly normal numbers of follicles were present. In this case it was because of a massive conversion to telogen hairs. Original magnification ×100

Figure 5.5 Miniaturized hairs in a patient with temporal triangular alopecia. Average hair shaft diameter is 0.02 mm. Original magnification ×200

Figure 5.6 Normal hairs from the same patient as in Figure 5.5 (normal, perilesional skin). Average shaft diameter is 0.12 mm. Original magnification ×200

- Trichotillomania (catagen/telogen increased)
- Androgenetic alopecia/hereditary balding (miniaturized hair increased)
• Alopecia areata (miniaturized/telogen hairs increased)
• Temporal triangular alopecia (miniaturized hair increased)
• Loose anagen hair syndrome (catagen/telogen increased)
• Most hair shaft disorders

MINIATURIZATION OF HAIRS

See Figures 5.5 and 5.6.

• Androgenetic alopecia/hereditary balding
• Temporal triangular alopecia
• Alopecia areata
• Patchy hair loss in systemic lupus erythematosus
• Patchy or diffuse hair loss in secondary syphilis
INCREASED PERCENTAGE OF CATAGEN AND/OR TELOGEN HAIRS

See Figures 5.7 and 5.8. Note: as mentioned earlier, the diagnostic significance of catagen and telogen hairs is the same, since all catagen hairs become telogen hairs within a few weeks. Therefore, follicles in either of these two phases can be referred to as catagen/telogen hairs.

- Trichotillomania
- Traction alopecia (acute)
- Postoperative (pressure-induced) alopecia
- Telogen effluvium
- Alopecia areata
- Androgenetic alopecia/hereditary balding (affected area only)

**Figure 5.9** In this patient with alopecia areata, the lower half of a follicle is affected by lymphocytic inflammation. Original magnification ×400

**Figure 5.10** The upper half of the follicle in Figure 5.9. This half is spared lymphocytic inflammation. Original magnification ×400

**INCREASED PERCENTAGE OF CATAGEN AND/OR TELOGEN HAIRS**
• Patchy hair loss in systemic lupus erythematosus
• Patchy hair loss in secondary syphilis
• Inflammatory, cicatricial alopecia

INFLAMMATION PREDOMINANTLY INVOLVING THE LOWER HALF OF THE FOLLICLES

See Figures 5.9 and 5.10.

• Alopecia areata (predominantly lymphocytic)
• Patchy hair loss in systemic lupus erythematosus (predominantly lymphocytic)
• Patchy hair loss in secondary syphilis (predominantly lymphocytic with occasional plasma cells)
• Dissecting cellulitis/perifolliculitis capitis abscedens et suffodiens (dense, mixed acute and chronic inflammation)

INFLAMMATION PREDOMINANTLY INVOLVING THE UPPER HALF OF THE FOLLICLES

With vacuolar interface alteration

See Figure 5.11.

• Lichen planopilaris
• Frontal, fibrosing alopecia
• Chronic, cutaneous (discoid) lupus erythematosus
Without vacuolar interface alteration

See Figures 5.12 and 5.13.

- Acne keloidalis
- Central, centrifugal scarring alopecia (also known as ‘follicular degeneration syndrome’ and ‘pseudopelade’)

**FOLLICULAR DROPOUT (REPLACEMENT OF FOLLICLES BY CONNECTIVE TISSUE)**

With loss of associated sebaceous glands

See Figures 5.14 and 5.15.

- Cicatricial alopecia: all forms
With associated sebaceous glands intact

Figure 5.17 In this specimen from a patient with central, centrifugal scarring alopecia there is a follicle whose inner root sheath has desquamated well below the isthmus (follicle at lower left). Normal follicles with intact inner root sheaths are present for comparison. Original magnification x100

Figure 5.18 An enlargement of the abnormal hair found in Figure 5.17. Original magnification x400

See Figure 5.16.

- Traction alopecia (end stage)
- Androgenetic alopecia (very advanced and long-standing)
- Alopecia areata (very advanced and long-standing)

PREMATURE DESQUAMATION OF THE INNER ROOT SHEATH

See Figures 5.17 and 5.18.
• Central, centrifugal scarring alopecia (also known as ‘follicular degeneration syndrome’ and ‘pseudopelade’)
• Acne keloidalis (occasionally)

**TRICHOMALACIA**

See **Figure 5.19**.

• Trichotillomania
• Acute traction alopecia
• Pressure-induced alopecia
• Alopecia areata
Clinicians who contribute even a brief clinical description and differential diagnosis are of considerable assistance to the pathologist. Some clinical features that may be described include:

- Pattern of hair loss
- Evidence of inflammation
- Evidence of hair breakage (hairs of uneven length)
- Evidence of permanent hair loss
- Obliteration of follicular ostia
- Abnormal hair density

The most important feature is the pattern of hair loss. Hair loss can be diffuse or 'patterned'. Patterned hair loss implies that the area of alopecia is confined to one or several portions of the scalp, leaving at least a portion of the scalp uninvolved. Truly diffuse hair loss suggests a uniform reduction in hair density over all portions of the scalp. Telogen effluvium (Figure 6.1) is an example of truly diffuse alopecia, with all areas of the scalp showing some thinning of the hair. Patchy alopecia areata is one example of patterned alopecia. The various patterns of thier loss are listed below.

**PATTERNS OF HAIR LOSS**

- Truly diffuse hair loss—e.g., telogen effluvium, alopecia totalis in evolution, chemotherapy/radiation therapy
- Diffuse hair loss over crown/vertex with relative sparing of parietal/occipital areas—e.g., androgenetic alopecia
- Nearly total hair loss with loss of follicular ostia, centered on crown or vertex—e.g., central centrifugal scarring alopecia
- Patches of hair loss with ‘bizarre’ or geometric shapes—e.g., trichotillomania
- Randomly scattered oval or circular patches of complete hair loss—e.g., alopecia areata
Androgenetic alopecia has been described as being both ‘diffuse’ and ‘patterned’. Although on rare occasions the balding process seems to affect the entire scalp, including the occiput, in the vast majority of cases the crown of the scalp (including frontal and vertex regions) is predominantly involved. Therefore, the condition has a distinctive ‘pattern’ of thinning (Figures 6.2 and 6.3).

Hair shaft disorders associated with hair fragility, such as trichorrhexis nodosa, tend to cause localized zones of alopecia containing hairs of uneven length. When the hair is particularly fragile, as is true in monilethrix, patches of hair stubble will be found. In disorders caused by hair shaft fragility, the shaft defect is usually readily apparent under the light microscope. In trichotillomania, the patches of alopecia are seldom totally ‘bald’. Usually there are several short hairs of varying lengths scattered over the involved area.

Clinicians should examine the scalp surface in all cases of hair loss. Abnormalities may or may not be related to the hair loss but need to be documented. Erythema, pustules, follicular papules and perifollicular scaling or hyperpigmentation suggest an inflammatory form of alopecia. Wide spacing between follicles, clusters of shafts exiting single follicular ostia (polytrichia or ‘tufting’) and the obliteration of follicular openings are all signs of scarring alopecia. Follicular ‘plugging’, with dilated ostia devoid of hair shafts, can be another indication of follicular destruction, as seen in discoid lupus erythematosus. Marked erythema and scaling or numerous papular lesions without a loss of follicular density suggests a primary scalp disorder (as opposed to a hair disorder). Psoriasis, seborrheic dermatitis and Langerhans cell

Figure 6.1 An example of truly diffuse hair loss (telogen effluvium). Hair thinning is uniform over the crown, occiput and sides of the head

- Randomly scattered, irregularly shaped patches of partial or complete hair loss —e.g., lichen planopilaris
- Marked thinning of both temples—e.g., burnt out traction alopecia
- Small, geometric patches of hair loss of anterior margin of temple (unilateral or bilateral) —e.g., temporal triangular alopecia
histiocytosis (histiocytosis X) of the scalp can cause dramatic scalp disease with little effect on hair density.

**Figure 6.2** An example of patterned hair loss in a woman with androgenetic alopecia. The crown is diffusely and symmetrically affected with relative sparing of the occiput as seen in Figure 6.3

BIBLIOGRAPHY

Figure 6.3 Occipital view of the patient in Figure 6.2
'Pure’ senescent alopecia is found in patients who boast a full head of hair well into middle age, and who typically deny a family history of balding. Patients with senescent alopecia note a very slow but steady, diffuse thinning of scalp hair starting at age 50 and older. Women who complain of a marked degree of thinning in the few years following menopause probably have a component of androgenetic alopecia. Many (and perhaps most) patients with senescent alopecia also have mild concomitant androgenetic alopecia, and the superimposed clinical and histological features of common balding and senescent alopecia may be impossible to separate.

Several authors have assessed the effect of aging on hair density by studying the scalp surface (i.e., clinical rather than histological evaluation). The results of these studies indicate that the density of hair follicles decreases steadily with aging. There is little information available on the histological evaluation of scalp biopsy specimens taken from ‘normal’ individuals of various ages.

**HISTOLOGICAL FINDINGS**

The ‘global impression’ when studying transverse sections is that there is only a slight numerical reduction in follicles that are otherwise normal. Compared to normal (or youthful) scalp, more follicles are in the telogen phase, but the telogen count may still be within the range of normal. Inflammation is uncommon, and fibrous streamers such as those found in androgenetic alopecia are absent. The follicles are not as long or as wide as normal, but ‘miniaturization’ such as that seen in androgenetic alopecia is absent. All these subtle findings would be uniform over the scalp surface.

The following combination of histological findings (based on 4-mm punch biopsy specimens) is typical of senescent alopecia (Figures 7.1 and 7.2):

1. A slight decrease in the total number of hairs (20–35 as opposed to the normal 30–45)
2. Numbers of telogen hairs within the range of normal (less than 15%)
3. A normal percentage of terminal hairs, with terminal hairs outnumbering vellus and indeterminate hairs by at least 2:1
Given these findings, androgenetic alopecia can be excluded because of the relative preponderance of terminal hairs, and the normal number of telogen follicles excludes telogen effluvium. If crown/vertex and midoccipital biopsy specimens from patients with senescent alopecia are compared, histological findings will be similar at both sites. This contrasts with androgenetic alopecia, where histological changes in the occiput (if present) are less dramatic than at the vertex or crown.
SUMMARY

Clinical correlation: an elderly person who admits to very gradual thinning of the hair; hair density appears normal (for age) or diffusely thinned over the entire scalp.

Histological findings:

- The specimen may appear entirely normal
- Findings from the crown/vertex resemble those found on the occipital scalp
- In advanced disease, the total number of hairs is reduced
- Terminal: vellus hair ratio and telogen count remain normal

BIBLIOGRAPHY

 CHAPTER 8
Androgenetic alopecia

Androgenetic alopecia is also known as common balding, hereditary balding, male-pattern balding and female-pattern balding. Some authors speculate that the pathogenesis of balding differs between men and women. However, the histopathological findings are similar in both sexes. Many women with a genetic predisposition to baldness first notice the onset or an acceleration of shedding and thinning around the time of menopause. However, some less fortunate women, especially those with strong family histories for balding, begin to thin in early adulthood. In the majority of cases, thinning is localized to the crown of the scalp, with sparing of the occipital and lower parietal fringe of hair (Figures 8.1 and 8.2). Two patterns of hair loss can be seen: Hamilton’s ‘male pattern’ and Ludwig’s ‘female pattern’. In fact, there is considerable overlap between the sexes, with many women demonstrating a ‘male’ pattern of hair loss, and some men showing a ‘female’ pattern, with diffuse crown thinning and retention of the frontal hairline.

In exceptional cases, the pattern of hair loss in both men and women with androgenetic alopecia can be truly diffuse (authors personal observation), with thinning of the occiput as well as the crown. In these cases, histological confirmation may be required to establish the diagnosis with certainty.

HISTOLOGICAL FINDINGS

Once considered a histological diagnosis of exclusion, androgenetic alopecia has distinctive and diagnosable features. The diagnostic ‘breakthrough’ has been the study of transverse sections of scalp biopsies, a technique popularized through the efforts of Headington and others. The microscopic diagnosis of androgenetic alopecia depends more on quantitative than qualitative features. Therefore, the ability to identify all follicles within a biopsy specimen is required to establish the diagnosis. Ideally, 4-mm punch biopsy specimens should be taken from both the involved and the relatively uninvolved sites (usually crown/vertex and occiput, respectively). All specimens should be sectioned horizontally. ‘Uninvolved’ skin can serve as the patient’s ‘normal control’. If the condition is advanced, a single specimen from balding skin may be sufficient.
Androgenetic alopecia is characterized by progressive miniaturization of hair follicles. This results in a mixture of hairs with various bulb depths and shaft diameters. When one examines a specimen from a normal scalp, sectioned transversely through the lower dermis, one gets the ‘global impression’ that all hairs are terminal hairs of similar size (diameter). A biopsy specimen from a patient with androgenetic alopecia, sectioned at the same level, will show hairs that appear vastly different in diameter (Figures 8.3–8.6). Below each miniaturized follicle is a ‘streamer’, the collapsed connective tissue sheath that once surrounded the formerly deep-seated, terminal hair. Streamers can also be found beneath follicles that have become temporarily miniaturized, as in alopecia areata. The streamers found in ‘early’ androgenetic alopecia can be difficult (if not impossible) to distinguish from the collapsed fibrous root sheath (‘follicular stela’) found below a normal terminal follicle that has temporarily entered the catagen/telogen phases. After a streamer has been present for years (as in ‘late’ androgenetic alopecia), the streamer becomes less vascular and assumes a grayish hue when stained with hematoxylin and eosin (Figures 8.7–8.9).
normal ratio of terminal: vellus hairs should be at least 2:1, but specimens
diagnostic of androgenetic alopecia show a reduction in this ratio. In advanced
cases, the number of vellus and indeterminate hairs will actually surpass
the number of terminal hairs (Figures 8.10 and 8.11). Early in the course of balding,
the total number of follicles present remains normal. Also, sebaceous glands
persist even when the hairs have greatly miniaturized (Figure 8.12). However, in
very long-standing balding, there is an actual decrease in follicular density as
well as follicular size. Therefore, androgenetic alopecia shows a biphasic pattern
of hair loss and may eventually resemble cicatricial alopecia.

As hairs miniaturize, the anagen phase becomes shorter. Therefore, the balding
scalp shows an increase in the telogen count, since miniaturized hairs spend a
greater proportion of each hair cycle in the telogen phase. In well-established
androgenetic alopecia, the majority of telogen follicles will be miniaturized
follicles (Figure 8.13). Successful treatment with minoxidil or finasteride will
increase the size of follicles and reduce the telogen count, presumably by
lengthening the duration of anagen.

Figure 8.2 The same patient as in Figure 8.1. There is relative sparing of the occiput

Inflammation has been described as a histological feature of androgenetic alopecia, but balding should be regarded as a form of non-inflammatory hair loss. Many biopsy specimens of androgenetic alopecia show mild, perifollicular, lymphohistiocytic, upper dermal inflammation, sometimes associated with mild perifollicular fibrosis. These changes are subtle and non-specific, and can also be found in many ‘normal’ scalp specimens. Mild, peri-infundibular chronic inflammation is especially common in African American women and can be regarded as normal. Peribulbar or destructive inflammation is absent in common balding.

Figure 8.3 Section (involved vertex) taken at the level of the dermal/fat junction. In this woman with early androgenetic alopecia, the number of follicles in the vertex specimen is similar to the number in the occipital specimen (Figure 8.4). However, follicles in the vertex specimen show considerable variation in diameter, and many are quite small. Original magnification ×40

Inflammation has been described as a histological feature of androgenetic alopecia, but balding should be regarded as a form of non-inflammatory hair loss. Many biopsy specimens of androgenetic alopecia show mild, perifollicular, lymphohistiocytic, upper dermal inflammation, sometimes associated with mild perifollicular fibrosis. These changes are subtle and non-specific, and can also be found in many ‘normal’ scalp specimens. Mild, peri-infundibular chronic inflammation is especially common in African American women and can be regarded as normal. Peribulbar or destructive inflammation is absent in common balding.
SUMMARY

Clinical correlation: symmetric thinning, predominantly affecting crown, vertex and frontal regions, with relative sparing of the occiput; no evidence of scarring. Family history of balding is usually elicited.

Histological findings (Figures 8.14 and 8.15):

- Normal total number of follicles (about 35 in Caucasians or 20 in African Americans per 4-mm plug)
- Reduced number of hairs (mixture of terminal and indeterminate) when counted at the dermal/fat junction
- Increased numbers and percentage of vellus and indeterminate hairs when counted at the level of the upper dermis
- Presence of fibrous ‘streamers’ below miniaturized hairs
- Slightly increased telogen count compared with ‘unaffected’ scalp
- Uninvolved scalp (e.g., occiput) appears normal or relatively normal

Figure 8.4 The same patient as in Figure 8.3, with this section taken from the normal occiput. Original magnification ×40
Figure 8.5 Involved vertex. The same specimen as in Figure 8.3, but here sectioned at the level of the mid- to upper dermis. The marked variation in hair size in the vertex specimen is evident. However, the total number of hairs in both vertex and occipital (Figure 8.6) specimens appears normal. Original magnification ×40

- No significant inflammation

BIBLIOGRAPHY


Whiting DA, Waldstreicher J, Sanchez M, Kaufman KD. Measuring reversal of hair miniaturization in androgenetic alopecia by follicular counts in horizontal sections of
Figure 8.6 Normal occiput. The same specimen as in Figure 8.4, but here sectioned at the level of the mid- to upper dermis. Original magnification ×40


Figure 8.7 Follicular stelae (‘streamers’) in a 50-year-old woman with androgenetic alopecia. ‘Old’ streamers, such as those seen here and in Figure 8.8, have a gray-blue hue and few vascular spaces. Original magnification ×400

Figure 8.8 ‘Old’ streamer, in the same woman as in Figure 8.7. Original magnification ×400
Figure 8.9 Relatively ‘new’ stela, in a 16-year-old girl with androgenetic alopecia. Such ‘streamers’, found in more recently miniaturized hairs, show more prominent vascularity. Original magnification ×400

Figure 8.10 Involved vertex. The same specimen as in Figure 8.3, sectioned at the level of the mid- to upper dermis. When multiple follicular units from the balding scalp are examined, most will show that vellus hairs equal or even outnumber terminal hairs. Original magnification ×200
Figure 8.11 Normal occiput. The same specimen as in Figure 8.5, sectioned at the level of the mid- to upper dermis. Here, when multiple follicular units from the normal scalp are examined, terminal hairs outnumber vellus hairs. Original magnification ×200

Figure 8.12 A follicular unit in which all hair shafts have miniaturized. Even so, the sebaceous glands remain intact. Original magnification ×200
Figure 8.13 Involved crown of a woman with androgenetic alopecia. The percentage of telogen hairs is increased, but the majority of telogen hairs are vellus or indeterminate hairs. Original magnification ×200
Figure 8.14 A 16-year-old girl with marked androgenetic alopecia. All the typical histological features are present, including: normal number of follicles; variation in follicular size; vellus and indeterminate follicles outnumbering terminal follicles; increased percentage of catagen/telogen follicles; and no significant inflammation. Original magnification ×40
Figure 8.15 The same specimen as in Figure 8.14. Original magnification ×200
A telogen effluvium occurs when abnormally large numbers of anagen hairs from all areas of the scalp enter the telogen phase. This may be caused by some sort of endogenous stress to the follicles, such as a metabolic disturbance, nutritional deficiency, or serious systemic illness. Other cases of telogen effluvium are ‘physiological,’ and not indicative of disease. The many possible causes are listed in Table 9.1. In response to the causative factor, many hairs prematurely enter the catagen phase. This is a committed step for follicles; having entered catagen they must proceed through the telogen phase and shedding before a new anagen hair can regrow.

Telogen effluvium is probably the most common form of hair loss associated with systemic diseases, especially chronic and debilitating conditions. However, only the most dramatic cases of telogen effluvium, resulting in more than 25% hair loss, are likely to come to clinical attention. Most drugs that have been associated with hair loss, with the exception of anticancer medicines and other toxins, cause hair loss by way of a telogen effluvium.

Physiological forms of telogen effluvium include postpartum and neonatal hair loss. In postpartum telogen effluvium, numerous hair follicles are artificially maintained in the anagen state under the influence of gestational hormones. This is reflected in the lower telogen counts that occur towards the end of pregnancy. After parturition, numerous follicles suddenly enter the catagen/telogen phase, and shedding of hairs begins about 3 months later.

Telogen effluvium of the newborn represents the nearly universal shedding of scalp hair during the first

Table 9.1 Causes of telogen effluvium

*Physiological (not pathological)*
- Physiological effluvium of the newborn
- Postpartum

*Injury or stress (pathological)*
- Post-febrile (extremely high fevers, e.g. malaria)
- Severe infection
- Severe chronic illness
- Severe, prolonged psychological stress
Post-surgical (implies major surgical procedure)
Hypothyroidism, hyperthyroidism and other endocrinopathies
Crash or liquid protein diets; starvation
Drugs
retinoids (e.g., acitretin, etretinate, isotretinoin)
anticoagulants (especially heparin)
antithyroid (e.g., propylthiouracil, methimazole)
anticonvulsants (e.g., phenytoin, valproic acid, carbamazepine)
heavy metals
- blockers (e.g., propranolol)

6 months of life. This may occur rapidly, resulting in obvious alopecia, or may proceed slowly and imperceptibly. In either case, large numbers of anagen hairs enter telogen within a period of months.

Many adult women suffer from a chronic telogen effluvium with no definable precipitating event. This has been termed ‘chronic telogen effluvium’, and is a diagnosis of exclusion. Many or most of these women have self-limited disease, although the condition may last for several years before spontaneous resolution.

The early stage of androgenetic alopecia has features of a chronic, localized form of telogen effluvium. The vertex and frontal hairs of the balding scalp experience a marked reduction in the length of anagen. A much higher proportion of hairs are thus entering telogen at any given time. Hair shedding is only obvious during the early stages of the balding process, when large, terminal hairs are being shed. Once hairs have miniaturized, the shedding of vellus telogen hairs is not apparent.

Patients with acute forms of telogen effluvium notice hair loss about 3 or 4 months after the precipitating event. This corresponds to the time it takes for a hair to move through catagen and the early stages of telogen. Scalp hair density may appear normal, despite the patient’s complaint of profuse hair loss. If alopecia is clinically obvious, the loss appears diffuse, affecting all parts of the scalp (Figure 9.1). A gentle hair pull will yield several hairs with the depigmented, cornified, clubbed morphology of telogen hair roots. A forcible hair pluck will produce a mixture of normal anagen and telogen hairs, as well as an occasional catagen hair (Figure 9.2). The percentage of telogen hairs will be increased to more than 20%, a criterion without which the diagnosis of telogen effluvium cannot be established with certainty. In the typical case of telogen effluvium, the telogen count does not exceed 50%. However, exceptions to this rule have been reported, and counts can reach 80%. Figures exceeding 80% are inconsistent with a simple case of telogen effluvium.

HISTOLOGICAL FINDINGS

Just as in androgenetic alopecia, the histological diagnosis of telogen effluvium depends more on quantitative than qualitative features. The only abnormality in a ‘pure’ case of telogen effluvium is an increase in the percentage of terminal
telogen follicles. Therefore, the total number of hairs in the specimen will be normal, but there will appear to be a reduced number of terminal hairs when counted at the dermal-fat junction. Fibrous ‘streamers’ (stelae) replace the ‘missing’ terminal hairs at this level (Figures 9.3 and 9.4). These streamers lie beneath the bulbs of the terminal telogen hairs, which are found in the mid-dermis. Once counted, the telogen hairs are increased in number, and the telogen count (number of terminal telogen hairs divided by the total number of terminal hairs) will be greater than 20%. However, a lower number does not exclude the diagnosis of telogen effluvium. If a patient’s normal telogen count happens to be 5%, a telogen count of 15% would be clearly abnormal for that patient. Unfortunately, we do not know the ‘baseline’ telogen counts for individuals, so numbers less than 20% must be regarded as equivocal. Values of 15–20% are often found in cases of chronic, low-grade telogen effluvium. Even lower numbers can be consistent with this diagnosis.

Figure 9.1 This patient with newly diagnosed hypothyroidism demonstrates the typical pattern of telogen effluvium—thinning of hair over the entire scalp, including the occipital and parietal regions.
The size of hairs is not affected in a telogen effluvium. Abnormally large numbers of terminal (large) anagen hairs are converted into terminal telogen hairs, but miniaturization does not occur (Figure 9.5). Therefore, the terminal : vellus hair ratio is normal (greater than 2:1).

In a simple case of telogen effluvium, both crown/vertex and occiput are involved (truly diffuse hair loss). In fact, all body hair will be affected by the disease, which may clinically be evident by thinned eyebrows, pubic hair and axillary hair. If paired biopsy specimens (crown and occiput) are obtained, the histological findings will be similar at both sites.

**Figure 9.2** Portion of a forcible hair pluck (trichogram) obtained from the scalp of a young woman with a telogen effluvium secondary to systemic lupus erythematosus. There is a mixture of normal-appearing anagen and telogen bulbs, but the telogen bulbs are over-represented. When all hairs were counted, the telogen count was 35%. Original magnification x40

**Figure 9.3** Telogen effluvium secondary to retinoid therapy. This specimen, sectioned at the level of the dermal-fat junction, demonstrates a reduced number of terminal anagen hairs. However, the terminal anagen hairs present are roughly similar in diameter. Several catagen/telogen hairs are seen, as well as a few stelae (underlying additional catagen/telogen hairs). After all follicles were counted, the total number was normal but the percentage of telogen hairs was elevated at 40%. Original magnification x40
Telogen effluvium is a non-inflammatory form of hair loss, and no significant inflammation is found. In particular, peribulbar inflammation and inflammation affecting the lower two-thirds of the follicles is absent.

**SUMMARY**

**Clinical correlation:** history of a precipitating event (2 or 3 months prior to hair loss) is often obtainable childbirth, major surgery or severe illness, certain medications, etc.

**Histological findings:**

- Normal *total* number of follicles
- Normal terminal: vellus hair ratio

**Figure 9.4** The same specimen as in Figure 9.3 (T, telogen follicle). Original magnification x100

**Figure 9.5** Telogen effluvium is characterized by an increase in the percentage of telogen hairs *without* a decrease in the overall size of hairs. The two terminal telogen hairs in this field have been sectioned through their secondary hair germs (SHG). Original magnification x200

Telogen effluvium is a non-inflammatory form of hair loss, and no significant inflammation is found. In particular, peribulbar inflammation and inflammation affecting the lower two-thirds of the follicles is *absent.*
• Marked increase in *percentage* of terminal telogen hairs
• Presence of fibrous ‘streamers’ indicating conversion to telogen hairs
• Normal size of follicles; no significant inflammation

**BIBLIOGRAPHY**

Guarrera M, Rebora A. Anagen hairs may fail to replace telogen hairs in early androgenic female alopecia. *Dermatology* 1996; 192:28–31


Trichotillomania is caused by the habitual, compulsive or intentional pulling and plucking of hair. Although it can be found in persons who are mentally or emotionally ill, many patients are healthy. However, with careful questioning, a history of significant stress or turmoil at school, home or work can often be obtained. Frequently, patients are school-aged children, and often both child and parents deny the possibility of pulling or plucking as a cause of hair loss. Therefore, a biopsy specimen can be crucial for establishing the diagnosis with certainty.

Lesions of trichotillomania are often irregularly shaped, with bizarre or geographic outlines. The bald patches are usually sharply demarcated from the surrounding normal scalp, and typically there are several retained short hairs of various lengths within the zone of alopecia (Figure 10.1). The scalp surface is usually not inflamed, but occasionally excoriations from scratching or picking may be present.

**HISTOLOGICAL FINDINGS**

The most distinctive and diagnostic histological feature of trichotillomania is incomplete, distorted follicular anatomy. In the process of plucking, some or all of the follicular epithelium may be removed from the follicle, and the residual tissue collapses to fill the void. Normally, the inner root sheath surrounds either a hair shaft or the oval space occupied by the shaft before processing. A collapsed inner root sheath indicates prior extraction of the hair shaft and is the most common histological defect encountered in trichotillomania (Figures 10.2 and 10.3). When both the shaft and the inner root sheath are extracted, the outer root sheath collapses upon itself. Infrequently, all or most of the epithelium is removed, and the fibrous root sheath along with any retained epithelial fragments and extravasated red blood cells form a vertical column.

Mechanical trauma to the hair frequently propels anagen follicles into the catagen phase. Therefore, increased numbers of catagen and telogen hairs are found in trichotillomania (Figure 10.4). Catagen hairs will be found in areas that have recently been plucked, and telogen hairs are present a few weeks after the traumatic event (Figures 10.5 and 10.6).
Frequently, chunks of pigmented hair matrix or cortex cells are torn from their moorings during the plucking process, and come to rest in superficial portions of the follicles (Figure 10.7). These cells then shrink to form a dark black homogeneous clump called a pigment cast (Figure 10.8). Pigment casts are simply the byproduct of fragmented, ectopic matrix or cortical epithelium.

If the hair matrix and suprabulbar epithelium is injured, but not severely disrupted, the follicle may remain in the anagen phase, producing a hair shaft. However, the shaft that is formed may be distorted in shape, smaller than normal and incompletely cornified. This is termed trichomalacia (Figures 10.9–10.11). Although trichomalacia is very characteristic of trichotillomania, it is not found exclusively in this condition (refer to the section on alopecia areata, Chapter 14).

Pulling or plucking does not incite inflammation but may cause some hemorrhage within the lower portion of the follicle (Figure 10.12). Even very dramatic cases of trichotillomania are remarkably free of an inflammatory

Figure 10.1 Typical lesion of trichotillomania. The site chosen for the biopsy samples is evident

Figure 10.2 Incomplete and distorted follicular anatomy in trichotillomania. A terminal anagen hair has lost its shaft from plucking. The inner root sheath has collapsed upon itself. Original magnification ×400
infiltrate. Very rarely, a few eosinophils are found surrounding the lower portion of a badly traumatized follicle.

**SUMMARY**

*Clinical correlation:* the patient is often a child or teenager, and a history of emotional stress at home, school or work may be elicited. The involved areas are irregularly shaped, sharply marginated patches with some retained short hairs of various lengths.

*Histological findings:*

- Follicles are of normal size
- Total number of hairs (both terminal and vellus) is normal
- Incomplete, disrupted follicular anatomy is *diagnostic*, but not always present
- Increased number of terminal catagen and/or telogen hairs (fibrous ‘streamers’ are also present if telogen hairs are increased in number)
- Trichomalacia and pigment casts often found
• No significant inflammation (peribulbar inflammation *absent*)

**Figure 10.5** Increased numbers of telogen hairs are seen in this specimen from a patient with trichotillomania. Original magnification ×400

**Figure 10.6** Increased numbers of telogen hairs (or underlying stelae) are seen in this specimen from a patient with trichotillomania. The histological differential diagnosis at this level would include telogen effluvium. Original magnification ×40
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Figure 10.7 A clump of non-cornified cortex cells has been stranded at an ectopic level during the plucking process. Eventually, such cells shrink and cornify to form pigment casts. Original magnification ×400

Figure 10.8 A pigment cast within the center of a telogen follicle. Original magnification ×400
**Figure 10.9** Trichomalacia. Two black pigment casts and two trichomalacic shafts can be seen. Original magnification ×400

**Figure 10.10** Trichomalacia. A single trichomalacic shaft is found. Original magnification ×400
**Figure 10.11** Trichomalacia. Another example with a single trichomalacic shaft. Original magnification ×400

**Figure 10.12** Extravasated red blood cells in the bulb of a catagen hair. Original magnification ×200
Like trichotillomania, traction alopecia is a form of mechanical, traumatic alopecia. However, trauma to the hair is usually mild and chronic. Although vigorous scratching or combing may cause traction alopecia, most cases are caused by hairstyles involving tight braiding or banding of the hair. The condition is most common among African American girls whose hair-styles involve nearly continual braiding. However, the identical pattern of hair loss can be seen in persons of all races. Some girls get the condition after wearing tight 'ponytail' type hairstyles.

The hair loss tends to be a peripheral or marginal form of alopecia, i.e., involving the frontal, temporal and parietal margins of the scalp (Figure 11.1). In girls who wear tight braids, perifollicular erythema and pustule formation may be seen. Whether this inflammation is caused by traction or by cosmetics used in conjunction with hair styling is unknown.

Traction alopecia is a biphasic form of hair loss. Initially the hair loss is temporary, hair regrowth occurs and the condition behaves like a non-scarring form of alopecia. However, if excessive traction is maintained for years, the hair loss may eventually become permanent (end-stage or ‘burnt out’; Figure 11.2). There may be a lag period of a decade or more between the period of traction and the onset of permanent hair loss. Therefore, many African American women present in their thirties and forties with a several-year history of persistent, bitemporal or frontal hair loss. These women may deny having worn tight braids since childhood, although often other forms of traumatic styling (e.g. ‘curlers’) have been employed.

**HISTOLOGICAL FINDINGS**

The histological findings in acute, reversible traction alopecia and permanent, long-standing, ‘burnt out’ traction alopecia are entirely different. The acute form, most commonly seen in African American girls or after a short-lived, traumatic hairstyle (such as a hair weave or ‘corn row’), resembles a mild form of trichotillomania. Occasionally, distorted or incomplete follicular anatomy is seen. More often, there is an increase in catagen/telogen hairs, pigment casts and subtle trichomalacia. Follicular numbers are normal.
In end-stage disease, most commonly found in young African American women, the total number of hairs is markedly reduced (Figures 11.3 and 11.4). This is accounted for by the loss of terminal hairs, because vellus hairs are still present in normal numbers (Figures 11.5–11.9). Sometimes the missing follicles appear to have disappeared without a trace, because the dermal collagen appears normal, but often some ‘old’ stelae can be found when deeper levels are examined (Figures 11.10 and 11.11). In many cases, distinct columns of connective tissue replace some follicles, leaving obvious ‘blank spaces’ (Figure 11.12).

The sebaceous glands associated with the remaining hairs are still intact, and often persist in follicular units that seem to have lost all their follicles. The presence of sebaceous glands and relative normality of the dermal architecture, despite marked hair loss, is highly characteristic of end-stage traction alopecia. No significant perifollicular inflammation is present in either early or late disease.

**Figure 11.1** Thinning of hair around the margins of the scalp in a girl with a history of tightly braided hair

**Figure 11.2** ‘Burnt out’ or end-stage traction alopecia in a 30-yeaf-old woman with permanent hair loss, confirmed by biopsy
Clinical correlation:

Early or acute disease—patient is often an African American child whose hair is tightly braided; there is alopecia of the scalp margin or around braids.

End stage disease—patient is usually an African American woman with symmetric thinning of the temples and/or frontal regions.

Histological findings:

- Normal size of follicles
- Total number of hairs (both terminal and vellus) is normal
- The most prominent finding is increased number of terminal catagen and/or telogen hairs (fibrous ‘streamers’ may be present if telogen hairs are increased in number)
- Trichomalacia and pigment casts occasionally found
- Incomplete, disrupted follicular anatomy rarely found

**Summary**

*Figure 11.3* Mild traction alopecia in an adult woman. The number of terminal anagen hairs is reduced in the alopecic zone (seen here) as compared with normal-appearing perilesional skin (see *Figure 11.4*). Original magnification ×40

*Figure 11.4* Normal-appearing perilesional skin contrasts with the lesional specimen shown in *Figure 11.3*. Original magnification ×40
• No significant inflammation (peribulbar inflammation absent)

End-stage or ‘burnt out’ disease (Figure 11.13)
• Marked decrease in total number of follicles and terminal follicles, with retention of vellus hairs
• Dermal collagen appears relatively normal except for occasional fibrous tracts at sites of former follicles
• Many or most follicular units still have associated sebaceous glands
• No significant inflammation (peribulbar inflammation absent)

**BIBLIOGRAPHY**

Figure 11.10 ‘Old’ stelae in a specimen from a patient with end-stage traction alopecia. When sectioned transversely, these stelae appear as roughly oval condensations of connective tissue containing a few small vascular spaces. Original magnification ×200

Figure 11.11 ‘Old’ stela in a specimen from another patient with end-stage traction alopecia. Original magnification ×400

Steck WD. Telogen effluvium: a clinically useful concept, with traction alopecia as an example. Cutis 1978; 21:543–8
Figure 11.12 Columns of connective tissue marking the sites of former follicles are quite obvious in this case of traction alopecia. In some cases, each column may represent the former site of an entire follicular unit. Original magnification ×100

Figure 11.13 Typical ‘end-stage’ traction alopecia. Original magnification ×200
CHAPTER 12
Postoperative (pressure-induced) alopecia

This form of hair loss is seen most commonly in patients who have undergone lengthy surgical procedures, and had one portion of their scalp (usually the occiput) in prolonged contact with the operating table. Postoperative alopecia can occur at any age and is often associated with gynecological and open-heart procedures requiring tracheal intubation. Less commonly, the condition is found in patients who sustain blunt trauma to the scalp. Postoperative alopecia typically presents as a solitary, roughly oval patch on the upper occiput (Figure 12.1). Early in the course of the condition, erythema and induration are found in the central portion of the lesion. Nearly total hair loss with fairly sharp demarcation from the surrounding scalp is found just a few weeks after the initial trauma. Usually complete hair regrowth occurs, although several cases of permanent (i.e. cicatricial) hair loss have been reported.

HISTOLOGICAL FINDINGS

The histological findings in postoperative alopecia change as the lesion evolves. Early in the course of the disease, before hair loss is complete, vascular thrombosis, inflammation and destruction may be seen in the dermis. Alopecia develops up to 28 days following the surgical procedure. In the typical case of postoperative alopecia, nearly all terminal follicles will be in the catagen or telogen phases (Figures 12.2 and 12.3). This synchronized conversion of most or all terminal hairs to the catagen/telogen phase is highly characteristic of postoperative alopecia. Trichomalacia may be present, but not the distorted or incomplete follicular anatomy sometimes found in trichotillomania (Figure 12.4). Pigment casts are also commonly found (Figure 12.5). Melanin pigment is usually found in the collapsed root sheaths below catagen/telogen follicles.

Variable degrees of dermal fibrosis and chronic inflammation are present in the papillary and upper reticular dermis. Focal vascular and tissue necrosis may be present along with an associated chronic inflammatory infiltrate. Fat necrosis is often found, associated with an infiltrate of foamy macrophages and mononuclear cells (Figure 12.6). Inflammation is mild relative to the degree of
apparent tissue damage. The inflammation does not seem to be centered around hair follicles, but is usually associated with foci of vascular and tissue necrosis.

**SUMMARY**

*Clinical correlation:* a patch of occipital hair loss occurring within a few weeks of a prolonged surgical procedure requiring general anesthesia.

*Histological findings:*

- Almost all hairs in the catagen or telogen phase
- Trichomalacia
- Melanin pigment in collapsed fibrous root sheaths
- Vascular thrombosis or necrosis with a relatively mild perivascular and perifollicular inflammatory infiltrate
- Fat necrosis with secondary infiltration by foamy macrophages and a few lymphoid cells
Figure 12.3 Postoperative alopecia, as in Figure 12.2. Two catagen hairs can be seen. Original magnification ×200

Figure 12.4 Trichomalacia in postoperative alopecia. Original magnification ×400

Figure 12.5 Pigment cast in a patient with postoperative alopecia. Original magnification ×400

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Figure 12.6 Fat necrosis with reactive granulomatous inflammation in a patient with postoperative alopecia. Original magnification ×400

CHAPTER 13
Temporal triangular alopecia

Also known as congenital triangular alopecia, temporal triangular alopecia may be present at birth or acquired during the first decade of life. Lesions outside the temporal area, and those acquired in adulthood, can rarely occur. The lancet-shaped lesions (Figure 13.1) are a few centimeters in width, may be unilateral or bilateral, and are oriented so that the tip of the ‘lancet’ points superiorly and posteriorly. Lesions appear hairless, but very fine vellus hairs can be seen with magnification. Once present, the patches of alopecia persist for life.

Microscopically, there are normal numbers of follicles, but almost all are vellus hairs. The small size of the hairs necessitates transverse sectioning for adequate assessment (Figures 13.2–13.6). All other features of the epidermis, dermis and other adnexae are entirely normal, and inflammation is absent. The condition appears to be a process of follicular miniaturization confined to a small, lancet-shaped area of the temporal region, resulting in the characteristic ‘bald’ spot. However, fibrous ‘streamers’ (stelae) as found in androgenetic alopecia are not found in temporal triangular alopecia (Figure 13.7). Congenital cases presumably undergo the process of follicular miniaturization in utero (or terminal hair formation never occurs).

SUMMARY

Clinical correlation: lancet-shaped bald spot (may be bilateral) discovered on the temporal region of a newborn or young child; the remainder of the scalp is normal.

Histological findings:

- Normal total number of hairs
- Few, if any, terminal hairs
- Almost all hairs are vellus hairs
- No fibrous ‘streamers’ (stelae)
- No significant inflammation or other epidermal or dermal abnormality
**Figure 13.1** Temporal triangular alopecia. This unilateral lesion was first noted when the patient was 3 years old. The condition may also be congenital and/or bilateral.

**Figure 13.2** Temporal triangular alopecia. A section through the deep dermis reveals very few follicles. Original magnification ×40

**BIBLIOGRAPHY**


Figure 13.3 A section through the upper dermis (compare with Figure 13.2) in temporal triangular alopecia demonstrates a normal number of follicles, almost all of which are very small. Original magnification ×40

Figure 13.4 A vertical specimen from a patient with temporal triangular alopecia reveals a few vellus follicles. It is impossible to quantify the total number and average size of follicles using vertical sections. Original magnification ×100

Figure 13.5 The small size of hairs in lesional skin from a patient with temporal triangular alopecia is easily appreciated when compared to hair diameters in normal, perilesional scalp (see Figure 13.6). Original magnification ×100

Figure 13.6 A specimen showing normal, perilesional scalp in a patient with temporal triangular alopecia. Original magnification ×100

Figure 13.7 Temporal triangular alopecia. The dermis below the miniaturized follicles appears normal, and stelae are not seen. Original magnification ×100

Alopecia areata is a very common form of non-scarring hair loss, affecting 1% or more of the population. The disease can affect any part of the body (Figure 14.1), but scalp hair loss is the usual complaint. Severity ranges from a small, circumscribed bald spot to total scalp hair loss (alopecia totalis), and even total body hair loss (alopecia universalis). Both the clinical course and histopathological features of relatively mild disease may differ significantly from severe disease (alopecia totalis or universalis).

Several patterns of partial hair loss can be found, including circumscribed (isolated oval patches; Figures 14.2 and 14.3), reticular (innumerable small patches), ophiasis (marginal) and diffuse. In the very unusual diffuse form, hair loss occurs over much of or the entire scalp, but circumscribed bald spots do not form. However, hair loss is seldom so uniform or symmetrical that it cannot be distinguished from a telogen effluvium. If the hair steadily and rapidly thins, the diagnosis of alopecia totalis in evolution is more appropriate than that of diffuse alopecia areata (Figure 14.4). Even in alopecia totalis or universalis, isolated hairs or tufts of hairs may continue to grow.

The involved scalp is usually normal in color but may show slight erythema or edema. Short hairs that taper as they approach the scalp are called ‘exclamation mark hairs’, and if present are very characteristic of alopecia areata. Because of shaft narrowing and hypopigmentation near the scalp surface, exclamation mark hairs appear to ‘float’ on the scalp surface (Figures 14.5 and 14.6). When hairs are gently pulled from the edges of an expanding lesion of alopecia areata, many telogen hairs and/or tapered ‘pencil point’ shafts can be easily extracted (Figure 14.7). Alopecia areata seems to affect pigmented hairs preferentially, and hair regrowth may occur with depigmented hairs (Figure 14.8).

**HISTOLOGICAL FINDINGS**

Very recent or rapidly progressive hair loss presents a very different histological picture from long-standing, well-established disease. For the purpose of this text, the pathology of alopecia areata will be divided into three ‘stages’, a concept that is accepted by other authorities such as David A. Whiting. These stages, which will be called acute, subacute and chronic, reflect the evolution of the
disease with the passage of time. Different stages of disease may be present at the same time at different sites on the same scalp. In any given patient, separate lesions of alopecia areata may begin, evolve, remit and recur independently of one another.

The **acute** stage is seen in rapidly progressive disease or disease of recent onset, as is found in evolving alopecia totalis or at the advancing margin of an enlarging bald spot. The total number of follicles appears normal. Several affected hairs are still terminal anagen follicles, with bulbs in the fat or deep

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**Figure 14.1** Alopecia areata affecting the beard area. This patient had no scalp hair loss

**Figure 14.2** Typical circumscribed lesion of alopecia areata

**Figure 14.3** Circumscribed lesion of alopecia areata
dermis. However, several follicles have entered the catagen/telogen phase (Figures 14.9–14.12). Some (but seldom all) anagen phase and early catagen phase hairs demonstrate a peribulbar mononuclear cell inflammatory infiltrate. Both CD4+ and CD8+ T cells are found, but the relative proportion of these cells has not yet been carefully correlated with the histological ‘stage’ (Figure 14.13). However, CD8+ cells are more likely to invade the epithelium (exocytosis) than

Figure 14.4 Alopecia totalis in evolution. This patient had rapid, diffuse hair loss progressing over a period of just a few months

Figure 14.5 Patch of alopecia areata with several ‘exclamation mark’ hairs
CD4+ cells. In some cases, ‘peribulbar inflammation’ may be subtle or even absent, and other histological features are required to establish the diagnosis. The presence of peribulbar inflammation is helpful but not essential to making the diagnosis of alopecia areata.

Small numbers of eosinophils may be present, but plasma cells are unusual (Figures 14.14 and 14.15). The infiltrate may invade the dermal papilla and hair matrix, resulting in damage to matrix cells. Especially when exocytosis of inflammatory cells into the hair matrix occurs, but sometimes in its absence as well, the matrix appears blurred and somewhat disorganized (Figures 14.16 and 14.17).

There may be both intercellular (spongiosis) and intracellular edema. This change may be subtle or quite prominent, and can affect the suprabulbar as well as the bulbar zone (Figures 14.18 and 14.19). Some anagen follicles show vacuole formation or necrosis of matrix cells located just above the upper pole of the dermal papilla, corresponding to the site of early hair cortex formation (Figure 14.20). This can result in small cystic spaces filled with acantholytic, necrotic cells. Focal matrix cell vacuolization and necrosis is a characteristic feature of alopecia areata, but it affects relatively few follicles and is found in

**Figure 14.6** ‘Exclamation mark’ hair. This hair was easily pulled from the scalp. The wider, distal end (right side) shows a fracture, and the proximal end is narrow and hypopigmented. Most exclamtion mark hairs are in the telogen phase

**Figure 14.7** Tapered, ‘pencil point’ hair gently extracted from a patient with rapidly progressive alopecia areata
only a minority of cases. Nuclear pyknosis and cell death (apoptosis) occurs not only in matrix keratinocytes, but also in outer root sheath cells and bulbar melanocytes. Amorphous clumps of pigment (pigment casts) are occasionally found within the follicular epithelium as a byproduct of hair matrix degeneration (Figure 14.21).

Figure 14.8 Alopecia totalis in evolution, with early sparing of hypopigmented hairs. This patient had black, but graying hair until his hair loss began. The gray hairs were initially spared, but eventually they too were shed.

Figure 14.9 ‘Acute’ alopecia areata. The number and size of hairs appears normal, but there is an increase in the number of catagen/telogen hairs. Original magnification ×40
As a result of damaged bulbar melanocytes and keratinocytes, pigment incontinence is often seen in the dermal papilla and follicular sheath of affected hairs.

The second or subacute ‘stage’ of alopecia areata does not correlate well with the extent or duration of clinical disease. This stage is the one most commonly encountered by pathologists. When all follicles are counted, requiring examination of transverse sections at both deep and superficial levels, the total number of follicles appears normal. However, in virtually all cases, there is a marked increase in the percentage of catagen/telogen hairs, often exceeding 50% of total follicles. Some of the catagen/telogen hairs are still terminal (large) hairs, and a few deeply seated, terminal anagen hairs may also be seen. Peribulbar inflammation tends to subside after the hair has entered the catagen phase, so that late catagen and telogen hairs are usually free of inflammation (Figure 14.22). However, a few inflammatory cells may persist near the lower portion of some catagen/telogen follicles, as well as within the collapsed sheaths (Figures 14.23–14.25).

Despite the peribulbar inflammation and matrix injury, some anagen hairs continue to produce hair shafts. Some of these shafts show trichomalacia, and are
smaller, incompletely cornified and distorted in shape. Many follicles produce shafts that become progressively smaller in volume and cross-sectional dimension. These hairs gradually taper down to a point (Figure 14.26), resulting in an extremely fragile constriction. They represent the ‘pencil point’ hairs which fall from the scalp in great numbers. A transverse section through the site of constriction may show a minute or absent (Figure 14.27) hair shaft. The term

Figure 14.12 ‘Acute’ alopecia areata, with several slightly inflamed stelae. Original magnification ×200

Figure 14.13 CD3+ T cells (all T lymphocytes stain brown) surround and invade the epithelium of an anagen hair. Varying proportions of CD4+ and CD8+ T cells may be present in such an infiltrate. Original magnification ×400

Figure 14.14 Eosinophils join the lymphocytic infiltrate surrounding the bulb of this catagen hair. Original magnification ×400
anagen arrest (or anagen effluvium) has been applied to those forms of alopecia characterized by the rapid tapering and shedding of large numbers of anagen hairs. Anagen arrest is characteristic of chemotherapy-induced alopecia, but the initial stages of alopecia areata (when terminal hairs are still present) also have features of an anagen arrest.

Figure 14.15 Eosinophils join the lymphocytic infiltrate surrounding the bulb of this anagen hair. Original magnification ×1000

Figure 14.16 The hair matrix of this inflamed bulb appears blurred and disorganized. Original magnification ×400

Figure 14.17 Despite the scanty peribulbar infiltrate, the hair matrix appears blurred and somewhat disorganized. Original magnification ×400
However, after producing a dystrophic (or no) shaft for a period of time, these anagen hairs eventually enter the catagen/telogen phase.

After remaining in the telogen phase for a period of time (about 100 days in a normal follicle; an unknown length of time in alopecia areata), hair follicles re-enter the anagen phase. Unless the disease has spontaneously subsided, an
inflammatory infiltrate again confronts the newly forming anagen hairs. This results in a repetition of the pathological process: peribulbar inflammation, disturbance of anagen hair growth and precipitation into the catagen/telogen phase. As this process repeats itself over and over, the duration of anagen becomes briefer, the anagen hairs miniaturize, and an increasingly large percentage of hairs will be found in the catagen/telogen phase, sometimes approaching 100% of the total (Figure 14.28).

The third histological stage, which here is referred to as chronic, is found in stable, long-standing bald patches and in well-established alopecia totalis or universalis. Terminal anagen hairs, with or without surrounding mononuclear infiltrate, are rare. Thus, the most familiar histological finding of alopecia areata may be absent. In addition, all the follicles may become miniaturized, but the total number of follicles remains normal (Figures 14.29 and 14.30). This remains true for years or even decades, but eventually follicular dropout may occur (an example of the ‘biphasic’ pattern of permanent alopecia; see Figure 4.1). When follicles are miniaturized, they are often missed on routine vertical sectioning. Transverse sections are required to examine and count all follicles.

The miniaturized anagen follicles found in ‘chronic’ disease are situated in the mid- to lower dermis, usually slightly deeper than normal vellus hairs.
Figure 14.31 Normally, anagen hairs develop through a series of developmental stages named anagen I-VI. The majority of the miniaturized, diseased follicles in alopecia areata develop to a stage resembling anagen III or IV, but no further. These small, abnormal follicles have been called ‘nanogen’ hairs (*nanos*, Greek for ‘dwarf’), and are a distinctive feature of long-standing alopecia areata. Nanogen hairs are not merely small. Their rapid and distorted life cycle makes them difficult to categorize as anagen, catagen or telogen hairs. The reasons for this are given below.

Nanogen hairs have an epithelial matrix that is small relative to the size of the dermal papilla, which is also reduced in volume. The nuclei of the papilla become rounder and the cells more compact, much as they do in the catagen/telogen phase (Figure 14.32). Nanogen hairs can be identified in transverse section because they have thin inner and outer root sheaths (two or three cell layers), but no central hair shaft, or at most an extremely fine, incompletely cornified shaft (Figures 14.33–14.37).

Furthermore, they may simultaneously demonstrate morphological features of both anagen and catagen/ telogen phases. The bulb may resemble that of an anagen hair (basophilic cells with mitotic activity) while the suprabulbar portion
shows features of a catagen hair. Conversely, the bulb may show features typical of a catagen hair while the suprabulbar zone possesses an anagen-like inner root sheath with trichohyaline granules (Figures 14.38–14.41). Nanogen hairs with ‘anagenlike’ bulbs may have some matrix cells in mitosis while others are undergoing apoptosis (Figures 14.42 and 14.43). Thus, features of active growth (mitotic cells) and involution (apoptotic cells) are seen simultaneously. The

**Figure 14.24** Residual inflammation surrounds the suprabulbar zone of an early catagen hair. Original magnification ×400

**Figure 14.25** The stelae below catagen/telogen hairs may also show residual inflammation. Original magnification ×400
strange histological characteristics of nanogen hairs signify a profound disorder of cell cycling that is peculiar to alopecia areata. In most cases, the majority of nanogen hairs will be found in the catagen/telogen phase (Figures 14.44–14.48), so that ‘anagen-like’ nanogen hairs may be difficult to find. Nanogen hair bulbs may be surrounded and sometimes invaded by inflammatory cells, but generally the degree of inflammation is mild and often subtle. The most inflamed nanogen bulbs are those with ‘anagen-like’ and early ‘catagen-like’ bulbs (Figures 14.49–14.51). Even in the ‘chronic’ stage of alopecia areata there can be a surprising amount of perifollicular inflammation and infiltration of hair follicles by mononuclear cells. However, as in the acute stage, the telogen follicles are relatively spared from an inflammatory infiltrate. In the chronic phase, the majority (or all) of the follicles in a specimen may be in the catagen/telogen phase, and so inflammation might be entirely absent (Figure 14.52). This results in a picture of ‘non-inflammatory’ alopecia areata, a surprising finding in patients who may have very severe and dramatic clinical disease.

Below each miniaturized follicle is a collapsed fibrous root sheath (stela) (Figure 14.53). It may contain numerous capillaries, a few inflammatory cells and clumps of melanin, but these findings are less common than in the fibrous sheaths of terminal hairs found in ‘acute’ disease. Unless they are inflamed, these collapsed sheaths appear identical to the ‘fibrous streamers’ described in androgenetic alopecia and telogen effluvium. Atrophy of sebaceous glands is seldom seen in alopecia areata.

**SUMMARY**

*Clinical correlation:* patchy or circumscribed hair loss without evidence of inflammation; may result in total scalp or body alopecia; lesions appear suddenly and may expand rapidly; ‘exclamation mark’ hairs may sometimes be seen.

*Histological findings:*
Early, severe or progressive disease (‘acute’ and ‘subacute’) –

- Normal total number of hairs
- Peribulbar, mononuclear cell infiltrate (with occasional eosinophils), predominantly affecting terminal anagen and catagen hair bulbs

Figure 14.27 This anagen hair has produced a cornified inner root sheath, but no cortex/shaft formation has occurred. Original magnification ×1000

Figure 14.28 The percentage of telogen hairs in ‘subacute’ disease can approach 100%. The remaining anagen hairs have begun to miniaturize. Original magnification ×100
Occasional exocytosis of inflammatory cells into bulbar epithelium

Degenerative changes (nuclear pyknosis, inter- and intracellular edema, vacuole formation) of hair matrix, especially the lower, central matrix

Increased number of terminal catagen and telogen hairs

Increased number of miniaturized hairs

Trichomalacia and marked narrowing of hair shafts

**Figure 14.29** Alopecia areata, ‘chronic’ stage. The majority of the follicles are in the telogen phase. Only a few terminal anagen hairs are present, and peribulbar inflammation may be difficult or impossible to identify. Original magnification ×40

**Figure 14.30** Alopecia areata, ‘chronic’ stage. See Figure 14.29. Original magnification×100

- Occasional exocytosis of inflammatory cells into bulbar epithelium
- Degenerative changes (nuclear pyknosis, inter- and intracellular edema, vacuole formation) of hair matrix, especially the lower, central matrix
- Increased number of terminal catagen and telogen hairs
- Increased number of miniaturized hairs
- Trichomalacia and marked narrowing of hair shafts
Long-standing and stable disease (‘chronic’)–

- Majority of hairs in catagen or telogen phases
- Numerous miniaturized, ‘arrested’, rapidly cycling hairs (nanogen hairs)

**Figure 14.31** The bulb of this miniaturized anagen hair is located in the mid-dermis. Original magnification ×100

**Figure 14.32** Nanogen hair. The bulbar epithelium of this inflamed hair is only a few cells in thickness, and is thin relative to the diameter of the papilla. Original magnification ×400
• Mild, peribulbar, mononuclear cell infiltrate around those nanogen hairs having anagen-like or catagenlike bulbs

**BIBLIOGRAPHY**


**Figure 14.34** Nanogen hair. Transverse section at the level of the suprabulbar zone. There is no central shaft formation. Original magnification ×1000
Figure 14.35 Nanogen hair. Transverse section at a higher level of the suprabulbar zone. The inner root sheath has cornified, but there is no shaft. (Same follicle as shown in Figures 14.33 and 14.34.) Original magnification ×1000

Figure 14.36 A vertically sectioned ‘anagen-like’ nanogen hair corresponds to the type of hair shown in Figures 14.33–14.35. The extremely small size of such a hair is best appreciated by comparing it to the bulb of a normal anagen hair (Figure 14.37), photographed at the same magnification. Original magnification ×400

Figure 14.37 Normal anagen hair. Original magnification ×400
Figure 14.38 This transversely sectioned nanogen hair has the bulb of a catagen hair. Original magnification ×400

Figure 14.39 Catagen-like suprabulbar zone of the same nanogen hair shown in Figure 14.38. Original magnification ×400
Figure 14.40 More superficial level of the same nanogen hair seen in Figures 14.38 and 14.39, but here showing the anagen-like features of inner root sheath and cortex formation. Original magnification x400

Figure 14.41 More superficial level of the nanogen hair shown in Figure 14.40. Original magnification x400
Figure 14.42 This greatly magnified ‘anagen-like’ nanogen hair has a matrix that is less basophilic than normal, yet it produces inner root sheaths (found on more superficial sections). In addition, both mitotic and apoptotic cells are found. Original magnification ×1000
Figure 14.43 Another ‘anagen-like’ nanogen hair bulb, as in Figure 14.42. Original magnification ×1000
Figure 14.44 A ‘catagen-like’ nanogen hair adjacent to a terminal anagen hair. Note the dramatic difference in size, and the more superficial location of the nanogen hair. Original magnification ×100
**Figure 14.45** Another vertically sectioned ‘catagen-like’ nanogen hair. Original magnification ×200

**Figure 14.46** A follicle similar to those in Figures 14.44 and 14.45, sectioned transversely through the bulb. Original magnification ×400
Figure 14.47 Another transversely-sectioned ‘catagen-like’ nanogen hair, showing the suprabulbar area. Original magnification ×400
Figure 14.48 Another transversely-sectioned ‘catagen-like’ nanogen hair, sectioned through the upper follicle. The central zone of cornification represents residual inner root sheath formation. Original magnification ×400
Figure 14.49 An unusually dense lymphocytic infiltrate surrounds the bulb of an ‘anagen-like’ nanogen hair. Original magnification ×400

Figure 14.50 An unusually dense lymphocytic infiltrate surrounds the bulb of a ‘catagen-like’ nanogen hair. Original magnification ×400
Figure 14.51 Immunoperoxidase stain for CD3+ T cells high-lights the lymphocytes surrounding the bulb of a nanogen hair. This stain may be helpful in cases where inflammation is scantly. Original magnification ×200

Figure 14.52 In this specimen demonstrating the ‘chronic’ histological phase of alopecia areata, the majority of the miniaturized follicles are in the telogen phase, and their bulbs are not inflamed. Original magnification ×100
Figure 14.53 Three transversely sectioned stelae (‘streamers’) in a patient with long-standing alopecia totalis. Original magnification ×200
CHAPTER 15
Syphilitic alopecia

Syphilis, the ‘great imitator,’ is capable of causing alopecia with several clinical and histological patterns. Alopecia found in secondary syphilis may be associated with other cutaneous lesions, or may be the only external manifestation of syphilis. The pattern of hair loss can be patchy (‘moth-eaten’), diffuse or a combination of the two (Figure 15.1).

When papulosquamous lesions of secondary syphilis are found on the scalp in association with alopecia (‘symptomatic’ syphilitic alopecia), the histological findings are usually those typically associated with lesions of secondary syphilis. These findings include: a perivascular and perifollicular infiltrate of lymphocytes, histiocytes and often numerous plasma cells; involvement of both superficial and deep dermal vascular plexuses; vascular dilatation and prominent, swollen endothelial cells; and frequent epidermal involvement with epidermal hyperplasia, spongiosis, infiltration with neutrophils and interface inflammation.

Hair loss in the absence of papulosquamous lesions of the scalp is termed ‘essential’ syphilitic alopecia. The clinical pattern of hair loss may be ‘moth-eaten’ or diffuse (as in telogen effluvium). Syphilis is one cause of telogen effluvium, and in some cases of diffuse syphilitic alopecia the histological pattern is that of a typical telogen effluvium (described in Chapter 9). However, more often the pattern is an inflammatory, predominantly peribulbar and suprabulbar, non-scarring process closely mimicking alopecia areata (Figures 15.2–15.6). Catagen/telogen follicles are increased in number, initially as terminal hairs, but eventually as miniaturized follicles. A mononuclear cell infiltrate surrounds the inferior segment of the follicle or involves the fibrous tracts below catagen/telogen hairs (Figure 15.5). A minority of specimens will show a solitary peribulbar lymphoid aggregate, and in some cases large numbers of lymphocytes infiltrate the outer root sheath at the isthmus level (Figure 15.6).

In well-established syphilitic alopecia, the majority or all the follicles have miniaturized (Figures 15.7 and 15.8). The peribulbar infiltrate tends to be rather scanty, and plasma cells may be difficult to find. Because so many follicles have miniaturized, finding and counting involved hairs is best achieved with transverse sectioning. Special stains for spirochetes do not reveal organisms.
Eosinophils in the infiltrate are unusual, but in many cases plasma cells are present in small numbers.

Syphilitic alopecia and alopecia areata may have the following features in common:

1. Sparing of the epidermis
2. Infiltration of lymphocytes around the lower segment of follicles, in fibrous tracts, or within the follicular epithelium
3. Presence of small or abnormal anagen hairs
4. Markedly increased numbers of catagen/telogen follicles

The presence of plasma cells and lymphocytic infiltration of the outer root sheath is more likely to be seen in syphilitic alopecia than in alopecia areata. Serological tests for syphilis and a prompt and complete response to antitreponemal antibiotics help to confirm the diagnosis.

**SUMMARY**

Clinical correlation: a diffuse (resembling telogen effluvium) or a diffuse but ‘moth-eaten’ alopecia. Other signs and symptoms of syphilis may be present.

Histological findings:

‘Symptomatic’ alopecia (associated with papulosquamous lesions)—

- Histological findings similar to typical papulosquamous lesions of secondary syphilis
- Superficial and deep perivascular and perifollicular infiltrate composed of plasma cells, lymphocytes and histiocytes
- Vascular dilatation and endothelial cell prominence

*Figure 15.1* Syphilitic alopecia that is both diffuse (affecting the entire scalp) and patchy (affecting some areas more than others). This pattern is typical. Photograph courtesy of Timothy Berger, MD
Figure 15.2 ‘Early’ syphilitic alopecia showing an increased number of catagen hairs. Original magnification ×40. Slide courtesy of James W. Patterson, MD

Diffuse alopecia secondary to telogen effluvium—

- Identical to ‘telogen effluvium’ as discussed earlier

‘Essential’ alopecia, ‘moth-eaten or diffuse, inflammatory—

- Histological findings associated with typical lesions of secondary syphilis are absent
- Resembles alopecia areata: peribulbar mononuclear cell infiltrate, miniaturization of hairs, increased number of catagen/telogen hairs (approaching 100% in some cases)
- Occasional plasma cells may help differentiate from alopecia areata
- Perifollicular and outer root sheath inflammation may extend up to the follicular isthmus
- Positive serological test for syphilis and favorable response to antibiotics
Figure 15.3 ‘Early’ syphilitic alopecia. Original magnification ×100. Slide courtesy of James W. Patterson, MD

BIBLIOGRAPHY

Figure 15.4 A relatively mild lymphocytic infiltrate with occasional plasma cells surrounds the lower portion of a catagen hair in ‘early’ syphilitic alopecia. Original magnification ×200. Slide courtesy of James W Patterson, MD
Figure 15.5 Residual inflammation is found in the collapsed root sheath (stela) below a catagen hair. Original magnification ×400
Figure 15.6 A collection of lymphocytes has entered the epithelium of this telogen hair. Original magnification ×400

Figure 15.7 Section from a patient with well-established syphilitic alopecia. The follicles are very small and inflammation is scanty. Original magnification ×40
Figure 15.8 Section from a patient with well-established syphilitic alopecia. Original magnification ×100
CHAPTER 16
Non-scarring alopecia from systemic lupus erythematosus

Hair loss in systemic lupus erythematosus (SLE) can occur in several forms. Discoid lesions causing scarring alopecia are the most familiar form of alopecia to both clinician and dermatopathologist, and will be discussed further in Chapter 21. Patients with SLE can be severely ill for long periods of time, and so diffuse hair loss can occur in the form of a telogen effluvium, as described in Chapter 9.

A form of hair loss in SLE that is fairly common but has received little attention in the literature is patchy, non-scarring alopecia. This form of hair loss occurs in patients with severe disease, and the underlying diagnosis of SLE has already been established or is suspected. Patches of partial or total hair loss are scattered on the scalp (Figure 16.1), and are associated with mild erythema, but without evidence of scarring (follicular ostia and surface texture appear normal). The hairs remaining in the balding patches are almost all telogen hairs or dystrophic ‘pencil-point’ anagen hairs (Figure 16.2), a finding diagnostic of an ‘anagen arrest’ (described in Chapter 14). If the underlying disease is promptly brought under control, complete hair regrowth occurs.

HISTOLOGICAL FINDINGS

A peribulbar mononuclear cell infiltrate is found around anagen hair bulbs, many of which become miniaturized with the passage of time (Figures 16.3–16.7). The inflammatory infiltrate may be more dense than that found in alopecia areata, but is not always so. The percentage of catagen and telogen hairs is greatly increased, and may approach 100% (Figure 16.8). Pigment incontinence and a mild inflammatory infiltrate are often found in the collapsed root sheaths (stelae) below telogen hairs.

These histological findings are similar to those found in alopecia areata and ‘essential’ syphilitic alopecia, and a diagnosis of lupus erythematosus may not be possible on histological grounds alone. Clinical findings and serological testing may be required to differentiate reliably between these three forms of non-scarring, reversible, inflammatory alopecia. However, some additional histological features may help to distinguish SLE from its histological ‘mimics’. For instance, an increase in dermal mucin is sometimes present (Figure 16.9).
Although increased mucin may be visible with routine staining, colloidal iron or similar stains can accentuate the mucin. Inflammation around eccrine glands and dermal vasculature is often seen in the alopecia of SLE. When inflammation involves dermal blood vessels, extravasated red blood cells are sometimes found (Figure 16.10). If present, an especially dense inflammatory infiltrate supports a diagnosis of lupus erythematosus (Figure 16.11). Finally, focal areas of vacuolar basilar degeneration, affecting the infundibula of follicles, may be found in some cases of SLE alopecia (Figure 16.12).

**SUMMARY**

*Clinical correlation:* the patient is usually a young adult with severe disease, and the underlying diagnosis of SLE has already been established or is suspected. Patches of partial or total hair loss are scattered on the scalp. Mild erythema of the scalp may be present.
Histological findings:

- May closely resemble alopecia areata and ‘essential’ syphilitic alopecia
- Increased number of catagen and telogen hairs in affected areas (may approach 100%)
- Peribulbar mononuclear cell infiltrate, often more dense than that seen in alopecia areata
- Most anagen hairs are small and surrounded by peribulbar inflammation
- Additional possible findings supporting the diagnosis of SLE include:
  
  (a) increased dermal mucin
  (b) vacuolar basilar degeneration of the infundibular epithelium
  (c) peri-eccrine and perivascular lymphoplasmacytic inflammation

Figure 16.3 Terminal anagen hairs still predominate in this ‘early’ lesion of patchy, non-scarring systemic lupus erythematosus. A moderately dense lymphocytic infiltrate surrounds the lower segments of several anagen hairs, but involves interfollicular blood vessels as well. Original magnification ×100

Figure 16.4 Patchy, non-scarring systemic lupus erythematosus (see Figure 16.3). The perivascular infiltrate contains some plasma cells. Original magnification ×400

Figure 16.5 In this ‘advanced’ lesion of patchy, non-scarring systemic lupus erythematosus, the majority of hairs have miniaturized, and their bulbs are found in the lower to mid-dermis. Lymphocytic inflammation is concentrated in the vicinity of follicular units. Original magnification ×40
Figure 16.6 Another example of an ‘advanced’ lesion of patchy, non-scarring systemic lupus erythematosus (see Figure 16.5). Original magnification ×40
(d) extravasated red blood cells around inflamed blood vessels

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**Figure 16.7** Miniaturized anagen hairs in an ‘advanced’ lesion of patchy, non-scarring systemic lupus erythematosus. These small follicles produce little or no hair shaft, and they closely resemble the nanogen hairs of alopecia areata. Original magnification ×200
Figure 16.8 In this ‘advanced’ lesion of patchy, non-scarring systemic lupus erythematosus, the majority of hairs are miniaturized and in the catagen/telogen phase. The inflammation is quite dense as compared with alopecia areata. Original magnification ×200

Figure 16.9 Even without special stains, interstitial dermal mucin is obvious in this specimen from a patient with patchy, non-scarring systemic lupus erythematosus alopecia. Original magnification ×200
Figure 16.10 Deep perivascular inflammation with extravasated red blood cells in a patient with patchy, non-scarring systemic lupus erythematosus alopecia. Original magnification ×400

Figure 16.11 Deep, dense, chronic inflammation in a patient with patchy, non-scarring systemic lupus erythematosus alopecia. Original magnification ×200
Figure 16.12 Mild and focal vacuolar interface change affecting the follicular infundibulum in a patient with patchy, non-scarring systemic lupus erythematosus alopecia. Original magnification ×400
Although loose anagen hair syndrome (LAHS) was first described in the late 1980s, it is a fairly common form of non-scarring hair loss. Lack of familiarity with the condition is the major obstacle to establishing the diagnosis. Most patients are first diagnosed at the age of 2–5 when they are brought to a physician with the complaint of thin, uneven hair with an abnormal texture (Figures 17.1–17.3). Parents often state that the child’s hair ‘won’t grow’.

Anagen hairs can be easily and painlessly extracted from the scalp by gentle traction. Extracted hairs show a characteristic ruffling of the hair shaft cuticle (Figure 17.4). The inner and outer root sheaths are left behind in the scalp when anagen hairs are extracted. Some authors have found an increased telogen count (> 20%) in patients with LAHS, but others have found a decreased count (< 5%). This issue will require more careful study in large numbers of patients.

HISTOLOGICAL FINDINGS

There are no reports of inflammation in biopsy specimens from LAHS patients. The internal root sheath has been reported to show a ‘crumbling degeneration’ as well as ‘premature keratinization’. Crumbling degeneration may refer to the numerous fractures that may occur within the cornified inner root sheath during tissue processing (Figures 17.5 and 17.6). This artifact of processing can also be seen in normal specimens, but is more common and prominent in LAHS. Varying degrees of abnormal (i.e., excessive) artifactual separation can be seen between the hair cuticle and cortex, between the hair cuticle and inner root sheath cuticle, within the inner root sheath, and between the inner and outer root sheaths (Figures 17.7–17.9). This clefting is not specific for LAHS, and similar changes can occasionally be found in specimens from normal scalps. However, artifactual clefting appears to be a prominent and consistent feature found in LAHS.

It must be emphasized that clefting found between the cornified hair shaft and inner root sheath is an artifact of processing that occurs in almost all normal as well as abnormal follicles. It is therefore irrelevant and should be ignored.

Occasionally, incomplete follicular anatomy resembling trichotillomania is found. When this occurs, only the hair shaft is absent, and the residual inner and
outer root sheaths are collapsed (Figure 17.10). This finding is hardly surprising, because the shafts can be easily and painlessly extracted in children with LAHS, and sites of recent depilation may be chosen for biopsy.

Although most hair shafts in LAHS retain the normal oval or circular shape, some shafts have unusual polygonal shapes (Figures 17.11 and 17.12). This corresponds to the grooving of hair shafts that can often be found in LAHS. A few patients with LAHS have more prominent hair shaft grooving, affecting most hairs. In some specimens from patients with LAHS, an increased telogen count is found.

**SUMMARY**

*Clinical correlation:* child with areas of slightly thinned, somewhat unruly hair. Parents complain that child’s hair ‘won’t grow’.

*Histological findings:*

- ‘Degenerative’ changes and abnormal cornification of inner root sheath
- Clefting within the inner root sheath (IRS), between the non-cornified cortex and IRS, between the inner and outer root sheaths, and between the outer root sheath and fibrous sheath
- In some, but not all specimens the following features may be seen: increased telogen count (>20%); absent hair shafts with collapsed root sheaths; unusual hair shaft shapes
- No significant inflammation
BIBLIOGRAPHY


Boyer JD, Cobb MW, Sperling LC, Rushin JM. Loose anagen hair syndrome mimicking the uncombable hair syndrome. *Cutis* 1996; 57:11–12

Chapman DM, Miller RA. An objective measurement of the anchoring strength of anagen hair in an adult with the loose anagen hair syndrome. *J Cutan Pathol* 1996; 23:288–92


*Figure 17.2* A 4-year-old girl with loose anagen hair syndrome. Most reported patients have been girls with blond hair. The hair is somewhat thinned and has a windblown appearance.
Tosti A, Piraccini BM. Loose anagen hair syndrome and loose anagen hair. *Arch Dermatol* 2002; 138:521–2

**Figure 17.3** Unruly, twisted hair in a girl with loose anagen hair syndrome (LAHS). This degree of hair shaft abnormality is exceptional for LAHS. Photograph courtesy of Jeffrey Miller, MD
Figure 17.4 Anagen hair gently extracted from the scalp of a child with loose anagen hair syndrome. The soft hair matrix and suprabulbar zone is prone to kinking during extraction, resulting in a ‘hockey stick’ appearance. There is a ruffling of the hair shaft cuticle where it separates from the inner root sheath. Original magnification ×100
Figure 17.5 Numerous fractures within the inner root sheath are commonly found in loose anagen hair syndrome. Original magnification ×400

Figure 17.6 Inner root sheath in loose anagen hair syndrome, showing numerous fractures. Original magnification ×400
Figure 17.7 Several variations of artifactual clefting can be seen in loose anagen hair syndrome. Here clefting has occurred between the hair cortex and the inner root sheath. Original magnification ×400

Figure 17.8 Artifactual clefting in loose anagen hair syndrome. Here clefts between inner and outer root sheaths are seen. Original magnification ×400
Figure 17.9 A circumferential cleft has occurred within the inner root sheath between Henle’s and Huxley’s layer. Original magnification ×400

Figure 17.10 In this specimen from a child with loose anagen hair syndrome, hair shafts have been previously extracted from two follicles, and the inner root sheaths have collapsed inward. Original magnification ×200. Slide courtesy of Jeffrey Miller, MD
The term _central, centrifugal scarring alopecia_ (CCSA) was recently coined to incorporate several variants of inflammatory, scarring alopecia. The difference between these variants may be due to racial differences or the well-recognized variability in immune responses between individuals. The entities grouped under CCSA include _pseudopelade_ (not Brocq’s pseudopelade), the _follicular degeneration syndrome_, and _folliculitis decalvans_. As variants of CCSA, they have the following features in common:

1. They are chronic and progressive, with eventual spontaneous ‘burn out’ after years or decades;
2. They are predominantly centered on the crown or vertex;
3. They progress in a roughly symmetrical fashion, with the most active disease activity occurring in a peripheral zone of variable width, surrounding a central, alopecic zone;
4. They show both clinical and histological evidence of inflammation in the active, peripheral zone.

Over the years, CCSA has been given several names, because the condition exhibits a spectrum of inflammation. Patients with highly inflammatory disease, as evidenced by pustule formation, crusting, intense erythema and bacterial superinfection, have usually been labeled with ‘folliculitis decalvans’. Patients with more indolent disease, characterized by perifollicular scaling and occasional papule formation, have been labeled with ‘pseudopelade’ or ‘the follicular degeneration syndrome’. Most patients described as having ‘pseudopelade’ have been Caucasians, and most patients labeled with ‘the follicular degeneration syndrome’ have been dark-skinned persons of African descent (Blacks; African Americans). However, clinically and especially histologically, the similarity of CCSA between racial groups far exceeds any differences.

CCSA is the most common form of scarring alopecia in any population that includes significant numbers of Black patients. Among African Americans, CCSA is responsible for more cases of scarring alopecia than all other forms combined. The majority of Black patients with CCSA are women, with a female:
male ratio of about 3:1. The average age at presentation is 36 years for women and 31 years for men, although most patients have had progressive disease for years or decades before they seek medical attention. The disease invariably begins and remains most severe on the crown or vertex of the scalp (Figure 18.1), gradually expanding in a centrifugal fashion. Even when the amount of hair loss is dramatic, symptoms may be mild or absent. Most patients note only mild, episodic pruritus or tenderness of involved areas. Virtually all African American women with CCSA are using or have used chemical hair relaxers for styling purposes, but few men have used anything except pomades. Patients experience progression of the disease even after all chemical treatments (if any) are discontinued. Caustic cosmetics may aggravate the disease or hasten its progression, but cannot fully explain its pathogenesis.

Figure 18.1 ‘Early’ central, centrifugal scarring alopecia emerges as a zone of partial hair thinning centered on the crown or vertex. Permanent hair loss is evidenced by the loss of follicular ostia between remaining follicles

Figure 18.2 Even in fairly advanced central, centrifugal scarring alopecia, evidence of clinical inflammation such as papules and pustules may be absent, and the patient may be misdiagnosed as having androgenetic alopecia male ratio of about 3:1. The average age at presentation is 36 years for women and 31 years for men, although most patients have had progressive disease for years or decades before they seek medical attention. The disease invariably begins and remains most severe on the crown or vertex of the scalp (Figure 18.1), gradually expanding in a centrifugal fashion. Even when the amount of hair loss is dramatic, symptoms may be mild or absent. Most patients note only mild, episodic pruritus or tenderness of involved areas. Virtually all African American women with CCSA are using or have used chemical hair relaxers for styling purposes, but few men have used anything except pomades. Patients experience progression of the disease even after all chemical treatments (if any) are discontinued. Caustic cosmetics may aggravate the disease or hasten its progression, but cannot fully explain its pathogenesis.

The crown and/or vertex of the scalp shows a symmetrical zone of partial or complete alopecia. When inflammation is subtle, CCSA may be incorrectly diagnosed as androgenetic alopecia (Figure 18.2). The follicular ostia between remaining hairs are obliterated, and the scalp is smooth and shiny, evidence of scarring alopecia.
A few isolated hairs, some showing polytrichia (tufting), may be stranded in the otherwise denuded central zone. Early or mild disease may be manifest as a partially bald patch only a few centimeters in diameter (as in Figure 18.1). Long-standing or severe disease can result in hair loss covering the entire crown of the scalp. As the examiner moves from the center of involvement, a gradual increase in hair density is noted, merging gradually with the surrounding normal scalp. In this transitional zone, a few inflammatory, follicular papules may be found. Even ‘normal’ scalp skin may show small foci of alopecia and an occasional follicular papule or some perifollicular scaling.

Pustules and crusting may be found in the minority of patients who suffer from rapidly progressive disease or bacterial superinfection (‘folliculitis decalvans’; Figure 18.3). It should be noted that some authors have used the term *folliculitis decalvans* to describe any form of dramatic or highly inflammatory scarring alopecia.

Pustular lesions come and go during the course of the disease. The pustules of CCSA are most likely a manifestation of bacterial superinfection and/or the immune response of the patient to degenerating follicular components. However, some authors have argued that ‘folliculitis decalvans’ is a primary staphylococcal infection. In any case, a short course of antibiotics or systemic
corticosteroids can temporarily eliminate the purulent component of CCSA. Inflamed lesions can be multifocal, but eventually merge into one larger patch on the crown or vertex. An expanding zone of alopecia with peripheral pustules and crusting is not diagnostic of CCSA, and can be seen in tinea capitis (kerion) and true bacterial infections of the scalp. When these latter conditions are excluded, the majority of patients will prove to have CCSA.

**HISTOLOGICAL FINDINGS**

If the central, bald zone is sampled, the histological findings will be those of a ‘burnt out’ scarring alopecia (see Chapter 22). The most productive area for biopsy is the peripheral, partially alopecic fringe, but even clinically ‘normal’ scalp may be diseased at the microscopic level. Not every follicle in a given area is involved simultaneously. A 4-mm punch biopsy specimen may contain only one or two ‘diagnostic’ follicles. This is because the involved follicles are selectively destroyed, with sparing of the relatively normal follicles. Although an occasional vertical section may reveal an involved hair, in most cases transverse sections at several levels are required for a definitive diagnosis.

**Figure 18.4** The sections shown in Figures 18.4–18.7 are well below the level of the isthmus and eccrine coils. Here, two follicles are shown that have undergone premature desquamation of the inner root sheath. Original magnification ×400.
The most distinctive and earliest histological finding is premature desquamation of the inner root sheath. This finding has been most often demonstrated in Black patients, but can be found in all races. Normally, the inner root sheath desquamates and disappears within the mid- to upper isthmus, which is usually located in the upper half of the dermis. In many cases of CCSA, the inner root sheath of an involved follicle desquamates in the lower half of the dermis, sometimes as low as (or below) the dermal-subcutaneous junction (Figures 18.4–18.7). Desquamation of the inner root sheath below the level of the eccrine coils can be regarded as ‘premature’ because it is rarely found in a normal scalp. Premature desquamation of the inner root sheath can be observed in follicles that are otherwise normal, suggesting that it represents a very early phase of the disease. It can occasionally be seen in other conditions in which follicles are subject to marked inflammation and degenerative changes. However, as a manifestation of early disease, premature inner root sheath desquamation is unique to CCSA. This anatomical defect is probably an important component in the pathogenesis of the disease and may predispose the affected follicles to injury.

Figure 18.5 One of four follicles, upper right, has undergone premature desquamation of the inner root sheath. Original magnification ×100
As the disease evolves in clinically abnormal scalp skin, involved follicles also demonstrate some or all of the following histological features: eccentric epithelial atrophy (thinning) with hair shafts in close proximity to the dermis (Figures 18.8 and 18.9); concentric lamellar fibroplasia (onion skin-like fibrosis) of affected follicles (Figures 18.10 and 18.11); variably dense lymphocytic perifollicular inflammation, primarily at the level of the upper isthmus and lower infundibulum (Figures 18.12–18.14); occasional fusion of infundibula (polytrichia; Figures 18.15 and 18.16); and in advanced lesions, total destruction of the follicular epithelium with retained hair shaft fragments and granulomatous inflammation. Inter-face alteration of the follicular epithelium is not found. To date, the immunofluorescent findings in patients with CCSA have not been critically studied.

When highly inflammatory disease with pustule formation is present (‘folliculitis decalvans’), the microscopic findings in pustular lesions include intrafollicular and perifollicular infiltrates rich in neutrophils as well as lymphocytes (Figures 18.17–18.21).

However, if non-pustular lesions at the active periphery are sampled, or if the biopsy is performed during a period of suppressive therapy, the histological
picture is identical to that of other forms of CCSA (Figure 18.22). If early lesions are sampled, all the histological changes described for CCSA can be found in ‘folliculitis decalvans’.

The only unique histological characteristic of CCSA is premature desquamation of the inner root sheath. The remainder of the histological features can be found in other forms of inflammatory, scarring alopecia, and are therefore suggestive, but not diagnostic.

**SUMMARY**

*Clinical correlation:* an adult, most often a Black woman, with a progressive, permanent loss of scalp hair starting on the central crown or vertex. A spectrum of clinical inflammation exists, ranging from minimal to highly inflamed, with marked erythema, pustules and crusting (‘folliculitis decalvans’).

*Histological findings:* the earliest finding is premature desquamation of the inner root sheath. This may be found even when normal-appearing scalp skin or perilesional skin is sampled. In more advanced lesions, some or all of the following features are usually seen:
• Variably dense lymphocytic perifollicular inflammation, primarily at the level of the upper isthmus and lower infundibulum
• Eccentric epithelial atrophy (thinning), with hair shafts in close proximity to the dermis
• Occasional fusion of infundibula (polytrichia)
• Concentric lamellar fibroplasia (onion skin-like fibrosis) of affected follicles
• In advanced lesions, total destruction of the follicular epithelium with retained hair shaft fragments and granulomatosus inflammation
• Replacement of the follicular epithelium by connective tissue (follicular scars)

**BIBLIOGRAPHY**

Figure 18.9 Two abnormal follicles in different stages of evolution; one follicle (lower left) shows eccentric epithelial atrophy and the second (lower right) shows total epithelial destruction with a retained hair shaft. A normal follicle (upper-most) is present for comparison. Original magnification ×200


Figure 18.10 Vertical section of a follicle showing concentric lamellar fibroplasia (as well as epithelial atrophy) at the level of the upper isthmus/lower infundibulum. Original magnification ×200
Figure 18.11 Transverse section of a follicle similar to that in Figure 18.10. Original magnification ×200

Figure 18.12 Perifollicular lymphocytic inflammation is usually found in association with epithelial thinning and lamellar fibroplasia, and is most intense at the level of the isthmus and lower infundibulum. Original magnification ×200
Figure 18.13 Another example of perifollicular lymphocytic inflammation. Original magnification ×200

Figure 18.14 One of the two follicles (left side) has an intact inner root sheath and is not affected by inflammation or fibroplasia. Original magnification ×200

Figure 18.15 The infundibula of several adjacent, inflamed follicles may merge, resulting in clinical polytrichia. Original magnification ×200
Figure 18.16 During the healing process a single, common enlarged infundibulum may form, resulting in polytrichia. Original magnification ×200
Figure 18.17 Biopsy specimen taken from a crusted, pustular lesion in a patient with the ‘folliculitis decalvans’ variant of central, centrifugal scarring alopecia. Much of the follicular epithelium has been damaged or destroyed by a dense inflammatory infiltrate composed predominantly of neutrophils. (See Figures 18.18–18.20.) Original magnification ×40
Figure 18.18 Higher power view of the section shown in Figure 18.17. Original magnification ×200
Figure 18.19 Special stain for bacteria reveals colonies of staphylococci. Original magnification ×400
Figure 18.20 Special stain for fungi reveals clusters of fungal spores typical of *Pityrosporum* contained within the infundibulum and overlying scale/crust. The findings in Figures 18.17–18.21 are non-specific ‘end stage’ changes, and can be found in any highly inflammatory form of scarring alopecia. Original magnification ×400
Figure 18.21 Pustular lesions in highly inflammatory cases of central, centrifugal scarring alopecia are rich in neutrophils and resemble a bacterial folliculitis. Original magnification ×100
CHAPTER 19
Lichen planopilaris

The clinical course of lichen planopilaris (LPP) may be insidious or fulminant, and the pattern of scalp hair loss is highly variable. Most commonly, there are several scattered foci of partial hair loss (Figures 19.1 and 19.2). Perifollicular erythema and scaling are almost always present (Figure 19.3). A pattern of hair loss suggestive of central, centrifugal scarring alopecia (Figure 19.4) or pseudopelade of Brocq can also occur. As in all cases of inflammatory scarring alopecia, LPP can heal with the formation of polytrichia (tufting; Figure 19.5). Clearly, there can be some clinical overlap with other forms of scarring alopecia. Therefore, the diagnosis of LLP cannot be based on clinical features alone.

Patients with indolent scalp disease may be asymptomatic, but itching and tenderness are often present. Patients who also have lesions of lichen planus on the skin or mucosa may have symptoms related to these lesions.

The Graham-Little syndrome (scarring alopecia of the scalp, loss of pubic and axillary hair, and the rapid development of keratosis pilaris) is considered by some to be a variant of LPP. However, typical lichen planus lesions are not found and the histological findings are usually not lichenoid. It is unclear whether this condition is related to LPP. In any case, the Graham-Little syndrome is rare.

HISTOLOGICAL FEATURES

Typical LPP shows a band-like mononuclear cell infiltrate obscuring the interface between follicular epithelium and dermis (Figure 19.6). Vacuolar alteration at the interface and wedge-shaped hypergranulosis within affected infundibula is typical. The epithelial-adventitial junction often shows prominent dyskeratosis with individually necrotic, polygonal basal keratinocytes (Figure 19.7). Colloid or Civatte bodies are occasionally found as part of the interface alteration, but this occurs less commonly in disease of the follicular epithelium than of the epidermis.

Inflammation affects the upper portion of the follicle (infundibulum and isthmus) most severely (Figures 19.8–19.10), but inflammation may occasionally extend down the length of the follicle. Perivascular and pericrinal lymphocytic infiltrates of the mid- and deep dermis (as seen in discoid lupus erythematosus) are absent in LPP. Occasionally, interfollicular changes of lichen planus are found
Figures 19.11–19.13. When present, interfollicular involvement strongly supports a diagnosis of LPP.

Eventually the infundibulum and isthmus become distended and plugged with keratinous debris. With time, vacuolar interface change subsides, the layer of perifollicular fibrosis becomes thicker, and the lymphocytic infiltrate seems to ‘back away’ from the follicle (Figures 19.14 and 19.15). Basilar keratinocytes

Figure 19.1 Most commonly, the scalp lesions of lichen planopilaris are irregularly shaped and widely scattered over the scalp

Figure 19.2 A ‘confetti-like’ pattern of numerous small and widely distributed lesions is a relatively common clinical variant of lichen planopilaris

Figure 19.3 Before the hair is completely destroyed, perifollicular erythema and scaling indicate the presence of active lichen planopilaris

(Figures 19.11–19.13). When present, interfollicular involvement strongly supports a diagnosis of LPP.

Eventually the infundibulum and isthmus become distended and plugged with keratinous debris. With time, vacuolar interface change subsides, the layer of perifollicular fibrosis becomes thicker, and the lymphocytic infiltrate seems to ‘back away’ from the follicle (Figures 19.14 and 19.15). Basilar keratinocytes
become pink and flattened, and an artifactual cleft between epithelium and stroma is often found. In fact, the epithelium may appear to be ‘floating’ within the cleft (Figure 19.16). Although these ‘later’ changes are often seen in patients with long-standing LPP, they are not pathognomonic and can be observed in other forms of inflammatory, scarring alopecia.

In time, the follicle is entirely destroyed. At first, retained hair shafts fragment and a granulomatous response is observed, but eventually the follicles are replaced by columns of connective tissue, giving the appearance of a ‘burnt out’ scarring alopecia (Figure 19.17; see also Chapter 22). As is the case in all forms of inflammatory, scarring alopecia, the healing process may result in clinical and histological polytrichia (Figure 19.18).

Grouped globular immunofluorescence (usually IgM), especially when found adjacent to the follicular epithelium, is the characteristic pattern seen in LPP (Figure 19.19). Linear deposits of immunoreactants are typical of lupus erythematosus. This distinction can be important, because the scarring alopecia of LPP and discoid lupus erythematosus may resemble each other both clinically and histologically.

Figure 19.4 This patient with lichen planopilaris has extensive hair loss centered on the crown of the scalp, resembling a severe case of central, centrifugal scarring alopecia.
Because of poor site selection or sampling error, often the only changes found are perifollicular fibroplasia and chronic inflammation. These changes are non-specific and cannot be considered diagnostic. Additional biopsies are required to establish the diagnosis whenever non-specific changes are found. However, typical clinical lesions of lichen planus on other parts of the body strongly support the diagnosis of LPP.

**SUMMARY**

*Clinical correlation:* various patterns of hair loss are seen, but most commonly there are scattered foci of partial hair loss. Perifollicular erythema and scaling are common to all cases. Lichen planus lesions elsewhere on the body support the diagnosis of LPP on the scalp.

*Histological findings:*

- Interface lichenoid dermatitis affecting the infundibulum and isthmus
- The presence of ‘squamatized’ basal layer with artifactual clefting between epithelium and dermis is often seen

![Figure 19.5](image.png) In this patient, lesions of lichen planopilaris have resulted in polytrichia.
Colloid bodies and interfollicular changes of lichen planus are sometimes found and support the diagnosis. Perifollicular chronic inflammation and fibroplasia are sometimes the only changes seen, but cannot be considered diagnostic. Grouped globular immunofluorescence (usually IgM), especially when found adjacent to the follicular epithelium, is the characteristic pattern seen in lichen planopilaris.

**BIBLIOGRAPHY**

Amato L, Mei S, Massi D, Gallerani I, Fabbri R Cicatricial alopecia: a dermatopathologic and immunopathologic study of 33 patients (pseudopelade of Brocq is not a specific clinicopathologic entity). *Int J Dermatol* 2002; 41:8–15


**Figure 19.8** Transverse sections (see also Figures 19.9 and 19.10) taken at multiple levels clearly demonstrate the depth of inflammation in lichen planopilaris. In this specimen, inflammation is most intense at the level of the upper isthmus. Original magnification ×40

**Figure 19.9** Inflammation is relatively sparse at the infundibular level. Original magnification ×40

**Figure 19.10** Transverse section at the suprabulbar level, showing relatively sparse inflammation. This is a typical pattern for lichen planopilaris. Original magnification ×40

Figure 19.11 This specimen from a patient with lichen planopilaris had typical follicular changes but also interfollicular epidermal involvement. Vacuolar interface alteration, numerous colloid bodies, and focal hypergranulosis can be seen. Original magnification x200

Figure 19.12 Another view of the specimen shown in Figure 19.11. Original magnification x400

Silvers DN, Katz BE, Young AW. Pseudopelade of Brocq is lichen planopilaris: report of four cases that support this nosology. Cutis 1993; 51:99–105
Figure 19.13 Another view of the specimen shown in Figures 19.11 and 19.12. Original magnification ×400

Figure 19.14 ‘Later’ stage histological findings in lichen planopilaris. Vacuolar interface change is less evident, the layer of perifollicular fibrosis is quite thick, and the lymphocytic infiltrate has moved outside the zone of fibrosis. Original magnification ×200
Figure 19.15 ‘Later’ stage lichen planopilaris, as in Figure 19.14. Original magnification x400

Figure 19.16 The more advanced changes seen in lichen planopilaris include prominent artifactual clefting between epithelium and perifollicular connective tissue. Original magnification x400
Figure 19.17 ‘End-stage’ lichen planopilaris. The epithelium has been completely destroyed. After the residual hair shaft has been resorbed, inflammation will subside and a column of connective tissue will remain. Original magnification ×400
Figure 19.18 In this ‘end-stage’ specimen of lichen planopilaris, the follicular infundibula of several adjacent follicles have merged (polytrichia) to form a single, enlarged ostium. Original magnification ×200
Figure 19.19 Immunofluorescent staining for IgG in a specimen from a patient with lichen planopilaris. Globular, clustered colloid bodies concentrated at the lower half of the infundibulum are highlighted bright green. Linear staining of the epithelium is absent. Slide courtesy of Kathleen David-Bajar, MD
CHAPTER 20
Frontal fibrosing alopecia

This condition was recently reported in several Australian, postmenopausal, Caucasian women. The disease has yet to be reported in the USA. Patients are elderly women (mean age 67 years) with progressive hair loss along the anterior hairline and the eyebrows. The histological features are similar to those found in lichen planopilaris. Frontal fibrosing alopecia appears to be just one of several clinical patterns of hair loss that can be seen in lichen planopilaris. However, lesions of lichen planus are not found in these patients and the lichenoid inflammation does not affect the interfollicular epidermis.

BIBLIOGRAPHY

CHAPTER 21
Chronic cutaneous lupus erythematosus
(discoid lupus erythematosus)

The term ‘chronic cutaneous lupus erythematosus’ (CCLE) is preferred to ‘discoid lupus erythematosus’ (DLE) because it is more inclusive. Nevertheless, DLE enjoys such common usage that the two terms are used interchangeably. CCLE usually occurs in adults, and is more common in women. Although lesions of CCLE are often found in patients with systemic lupus erythematosus (SLE), the majority of patients with CCLE do not have systemic disease. Among patients with skin disease only, about 50% will have scalp lesions, and very few of these patients will ever progress to SLE. Although itching or tenderness is common, the condition may be asymptomatic. The presence of lesions outside the scalp greatly simplifies diagnosis.

Scalp involvement may resemble classic DLE lesions, with alopecia, erythema, epidermal atrophy and dilated, plugged follicular ostia. Central hypopigmentation and peripheral hyperpigmentation are characteristic of lesions in dark-skinned individuals (Figure 21.1). Plaques of CCLE may merge to form large, irregularly shaped zones of scarring alopecia, and in some cases may be centered on the crown or vertex, resembling central, centrifugal scarring alopecia (Figure 21.2). However, some cases of CCLE first present as clinically ‘non-scarring’ or even ‘non-inflammatory’ alopecia that may resemble alopecia areata (Figure 21.3). Long-standing, ‘burnt out’ disease confined to the scalp may be impossible to differentiate from lichen planopilaris or Brocq’s pseudopelade (Figure 21.4). Because CCLE of the scalp can have many clinical patterns, the diagnosis cannot be based on clinical findings alone.

HISTOLOGICAL FINDINGS
Vacuolar interface alteration of the epidermis and follicular epithelium is typical, although the epidermis may be spared in scalp lesions (Figures 21.5–21.9). The degree of perifollicular inflammation associated with the vacuolar interface change is highly variable. There can be dramatic vacuolar alteration with almost no inflammation, or there can be a dense, band-like lymphocytic infiltrate suggestive of lichen planopilaris (LPP). Dyskeratosis and colloid (Civatte) bodies are occasionally seen, but less commonly than in LPP. However, moderate to dense, superficial and deep chronic inflammation, often including
plasma cells, is seen in both perivascular and periadnexal locations (Figure 21.10). Perifollicular inflammation is often most severe at the level of the infundibulum but can extend down to the follicular bulbs (Figure 21.11), and inflammatory cells may invade the follicular epithelium. Residual inflammation may be found in and around the follicular tracts below follicles that have entered the telogen phase or have been destroyed (Figures 21.12 and 21.13). Increased dermal mucin is often present (Figure 21.14) and is helpful in differentiating CCLE from LPP. Additional findings that support a diagnosis of CCLE over LPP include the presence of a focally thinned but hyperkeratotic epidermis, a thickened basement membrane, the absence of wedge-shaped hypergranulosis and an infiltrate containing numerous plasma cells (Figure 21.15).

Although artifactual clefting between the follicular epithelium and the stroma is typical of LPP, it can also be seen in CCLE (Figure 21.16). Therefore, this histological feature cannot be used to separate the two entities. Granular deposits of IgG and C3 (less commonly IgM or IgA) at the dermoepidermal junction and/or the junction of the follicular epithelium and dermis are typical of CCLE.
Globular deposits of IgM representing colloid bodies may be present, but not as commonly as in LPP.

**SUMMARY**

*Clinical correlation:* The diagnosis of CCLE requires histological confirmation and cannot be based solely on the clinical appearance of scalp lesions. These may resemble classic DLE lesions, with alopecia, erythema, epidermal atrophy and dilated, plugged follicular ostia. Central hypopigmentation and peripheral hyperpigmentation are commonly seen in dark-skinned individuals. The distribution and degree of clinical inflammation varies among patients, and plaques of CCLE may result in patterns resembling alopecia areata, LPP, central, centrifugal scarring alopecia and Brocq’s pseudopelade.

*Histological findings:*
Vacuolar interface alteration of the epidermis and follicular epithelium is typical, although the epidermis may be spared in lesions of DLE involving the scalp. The interface change is usually vacuolar rather than lichenoid. Dyskeratosis and colloid (Civatte) bodies are occasionally seen, but less commonly than in LPP. Moderate to dense chronic inflammation, often including plasma cells, is seen in both perivascular and periadnexal locations. When perifollicular inflammation is noted, it is usually most severe at the level of the infundibulum, and inflammatory cells may invade the follicular epithelium. Similar inflammation may be found in and around the follicular tracts that lie below follicles that are in the telogen phase or have been destroyed. Increased dermal mucin is often present and is helpful in differentiating DLE from LPP. Granular deposits of IgG and C3 (less commonly IgM or IgA) at the dermoepidermal junction and/or the junction of the follicular epithelium and
dermis are typical of CCLE. Globular deposits of IgM representing colloid bodies may be present, but not as commonly as in LPP.

**BIBLIOGRAPHY**


**Figure 21.7** Chronic cutaneous lupus erythematosus, with moderate inflammation. Original magnification ×400
**Figure 21.8** Chronic cutaneous lupus erythematosus, with dense inflammation. Original magnification ×200

Figure 21.9 Chronic cutaneous lupus erythematosus, with dense inflammation, as in Figure 21.8. Original magnification ×400

Figure 21.10 Lymphoplasmacytic inflammation often surrounds eccrine glands as well as follicles. Original magnification ×400
Figure 21.11 Inflammation may extend down to the follicular bulbs. Original magnification ×40

Figure 21.12 Follicular stela infiltrated by copious plasma cells and lymphocytes. Original magnification ×400
Figure 21.13 Follicular stela infiltrated by copious plasma cells and lymphocytes. Original magnification ×400
Figure 21.14 An increase in interfollicular dermal mucin (here stained blue) is commonly found in chronic cutaneous lupus erythematosus. Colloidal iron stain, original magnification ×400
Figure 21.15 Epidermal involvement in a scalp biopsy from a patient with chronic cutaneous lupus erythematosus (CCLE). Hyperkeratosis with hypogranulosis and epidermal thinning help to differentiate CCLE from lichen planopilaris. Original magnification ×200

Figure 21.16 Artifactual clefting between the follicular epithelium and the stroma can be seen in this example of chronic cutaneous lupus erythematosus. Original magnification ×200
Figure 21.17 Immunofluorescent staining for IgM in a patient with chronic cutaneous lupus erythematosus. A bright, green, granular band of staining highlights the epidermal and follicular basement membrane zones. Slide courtesy of Kathleen David-Bajar, MD
Figure 21.18 Gray-scale rendition of Figure 21.17 identifies the epidermal basement membrane zone (EP B Z) and the follicular basement membrane zone (F B Z)
CHAPTER 22

Brocq’s alopecia (pseudopelade of Brocq) and ‘burnt out’ scarring alopecia

BROCQ’S ALOPECIA

The term ‘pseudopelade of Brocq’ is a source of much confusion and fruitless debate, and should be abandoned. Pseudopelade has been used in recent decades to describe some patients with central, centrifugal scarring alopecia (CCSA), a very different condition from that described by Dr Brocq. To avoid confusion, we can refer to the entity described by Brocq as Brocq’s alopecia. Brocq’s alopecia is not a distinct disease but a clinical pattern of scarring alopecia. It is an end-stage or clinical variant of several other forms of scarring alopecia and a diagnosis of exclusion. The same pattern of hair loss can be seen in ‘burnt out’ lichen planopilaris (LPP), discoid lupus erythematosus (DLE) and other forms of cicatricial hair loss. If a definitive diagnosis of DLE, LPP, CCSA or another form of scarring alopecia can be made based on clinical, histological or immunofluorescent features, then the term Brocq’s alopecia cannot be used. If a ‘primary’ form of Brocq’s alopecia exists, it has yet to be convincingly described.

The Brocq’s alopecia pattern of hair loss is very uncommon. The typical patient is a Caucasian adult who is surprised to discover discrete, asymptomatic areas of scalp hair loss. In some patients, the disease is slowly progressive, and new areas of alopecia develop over a period of months to years. However, the condition often worsens in ‘spurts’, with periods of activity followed by ‘dormant’ periods. This is distinctly different from the slow but steady disease progression seen in forms of CCSA described in Chapter 18. Disease progression in Brocq’s alopecia eventually terminates spontaneously.

Unlike CCSA, Brocq’s alopecia results in irregularly shaped and often widely distributed and grouped bald patches on the scalp. Cases with exclusive crown or vertex involvement may actually represent examples of ‘burnt out’ CCSA.

The individual lesion is hypopigmented (‘porcelain white’ is the classic description) and slightly depressed (atrophic). Lesions are often irregularly shaped, as opposed to the round or oval patches usually seen in alopecia areata and most cases of CCSA (Figure 22.1). The classic description of ‘footprints in the snow’ refers to dermal atrophy causing a slight depression below the surrounding normal scalp. In fact, many cases of Brocq’s alopecia do not
demonstrate atrophy. Usually only mild erythema and slight perifollicular scaling are present, and often there is no clinical evidence of inflammation. In fact, some authors have argued that any inflammation excludes Brocq’s alopecia from the clinical differential diagnosis. Just as in other forms of scarring alopecia, a few isolated hairs may remain within an otherwise smooth, shiny, denuded patch.

The histological findings of Brocq’s alopecia have yet to be clearly defined. The criteria established by Pinkus in 1978 are not correlated in any way with clinical features. Thus, ‘pseudopelade’ as described by Pinkus is a histological and not a clinical entity. In most cases of Brocq’s alopecia, the ‘active’ lesion is elusive, and the typical histological findings are those of a ‘burnt out’ scarring alopecia (see below).

The histological findings of ‘pseudopelade’ more recently described apply to a subset of CCSA rather than Brocq’s alopecia. It would not be surprising to find an occasional case of Brocq’s alopecia demonstrating the typical histological findings of CCSA. Brocq’s alopecia is, after all, the end stage of several different forms of scarring alopecia. A prospective study of Brocq’s alopecia with sound clinical correlation has yet to be performed.

‘BURNT OUT’ SCARRING ALOPECIA

All too often, pathologists must render the diagnosis of a ‘burnt out’ or ‘end stage’ scarring alopecia. The histological term burnt out scarring alopecia, like the clinical term Brocq’s alopecia, does not indicate a specific disease but a pattern common to several entities. There are two possible reasons for finding this pattern. First, the patient’s disease may, in fact, have truly ‘burnt out’ like an old forest fire, with no further follicular destruction. More commonly, however, the ‘end stage’ pattern is the result of clinicians’ unfortunate choice of biopsy sites. Frequently, bald or nearly bald areas are sampled, instead of spots where numerous hairs are still present. The advancing border of a zone of scarring alopecia is always a more productive site than a bald zone. Areas with subtle
findings such as perifollicular erythema or scaling prove more fruitful than places showing the ‘advanced’ findings of pustules, papules, extensive hair loss or obliteration of follicular ostia. Just a few millimeters may separate a ‘perfect’ from a suboptimal biopsy site, but even experienced clinicians cannot see below the surface of the skin. In patients with progressive hair loss, the diagnosis of ‘burnt out scarring alopecia’ should serve as an invitation for additional biopsy specimens.

An ‘end stage’ or ‘burnt out’ scarring alopecia is characterized by: a decreased total number of hairs, especially terminal hairs (Figure 22.2); loss of the sebaceous glands; residual, ‘naked’ hair shafts surrounded by mild, granulomatous inflammation (Figure 22.3); follicular stelae without overlying follicles; and cylindrical columns of connective tissue representing the sites of former follicles (Figures 22.4–22.6).

Dense, superficial, perifollicular inflammation, with destruction of the follicular epithelium (Figure 22.7; see also Figures 18.17–18.20), may be interpreted as a non-specific, ‘late’ finding heralding impending ‘burn out’ at the
biopsy site. However, this ‘end stage’ histological finding indicates that the patient has active, progressive disease.

**Figure 22.4** Transverse section of columns of connective tissue that mark the sites of former follicles (arrowhead). The absence of associated inflammation indicates that the destructive process has truly ‘burnt out’, at least at the site chosen for biopsy. Original magnification ×100
SUMMARY

Brocq’s alopecia

Clinical correlation: usually in a Caucasian adult, who discovers discrete, asymptomatic, hypopigmented, slightly depressed (atrophic) and irregularly shaped bald patches affecting any portion of the scalp.

Histological findings: Brocq’s alopecia represents the end stage of various forms of scarring alopecia and usually shows features of a ‘burnt out’ scarring alopecia (see below). If typical histological findings of LPP, CCSA or other types of scarring alopecia are found, the diagnosis of Brocq’s alopecia can be excluded.
`Burnt out’ scarring alopecia

Clinical correlation: most often indicative of bald or nearly bald plaques in patients with various forms of scarring alopecia. Biopsy site sampling error (and sometimes disease resolution) is the most common explanation.

Histological findings:

- Decreased total number of hairs, especially terminal hairs
- Loss of the sebaceous glands
- Residual, ‘naked’ hair shafts surrounded by mild, granulomatous inflammation
- Follicular stelae without overlying follicles
- Cylindrical columns of connective tissue at the sites of former follicles

Figure 22.6 Higher power view of the follicular scar shown in Figure 22.5. Original magnification ×200
Figure 22.7 Dense, acute and chronic inflammation surrounds a residual hair shaft (lost during processing). This histological pattern often corresponds to the clinical finding of pustules or crusted papules. These are ‘late’ and non-specific findings. Original magnification x200

BIBLIOGRAPHY

Amato L, Mei S, Massi D, Gallerani I, Fabbri P. Cicatricial alopecia; a dermatopathologic and immunopathologic study of 33 patients (pseudopelade of Brocq is not a specific clinicopathologic entity). Int J Dermatol 2002; 41:8–15


CHAPTER 23
Acne keloidalis (folliculitis keloidalis)

Often called *acne keloidalis nuchae*, this disorder typically affects young Black men and occasionally young Black women. Acne keloidalis (AK) is at least ten times more common in Blacks than in Caucasians and comprises nearly 0.5% of all dermatological cases in the Black population. However, the condition can occur in Caucasian men and on rare occasions in Caucasian women. It begins as small, smooth, firm papules with occasional pustules on the occipital scalp and posterior neck. In a minority of cases, lesions are more numerous on the vertex and crown, and so it is best to use the term *acne keloidalis* without the modifier *nuchae*. Initially hairs can be seen exiting the papules, but the hair shafts are soon shed. With time, the papules resolve and leave small zones of alopecia within a field of papular lesions. In many patients, the papules coalesce and form firm, hairless, keloid-like protuberant plaques (hence the term *keloidalis*) that can be painful and cosmetically disfiguring. Abscesses and sinuses exuding pus can be present in advanced cases. Although AK may be asymptomatic, mild symptoms of burning and itching are often present.

The cause of AK remains unclear. The notion that lesions of AK are caused by ‘ingrowing’ hairs, analogous to the situation in pseudofolliculitis barbae, has been disproved. AK is frequently found in association with central, centrifugal scarring alopecia (CCSA), suggesting a common or related pathogenesis (Figures 23.1–23.4). Occasionally, the papular lesions and hair loss of AK extend up onto the vertex of the scalp, producing a clinical ‘overlap’ of AK and CCSA.

**HISTOLOGICAL FINDINGS**

Most descriptions of the pathology of AK are based on late-stage, nodular lesions. When early, papular lesions or samples of perilesional skin are studied, the findings are those of a chronic, lymphocytic folliculitis. AK is a primary form of scarring alopecia, and many of the histological findings closely resemble those found in certain other forms of cicatricial alopecia. The most common findings, in order of frequency found, are: perifollicular, chronic inflammation (lymphocytic and plasmacytic), most intense at the level of the isthmus and lower infundibulum (Figures 23.5 and 23.6); lamellar fibroplasia, most marked at the level of the isthmus; complete disappearance of sebaceous glands
associated with inflamed or destroyed follicles; thinning of the follicular epithelium, especially at the level of the isthmus; polytrichia (several hairs sharing a common, fused infundibulum; Figure 23.7); and total epithelial destruction (superficial and deep) with residual ‘naked’ hair fragments (Figures 23.8–23.12). Some of these retained shafts are slowly resorbed, resulting in a

Figure 23.1 Typical acne keloidalis lesions on the nuchal region of a woman with typical central, centrifugal scarring alopecia on the crown

Figure 23.2 Central, centrifugal scarring alopecia on the crown of the woman in Figure 23.1

Figure 23.3 Acne keloidalis lesions on the nuchal region of a man with central, centrifugal scarring alopecia on the vertex/crown
focal bald spot. Others, however, can serve as a nidus for marked fibrosis, persistent inflammation and bacterial superinfection. The resulting tissue hypertrophy traps adjacent follicles, and eventually leads to the ‘keloidal’, hypertrophic scarring seen in some patients with AK.

Even some specimens from ‘normal-appearing’ perilesional skin may contain true follicular scars, demonstrating subclinical disease. Special stains for bacteria demonstrate relatively few organisms, suggesting that bacterial overgrowth is not important in the pathogenesis of the disease.

Many of the histological features of AK are shared by CCSA. This overlap of histological features suggests a common pathogenesis for AK and CCSA, a notion that is supported by the frequent occurrence of both conditions in the same patient. As mentioned above, clinical features sometimes overlap as well.

**SUMMARY**

*Clinical correlation:* small, smooth, firm follicular papules with occasional pustules, eventually leading to partial hair loss; most common on the nuchal
region (occipital scalp and posterior neck). With advanced disease, there are coalescent papules forming firm, hairless, keloid-like protuberant plaques.

**Histological findings:**

- Perifollicular, chronic inflammation (lymphocytic and plasmacytic)
- Perifollicular, lamellar fibroplasia
- Complete disappearance of sebaceous glands associated with inflamed or destroyed follicles
- Thinning of the follicular epithelium, especially at the level of the isthmus
- Eventually, total epithelial destruction, with residual ‘naked’ hair fragments

**BIBLIOGRAPHY**


Sperling L, Homoky C, Pratt L, Sau P. Acne keloidalis is a form of primary scarring alopecia. *Arch Dermatol* 2000; 136: 479–84

Figure 23.6 Higher power view of the section shown in Figure 23.5. Original magnification ×200
Figure 23.7 In a specimen taken from a small papular lesion of acne keloidalis nuchae, two follicles (sharing a fused infundibulum) are surrounded by dense, lymphocytic inflammation. Original magnification ×200
Figure 23.8 Specimen taken from a papular lesion of acne keloidalis nuchae shows dense perifollicular chronic inflammation, most intense at the level of the lower isthmus. The epithelium is disrupted at this level, and such follicles are doomed to destruction. Original magnification x40
Figure 23.9 Higher power view of the specimen in Figure 23.8. Original magnification ×100
Figure 23.10 When the epithelium of follicles has been disrupted or destroyed, the inflammatory infiltrate is often rich in plasma cells as well as lymphocytes. Original magnification ×400

Figure 23.11 Two follicles in a papular lesion of acne keloidalis nuchae, sectioned at the level of the upper isthmus. The epithelium of one follicle has been completely destroyed below the level of the isthmus, which would result in permanent hair loss. Original magnification ×200
Figure 23.12 Three hairs in a papular lesion of acne keloidalis nuchae, sectioned at the level of the lower isthmus. Original magnification ×200
CHAPTER 24
Dissecting cellulitis of the scalp
(perifolliculitis capitis abscedens et suffodiens)

Dissecting cellulitis is an uncommon but distinctive disease. It most commonly affects young adult men, especially Black men. Dissecting cellulitis is part of the ‘follicular occlusion triad’ that includes hidradenitis suppuritiva and acne conglobata, but scalp disease often occurs alone.

Lesions begin as multiple, firm scalp nodules, most commonly on the crown, vertex and upper occiput. The nodules rapidly develop into boggy, fluctuant, oval and linear ridges that eventually discharge purulent material (Figure 24.1). Lesions often interconnect, so that pressure on one fluctuant area may result in a purulent discharge from perforations several centimeters away (Figure 24.2). Despite massive, deep inflammation, there can be surprisingly little pain, and patients often seek help because of hair loss and a foul-smelling discharge.

The disease waxes and wanes for years, but eventually leads to dense dermal fibrosis, sinus tract formation, permanent alopecia and hypertrophic scarring. Squamous cell carcinoma has rarely been reported to arise in the setting of long-standing disease.

HISTOLOGICAL FINDINGS
The very earliest findings have seldom been described, probably because patients first consult physicians after the disease is fairly well developed. Also, clinicians are more likely to take biopsy specimens from nodular or cystic lesions that represent more advanced, long-standing disease. In the author’s experience, the earliest change is moderately dense, perifollicular lymphocytic inflammation affecting the lower half of the dermis and extending down into the fat (Figures 24.3 and 24.4). At this stage, numerous intact and seemingly undamaged follicles are bathed in a sea of acute and chronic inflammation. Terminal anagen hairs are reduced in number, because many have converted to the catagen/telogen phase (Figure 24.5). Initially, lymphocytes dominate the infiltrate, but eventually neutrophils become more numerous, especially in long-standing, fluctuant lesions. Still later, plasma cells increase in number. Vascular proliferation accompanies the infiltrate as the disease evolves, and the deep dermis and subcutaneous fat begin to exhibit the histological findings of granulation tissue (Figures 24.6 and 24.7).
In most other forms of inflammatory, scarring alopecia, the sebaceous glands are often the first structure to disappear. However, sebaceous glands remain intact well into the course of dissecting cellulitis. This relative sparing of sebaceous glands may be related to the depth of the infiltrate in the disease (Figures 24.8 and 24.9).

The intense inflammation is associated with the conversion of anagen hairs into catagen/telogen hairs, whose shafts are ultimately shed. Therefore, the hair loss seen in the early stages of dissecting cellulitis is temporary. However, with
the passage of time, inflammation extends throughout the dermis and invades the follicular epithelium, and there is subsequent epithelial injury and follicular destruction. Repetitive injury to the follicles results in acniform dilatation of the infundibula, with perifollicular neutrophilic inflammation (Figure 24.10).

If fluctuant nodules and sinuses are sampled, large perifollicular and mid- to deep dermal abscesses composed of neutrophils, lymphocytes and often numerous plasma cells are seen. Eventually, chronic abscesses become lined with squamous epithelium derived from the overlying epidermis, and true sinus tracts are formed. As follicles are completely destroyed, inflammation subsides and is replaced by dense fibrosis of the dermis and the superficial fat.

**SUMMARY**

*Clinical correlation:* young adults, usually male and often African American, with boggy nodules and a purulent discharge on the scalp. Lesions are scattered, but most are concentrated on the crown, vertex and upper occiput.
**Histological findings:**

- Very early lesions are seldom evaluated, but may show moderately dense, lymphocytic, perifollicular inflammation surrounding the lower half of the follicle.

**Figure 24.4** Another section showing ‘early’ lesions of dissecting cellulitis (see Figure 24.3). Original magnification ×40

**Figure 24.5** A terminal catagen hair bulb is surrounded, but not invaded, by a dense lymphocytic infiltrate. Original magnification ×400

*Histological findings:*
• Increase in catagen/telogen hairs
• Sebaceous glands may persist well into the course of the disease
• Fully developed, fluctuant lesions show perifollicular and mid- to deep dermal abscesses composed of neutrophils, lymphocytes and often copious plasma cells
• Eventually, granulation tissue, epithelium-lined sinus tracts and fibrosis are seen

BIBLIOGRAPHY

Figure 24.8 Despite the loss of follicles and the severity of deep inflammation, sebaceous glands persist well into the course of the disease. Original magnification ×40
Figure 24.9 Dissecting cellulitis. Sebaceous glands are spared. Original magnification ×100
Figure 24.10 Acneiform dilatation of the infundibulum. Original magnification ×100
The ‘tufting’ seen in this pattern of hair disease is common to several forms of scarring alopecia (Figure 25.1). It is not a specific disease, but an end stage of several different conditions. In spite of this, ‘tufted folliculitis’ continues to be applied as a diagnostic term in the medical literature. Such usage is incorrect and misleading.

Tufting occurs because the infundibular epithelium of damaged follicles often heals with the formation of a large, common infundibulum (Figure 25.2). A zone of fibrosis separates individual tufts. Another name for this phenomenon is polytrichia. Although tufting is fairly common in cases of central, centrifugal scarring alopecia, it can occasionally be seen in a wide variety of other disorders, including folliculitis keloidalis, dissecting cellulitis and inflammatory tinea capitis.

**Figure 25.1** Dramatic example of polytrichia (‘tufting’) in a patient with central, centrifugal scarring alopecia

**BIBLIOGRAPHY**


Figure 25.2 A histological view of polytrichia (‘tufting’). Whenever the upper halves of adjacent follicles become inflamed and damaged, a large, common infundibulum may form. Original magnification ×100


CHAPTER 26
Tinea capitis

Most cases of tinea capitis (fungal infection of scalp hair) seen in North America are caused by *Trichophyton tonsurans*, a large-spore endothrix infection. *Microsporum canis* (ectothrix) is also found occasionally. Different species may be more prevalent in other parts of the world. The fungus invades and multiplies within the hair shaft below the surface of the skin, causing hair fragility and breakage. This results in bald spots with follicular ostia filled with keratinous debris or ‘black dots’, the residua of infected, pigmented hair shafts (Figure 26.1). Variable degrees of inflammation may be seen, ranging from none (no erythema or scaling) to highly inflamed, purulent, edematous and crusted plaques (kerion; Figure 26.2). Pustular lesions on the scalp should always raise suspicion for tinea capitis. Tinea capitis almost always affects children, and African American children seem to be especially prone to infection. Any inflamed or scaly bald spot on the scalp of an African American child should be considered to be tinea capitis unless proven otherwise. However, adults of all races (but especially African American women) can also be affected. If treated early, tinea capitis heals without scarring or hair loss. However, highly inflammatory lesions or untreated lesions may eventuate in permanent hair loss. Inflammatory tinea capitis is one of several causes of ‘tufted folliculitis’ (polytrichia).

HISTOLOGICAL FINDINGS

The common denominator and pathognomonic finding in all cases of tinea capitis is the presence of spores (and sometimes hyphae) within the hair shaft (Figures 26.3 and 26.4). Only a few follicles within a specimen may demonstrate this finding, so that transverse sectioning is the most reliable way to establish the diagnosis. The histological degree of inflammation is highly variable. In many cases, hairs packed with spores are almost free of perifollicular inflammation (Figure 26.5). A superficial and/or deep perifollicular infiltrate rich in neutrophils, eosinophils, lymphocytes and plasma cells may be present (Figures 26.6–26.9).

Exceptional cases of inflammatory tinea capitis resembling dissecting cellulitis (Figure 26.10) have a deep, dense lymphocytic infiltrate that surrounds
hair bulbs and extends into the fat (Figures 26.11–26.14). Surprisingly, although clinical findings and degree of histological inflammation are quite dramatic, infected follicles may be difficult to find. In these cases, fungal cultures may be required to establish the diagnosis.

**SUMMARY**

*Clinical correlation:* patients are most often children with bald, scaly, erythematous patches on the scalp. Dark and widened follicular ostia (‘black dots’) may be present. The clinical manifestations of inflammation are highly variable and range from mild scaling without erythema to intense erythema, induration, pustules and a purulent discharge.

*Histological findings:*

- Spores (and sometimes hyphae) within hair shafts are a diagnostic feature, but not every follicle in a specimen may be involved
- The amount of perifollicular inflammation is highly variable
A superficial and/or deep perifollicular infiltrate rich in neutrophils, eosinophils, lymphocytes and plasma cells may be present.

**BIBLIOGRAPHY**


**Figure 26.3** Transverse section of follicle infected with fungal spores. An affected shaft (right side) is adjacent to one that is totally spared. Original magnification ×400.
Figure 26.4 Vertical section of follicle infected with fungal spores. Original magnification x400
Figure 26.5 Same follicles as in Figure 26.3. Although one hair shaft is filled with spores, perifollicular inflammation is scanty. Original magnification ×200
Figure 26.6 An example of highly inflammatory tinea capitis. Such cases are often associated with pustules or a purulent discharge and crusting. Original magnification ×40
Figure 26.7 Perifollicular infiltrate containing numerous eosinophils as well as lymphocytes. Original magnification ×400

Figure 26.8 Inflammation often extends into the follicular wall, and in some cases eventuates in follicular destruction. Original magnification ×400
Figure 26.9 The deep portion of the infiltrate seen in tinea capitis is predominantly composed of lymphocytes and plasma cells. Original magnification ×400

Figure 26.10 Tinea capitis resembling dissecting cellulitis in an adolescent boy. A culture identified the causative organism as *Trichophyton tonsurans*
Figure 26.11 Tinea capitis resembling dissecting cellulitis (same adolescent boy as in Figure 26.10). Inflammation is intense but quite deep, predominantly involving the superficial fat. Original magnification ×40

Figure 26.12 Same patient as in Figure 26.11. Original magnification ×200

Figure 26.13 Hairs adjacent to the zone of inflammation may be converted to the catagen/telogen phase. Original magnification ×100
Figure 26.14 Even anagen hairs may remain unaffected although surrounded by a sea of inflammation. Original magnification ×400
Aplasia cutis congenita (ACC) is the congenital absence of skin. The scalp is by far the most common site for solitary lesions of ACC. Occasionally two or even three lesions of ACC can occur together on the scalp. Lesions are often located near the hair whorl on the vertex of the scalp, and can be various shapes and sizes (0.5–10 cm). At the time of birth the defects may be deeply ulcerated, superficially eroded or completely healed but scarred (Figure 27.1).

**HISTOLOGICAL FINDINGS**

Biopsy specimens from patients with ACC are often obtained well after birth, when the lesions are excised for cosmetic reasons. The epidermis usually demonstrates some flattening of the rete ridges. A few residual hair follicles may remain stranded in a dermis that is otherwise devoid of adnexal structures, including follicles and sweat glands. When the central portion of the lesion is compared with the surrounding normal scalp skin, the dermis usually appears thickened, because the superficial subcutaneous fat has been replaced by collagenous tissue (Figures 27.2–27.4). However, in some instances, the dermis is relatively thinned.
Collagen bundles are usually thickened and sclerotic in appearance, and are often oriented in bundles parallel to the skin surface, as would be expected in a well-healed scar (Figures 27.5 and 27.6). Occasionally, however, the thickened dermal collagen appears deceptively normal, because of the random orientation of collagen bundles. However, even in these cases, adnexal structures are absent.

**Figure 27.2** Dermal thickening and loss of follicles is evident in the central portion (right side) of this lesion of aplasia cutis congenita as compared with the border (left side). Original magnification ×40

**Figure 27.3** Flattening of the rete ridges is evident in the central portion of this lesion of aplasia cutis congenita (right side) as compared with the border (left side). Loss of follicles and thickening of the dermis are also seen. Original magnification ×40

**Figure 27.4** Same section as in Figure 27.3. Original magnification×100

Collagen bundles are usually thickened and sclerotic in appearance, and are often oriented in bundles parallel to the skin surface, as would be expected in a well-healed scar (Figures 27.5 and 27.6). Occasionally, however, the thickened dermal collagen appears deceptively normal, because of the random orientation of collagen bundles. However, even in these cases, adnexal structures are absent.
or rare (Figures 27.7 and 27.8). Inflammation is not seen in completely healed lesions of ACC.
Clinical correlation: a single (rarely multiple) hairless plaque, most often located near the hair whorl on the vertex of the scalp. Lesions are present at birth, and can be of various shapes and sizes. At birth, the defects may be deeply ulcerated,
superficially eroded or completely healed but scarred. Most lesions are excised well after birth, when they are completely healed.

*Histological findings:*

- Flattening of the rete ridges
- Thickening of the dermis with replacement of fat by collagen
- Absence of most or all adnexal structures including follicles and sweat glands
- Thickening, sclerosis and parallel orientation of dermal collagen fibers

**BIBLIOGRAPHY**

Itin P, Pletscher M. Familial aplasia cutis congenita of the scalp without other defects in 6 members of three successive generations. *Dermatologica* 1988; 177:123–5
A detailed review of hair shaft abnormalities is beyond the scope of this text. Several excellent reviews of hair shaft disorders are available and are listed at the end of the appropriate following sections. However, pathologists, dermatopathologists and dermatologists are sometimes called upon to identify a hair shaft disorder from a submitted sample of hair. This section will focus on describing the morphological features of shaft disorders as seen by light microscopy.

**TRICHRORHEXIS NODOSA**

The basic cause of trichorrhexis nodosa is trauma to the hair shafts, but any inherent weakness of the shaft may result in this defect. All the various forms of weathering (grooming, washing, styling, etc.) can cause or worsen the disorder. Examples of physical trauma include excessive brushing, the application of heat, hair pulling in trichotillomania and scratching due to pruritic scalp disorders. Examples of chemical trauma include permanent straightening (‘relaxing’; Figure 28.1), permanent waving, dyeing and shampooing. Clinically, trichorrhexis nodosa may appear as small, white or gray specks on the hair shafts. Affected hairs are susceptible to fracture, which can result in patchy or diffuse alopecia, depending on the extent of involvement. Although scalp hair is usually affected, trichorrhexis nodosa can be found on pubic hair and other hairy areas of the body.

The ‘nodes’ of trichorrhexis nodosa represent foci of frayed cortical fibers that bulge out through a ruptured cuticle. Scanning electron microscopy has shown that the earliest change is a focal loss of cuticular cells, which eventually results in hair fiber separation and fracture. Each node then takes on the appearance of two paintbrushes whose bristles have been pushed together (Figures 28.2 and 28.3). Each ‘bristle’ represents a cornified cortical cell, which can separate from adjacent cells once the binding function of the cuticle is impaired.

Trichorrhexis nodosa is often found in association with other types of hair shaft abnormalities caused by weathering or inherited hair fragility. Therefore, *trichoclasia* (‘greenstick fractures’), *trichoptilosis* (‘split ends’) and *trichoschisis*...
TRICHOSCHISIS

The term *trichoschisis* is used for a clean, transverse fracture across the hair shaft through cuticle and cortex. A localized absence of cuticle cells can be found at the site of the fracture. Although trichoschisis is occasionally found in normal hair, it usually occurs in the setting of *trichothiodystrophy* (see below).

**BIBLIOGRAPHY**

Caserio RJ. Diagnostic techniques for hair disorders. Part I: Microscopic examination of the hair shaft. *Cutis* 1987; 40: 265–70


**Figure 28.1** Hair loss in an African American woman caused by chemical relaxers. Longer hairs along the margins of the ‘bald spot’ showed dramatic trichorrhexis nodosa of the proximal shafts. Hair loss was due to fractures of the damaged shafts (see below) are often found in hairs from the same patient or even the same shaft (Figure 28.4).
Patients with trichothiodystrophy have congenitally brittle hair with low sulfur content. This hair defect serves as the common denominator for patients with neuroectodermal abnormalities and mutations in the xeroderma pigmentosum XP-B and XP-D helicase subunits of the dual functional DNA repair/basal transcription factor TFIIH. Depending on the precise mutation, patients may

**TRICHOThIODYSTROPHY**

Figure 28.2 Focal loss of the hair cuticle results in cortical fiber separation and the fragile ‘nodes’ of trichorrhexis nodosa

Figure 28.3 Fragile ‘node’ of trichorrhexis nodosa

Figure 28.4 Trichoclasis (‘greenstick fracture’ of hair) and trichorrhexis nodosa are often found in the same weathered shaft
have some or all of the following clinical manifestations: photosensitivity, ichthyosis, brittle hair, impaired intellect, decreased fertility, short stature and a wide variety of other abnormalities. The 2001 review by Itin, Sarasin and Pittelkow is particularly useful.

Hair from patients with trichothiodystrophy demonstrates several abnormalities that are visible with light microscopy. The first is trichoschisis, as described above, which accounts for the brittleness of the hair (Figure 28.5). Hair shafts may be focally flattened and ribbon-like (Figures 28.6 and 28.7). One prominent and consistent finding is an alternating light and dark banding seen with polarized light (Figures 28.8 and 28.9). The hair is placed between polarizing filters, and the banding is most evident when the filters are rotated into the ‘crossed’ or ‘extinguished’ position (background is dark). Banding can be transverse or diagonal, and can be reversed (i.e., light becomes dark) by rotating one of the filters about 45°. This banding may be due to the undulating nature of cortical fibers found in trichothiodystrophy (as seen in Figures 28.5 and 28.9).

**Figure 28.5** Trichoschisis in a patient with trichothiodystrophy. The sharply defined transverse fracture results in hair fragility and fracture.

**Figure 28.6** Ribbon-like flattening and twisting of hairs in a patient with trichothiodystrophy.
Figure 28.7 Flattened hairs in a patient with trichothiodystrophy

Figure 28.8 Alternating light and dark banding (‘tiger tail phenomenon’) in a patient with trichothiodystrophy. A polarized hair with the filters in the ‘crossed’ position is shown here

Figure 28.9 The same shaft as in Figure 28.8, without filters in place

BIBLIOGRAPHY

Coin F, Marinoni JC, Rodolfo C, Fribourg S, Pedrini AM, Egly JM. Mutations in the XPD helicase gene result in XP and TTD phenotypes, preventing interaction between XPD and the p44 subunit of TFIH. *Nature Genet* 1998; 20:184–8

**TRICHORRHESIS INVAGINATA**

This rare, congenital hair shaft abnormality is seen in Netherton’s syndrome, the combination of short, brittle hair and an ichthyosiform erythroderma (especially ichthyosis linearis circumflexa). Atopic dermatitis is also found in the majority of patients. The defective gene in Netherton’s syndrome is SPINK5, which encodes a serine protease inhibitor.

Also called ‘bamboo hair’, trichorrhexis invaginata appears as multiple small swellings spaced along the shaft at irregular intervals. Each swelling consists of a cup-like expansion of the proximal hair cortex that surrounds a rounded distal fragment, giving the defect the appearance of a ball-and-socket joint (Figure 28.10). It appears as if the distal segment has intussuscepted down into the proximal segment. These nodes are extremely fragile and susceptible to fracture (Figures 28.11 and 28.12), and consequently the hair of affected persons is short, thin and friable.

**BIBLIOGRAPHY**

PILI ANNULATI

Also called ‘ringed hair’, pili annulati is characterized clinically by alternating light and dark bands of color along the hair shafts. The colors are reversed when the hairs are viewed by light microscopy. The banded appearance is obvious only in blond or lightly pigmented hair, which often has a sandy appearance. The condition may be present at birth or appear during infancy. Hair growth is normal and in the majority of cases the hair is strong and sound, but there may be some degree of shaft fragility. Trichorrhexis nodosa-like fractures may be produced in the dark bands. There are no other associated abnormalities and treatment is not required. The condition appears to be caused by a defect in hair cornification.

The cortex of the abnormal bands contains zones of air-filled spaces (Figure 28.13), between cortical fibers and within cortical cells. Medullary cells are not involved in the process. The air-filled spaces can be confined to the central portion of the shaft, or may involve the full thickness with associated cuticle disruption.

Pili annulati has been associated with alopecia areata on more than one occasion. It is not known whether or not this is pure coincidence.
**BIBLIOGRAPHY**


**‘BUBBLE HAIR’**

This distinctive hair shaft abnormality is usually seen in young women with a localized area of uneven, fragile hairs. The involved hair is straighter and stiffer than normal. Light microscopy has demonstrated that the hair shafts contain large, irregularly spaced ‘bubbles’ that expand and thin the hair cortex (Figure 28.14). Hair fractures occur at the site of larger bubbles.

The problem is caused by traumatic hair care techniques involving heat, as from a malfunctioning hair dryer. Once the damaged hair is trimmed, the condition resolves completely with gentle hair styling.

**BIBLIOGRAPHY**


**MONILETHRIX**

Patients with monilethrix (‘beaded hair’) have extremely brittle, beaded hairs that emerge from keratotic, follicular papules. Hair shafts rarely grow more than 2–3 cm. The hairs fracture easily, usually resulting in severe alopecia. The course is variable and seasonal, and some patients may show improvement in adult life, while others seem to worsen. The condition usually appears in early childhood, predominantly on the occiput and nape, but it can affect the entire
sculpt. Occasionally the child is born bald, with beaded hairs appearing several months or years later. Facial and body hair are also involved in severe cases.

Monilethrix is usually inherited as an autosomal dominant trait with high penetrance but variable expressivity. Recessive phenotypes have also been described, as have sporadic cases. Mutations in two of the 11 known hair keratins have been associated with monilethrix. Mutations in the hHb6 gene are most common, but mutations in other genes (e.g., hHb1) can lead to the disease.

Hair shafts in patients with monilethrix show characteristic, evenly spaced, elliptical nodes that are 0.7–1 mm apart (Figure 28.15). The intervals between nodes will vary in different hairs from the same patient. Not every hair follicle is affected in a synchronized fashion. The segments of hair shaft between nodes are non-medullated, tapered constrictions. Imbricated cuticular scales are present on the nodes, whereas the internodes show abnormal longitudinal ridging and often absent scales. Scanning electron microscopy has demonstrated that the nodes correspond to the normal caliber of the hair and that the defective portion resides in the constrictions.

The hair roots have an architecture that conforms to the shaft abnormality, with alternating constrictions, so that the hair is deformed as it is being produced. Matrix cells destined to become cortical cells are particularly affected. This seems to result in a decrease in number of cortical cells and thinning of the hair shaft.

**BIBLIOGRAPHY**


Pili torti is the most misdiagnosed structural hair shaft defect, and it is often confused with monilethrix when studied with light microscopy. Hairs in pili torti are flattened and twisted on their longitudinal axes (Figures 28.16 and 28.17).

Each twist may be 90°, 180°, or up to 360°. Typically, runs of four or five twists are found at irregular intervals along the hair shaft. Involved hairs are brittle, break off easily, do not achieve normal length and often have a spangled or beaded appearance. The hairs are fragile and usually fracture within the twists. The fractured distal tip often shows longitudinal fraying (trichoptilosis). The hair follicles show no histological abnormalities other than some curvature and twisting. Although it can be present at birth or emerge after puberty, pili torti classically appears in early childhood when normal scalp hairs are replaced by brittle, spangled hairs, especially in the occipital and temporal regions. The spangled appearance is due to unequal reflection of light from the twisted surface. The eye-brows and eyelashes may also be involved. There is a variable degree of hair fragility ranging from patchy alopecia with coarse stubble to hairs of 5 cm or more. Pili torti may improve after puberty or may persist throughout life.

Pili torti is also found in various syndromes, such as Menkes’ kinky hair disease, Bjornstad syndrome and Rapp-Hodgkin ectodermal dysplasia. The numerous other distinctive features of these syndromes allow for proper diagnosis, with pili torti offering an additional clue.
Figure 28.17 Scanning electron micrograph of a shaft showing pili torti
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PILI MULTIGEMINI

This condition is typically found in the beard area, especially along the jaw. Anywhere from two to eight hair matrices, each with its own papilla and separate inner root sheath (Figure 28.18), form clusters of shafts that emerge from a single follicular canal. Surrounding the cluster of hair bulbs is a common outer root sheath that separates to invest each ascending hair shaft (Figures 28.19 and 28.20).

The shafts have various shapes in cross-section (flat, ovoid, triangular or grooved), which probably results from the pressure between hairs in the same follicle (Figure 28.21). Usually the condition is totally benign, but it may be associated with perifollicular erythema resembling folliculitis. There is no specific treatment for pili multigemini, which tends to be persistent. The abnormal hairs will simply regrow after plucking.

BIBLIOGRAPHY

Pinkus H. Multiple hairs (Flemming-Giovannini): report of two cases of pili multigemini and discussion of some other anomalies of the pilary complex. *J Invest Dermatol* 1951; 17:291-301

UNCOMBABLE HAIR SYNDROME (PILITRIANGULI ET CANALICULI; OR 'SPUN GLASS’ HAIR)

This condition was first described in 1973 as 'cheveux incoiffables' (unstylicable hair). It has also been termed 'spun glass hair' and pili trianguli et canaliculi. Many familial cases of uncombable hair have been reported, and in those instances an autosomal dominant pattern of inheritance appeared most likely. The typical clinical picture includes onset in infancy or early childhood; blonde to light brown hair; dry, frizzy and spangled hair texture; and slow to normal growth rate. The hair is disorderly, stands out from the scalp, and cannot be combed flat. Some spontaneous improvement is often noted later in childhood.
The hair shafts in uncombable hair are usually triangular in cross-section, but can be kidney-shaped, flat, or irregular. Most of the hair shafts will demonstrate the irregular, triangular shape. Longitudinal grooves running along the entire shaft are common, hence the name pili trianguli et canaliculi. Although grooving of a few hair shafts can be found on the normal scalp, in the uncombable hair syndrome at least 50% of hair shafts demonstrate this abnormality. Grooves can often be seen with the light microscope (Figure 28.22), although they are best visualized with the scanning electron microscope (Figure 28.23). Scalp biopsy reveals that the inner root sheath has the same configuration as the shaft.

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TAPERED HAIRS (‘PENCIL POINT’ HAIRS)

Tapered hairs that can be easily pulled from the scalp are always a pathological finding. Tapered hairs are found in all forms of anagen arrest. Anagen arrest is a pattern of hair loss caused by a sudden insult to the metabolic machinery of the hair follicle. The classic form of anagen arrest is seen when patients receive radiotherapy or systemic chemotherapy for the treatment of malignancy. The hair matrix is very sensitive to the toxic effects of X-irradiation and chemotherapy. Affected follicles produce shafts that become progressively smaller in volume and cross-sectional dimension. These hairs quickly taper down to a point, resulting in an extremely fragile constriction. Hair shafts easily fracture at the constrictions, and ‘pencil point hairs’ fall from the patient’s scalp in great numbers (Figure 28.24).

Several diseases cause hair loss with the features of an anagen arrest. Severe, rapidly progressive alopecia areata (e.g., alopecia totalis in evolution) has a similar effect on the metabolic machinery of hair follicles, and tapered hairs are often seen
Figure 28.25 When patients with systemic lupus erythematosus or secondary syphilis experience rapid hair loss, it sometimes has features of an anagen arrest.

PERIPILAR CASTS (‘PSEUDONITS’, ‘HAIR CASTS’)

Kligman and Brunner described this abnormality independently in 1957. Peripilar casts are often referred to as ‘hair casts’, although the latter designation is imprecise. They are tubular masses of amorphous material that are perforated centrally by the hair shaft. Hair casts are conical proximally, and their distal end may taper or in larger casts may expand into a funnel shape. Some casts conform to the shape of the follicular canal and infundibulum. True (‘classic’) peripilar keratin casts are commonly composed of cornified external root sheath (Figure 28.26), rarely composed of internal root sheath, and sometimes composed of both external and internal root sheaths. These types can be distinguished by staining with 4-dimethylaminocinnamaldehyde, which stains the inner root sheath a reddish color, or with toluidine blue, which stains the inner root sheath an intense blue (Figure 28.27).
Figure 28.22 The prominent grooving seen in the ‘uncombable hair syndrome’ is visible even with light microscopy

Figure 28.23 The grooving seen in the ‘uncombable hair syndrome’ is best visualized with scanning electron microscopy

Figure 28.24 Gently pulling on the hair was sufficient to remove numerous ‘pencil point’ hairs from a patient receiving systemic chemotherapy for a malignancy

BIBLIOGRAPHY

Brunner MJ, Facq LM. A pseudoparasite of the scalp hair. Arch Dermatol 1957; 75:583
Kligman AM. Hair casts. Arch Dermatol 1957; 75:509–11
Figure 28.25 A tapered hair gently pulled from the scalp of a patient with rapidly worsening alopecia areata

Figure 28.26 A peripilar cast composed of infundibular keratin. This is the most common kind of hair cast
Figure 28.27 Peripilar casts composed of distal fragments of inner root sheath. These casts stain a dark blue with toluidine blue
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