Trichoscopy in Hair Shaft Disorders

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KEYWORDS

• Trichoscopy • Dermoscopy • Dermatoscopy • Alopecia • Hair shaft • Ectodermal dysplasia • Classification • Hair fragility

KEY POINTS

- Trichoscopy allows the practitioner to analyze the structure and size of growing hairs without the need to pull hair for examination.
- Trichoscopy allows establishing the diagnosis of most of the known hair shaft disorders.
- Some structures are only visible with dry trichoscopy, whereas other may require an immersion fluid.
- In patients suspected of trichothiodystrophy, a polarized dermoscope should be used.

INTRODUCTION

Trichoscopy (hair and scalp dermoscopy) has been successfully applied in practical dermatology in recent years.[1,2] One of the major fields of progress is the use of trichoscopy for evaluation of hair shaft diseases in children.[3,4] This noninvasive technique replaced light microscopy, which required pulling of multiple hairs for investigation. This was in particular burdensome for patients with hairs prone to fracturing and in diseases, where only few hairs might be affected, but the examination is crucial for establishing a diagnosis. A best example is Netherton syndrome, which occasionally required pulling a few hundred hair shafts to establish a diagnosis. In 2007 and 2008, a Polish group first described the application of trichoscopy in hair shaft disorders.[5,6] Now trichoscopy may be successfully applied in most of the inherited and acquired hair shaft disorders.

NORMAL HAIRS

Normal hair shafts are uniform in thickness and color throughout their length.[7,8] Terminal hairs may have a medulla, which is continuous, interrupted, fragmented, or absent.[9] Up to 10% of normal human scalp hairs are vellus hairs.[7,8] These are hairs that are less than 3-mm long and less than 30-μm thick.

CLASSIFICATION OF HAIR SHAFT ABNORMALITIES IN TRICHOSCOPY

A classification of hair shaft abnormalities in trichoscopy was proposed by Rudnicka and colleagues.[18] It distinguishes the following groups of hair shaft features observed by trichoscopy: (1) hair shafts with fractures, (2) hair narrowings, (3) hairs with nodelike structures, (4) curls and twists, (5) bands, and (6) short hairs. A short hair is defined as a hair in which an entire hair shaft is visible in 1 field of view of a dermoscope (10-fold to 20-fold magnification). These hairs are usually less than 10-mm long.

HAIR SHAFT DISEASES

Monilethrix and Monilethrix-Like Hairs

Monilethrix is characterized by regular, periodic thinning of hair shafts and a tendency to fracture

Disclosure Statement: No conflicts of interest.

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Dermatol Clin ■ (2018) ■–■
https://doi.org/10.1016/j.det.2018.05.009

at constricted points. Nodosities correspond to the normal hair caliber, whereas the defect is in the constricted sections.[10]

Monilethrix is a hereditary disorder, typically caused by autosomal dominant mutations in type II hair keratin genes KRT 81, KRT 83, and KRT 86.[11] Mutations in the desmoglein 4 (DSG4) gene are associated with autosomal recessive monilethrix and monilethrix-like congenital hypotrichosis, which differ from classic monilethrix by barely visible internodes, which do not show constant periodicity.[12]

From early childhood, patients with monilethrix present with short and fragile hairs that never grow long enough to require a haircut. Noninvolved hairs are seldom longer than 5 cm to 8 cm. Other hairy areas, such as eyebrows; eyelashes; or axillary, pubic, and body hair, may also be involved.[13] The disease tends to improve with age.

The hair shaft fragility is associated with perifollicular abnormalities, which range from subtle perifollicular erythema to large hyperkeratotic follicular papules. Other, rare, ectodermal symptoms in these patients may include koilonychia, brittle nails, syndactyly, juvenile cataract, decreased visual field, and dental abnormalities.[13]

Several studies have investigated the application of trichoscopy in diagnosing monilethrix. The first study by Rakowska and colleagues[6] showed that trichoscopy allows visualizing abnormalities in both terminal and vellus hairs of the scalp. Hair shafts show uniform elliptical nodosities and intermittent constrictions causing variation in hair shaft thickness (**Fig. 1**). Hairs are bended regularly at multiple locations and have a tendency to fracture at constriction sites.[5,6] The term, *regularly bended ribbon sign*, was suggested to differentiate trichoscopy features of monilethrix from pseudomonilethrix and other causes of hair loss.[6] Horny follicular papules appear as big yellow dots, when evaluated in trichoscopy with immersion fluid, while perifollicular scaling and keratotic follicular plugs may be observed in dry trichoscopy.

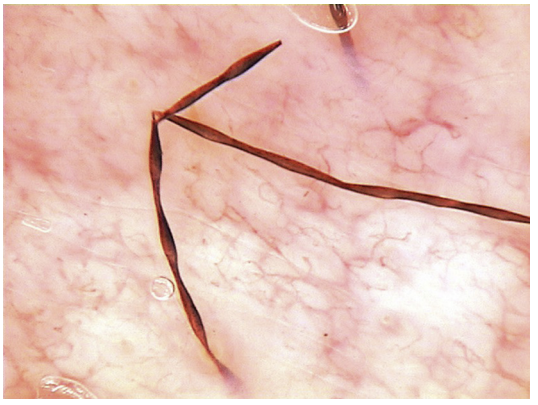

Fig. 1. Monilethrix. Trichoscopy shows hair shafts with regularly distributed nodes (correspond to normal hair shaft thickness) and internodes (correspond to narrowing of hair shaft). If only small proportion of hair shafts is affected, this abnormality is more likely to found in occipital area (×70).

A later report showed that beaded hairs arise from the keratotic papules on neck.[14] Typical trichoscopy findings were also observed in the affected body hairs of the forearms.

Not all hairs are affected by monilethrix. Probably the best areas for searching for typical abnormalities are the temporal and occipital areas, where most hair shafts show features of the disease. In rare cases, however, other locations are predominantly affected. The development of hair loss mimicking androgenetic alopecia with typical monilethrix hairs in the androgen-dependent areas of the scalp was reported.[15]

The term, *pseudomonilethrix*, was used to describe irregular, square-shaped, flattening of hair shafts. It remains controversial whether pseudomonilethrix is a true disease[16] or an artifact produced by either procedure of preparing hairs for microscopic examination or by excessive use of cosmetic hair care products.[17] There was no reported case of pseudomonilethrix on trichoscopy despite massive use of this diagnostic method in recent years. This may be an indirect confirmation that pseudomonilethrix is an artifact, which may be visible in light microscopy but is easy to identify as excessive use of hair cosmetics in the trichoscopic evaluation.

A pseudomonilethrix effect may be observed in patients who use hair styling gels.[6] Thus, patients should be advised not to use these products between hair washing and trichoscopy. Also, ultrasound gel used as immersion fluid can make hair shafts appear irregularly flattened.[6]

Pseudomonilethrix has to be distinguished from monilethrix-like hairs, which show the same type of ovoid constrictions as in monilethrix but with no regularity characteristic for true monilethrix. These constrictions have been also called Pohl-Pinkus constrictions. Monilethrix-like hair shafts may be observed in diseases with variable course, such as alopecia areata and lichen planopilaris, or in patients undergoing chemotherapy (**Box 1**).[18–20]

Trichorrhexis Nodosa

Trichorrhexis nodosa is a condition in which the shaft splits longitudinally into numerous small fibers within a restricted area of the shaft. The outer fibers bulge out, what causes a segmental increase in hair diameter. Macroscopically this may resemble nodules located along the hair