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Authors

Flanigan, Kendall
Greer, Jeremy
Maruthur, Mario

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Loose anagen syndrome in one identical twin girl

Kendall Flanigan BS BA, Jeremy Greer MD, Mario Maruthur MD

Affiliations: Department of Dermatology, Medical College of Georgia, Augusta University Medical Center, Augusta, Georgia, USA

Corresponding Author: Jeremy Greer, 610 Surrey Lane, Martinez, GA 30907, Tel: 423-747-1450, Email: jegreer@augusta.edu

Abstract

Loose anagen syndrome (LAS) is a hair disorder involving insufficient anchoring of the hair follicle to the scalp owing to an autosomal dominant or sporadic mutation in the gene encoding keratin 6. There are three phenotypes of LAS, including type B, which presents in young, light-haired girls as unruly, uncombable hair with diminished growth. We present a 2-year-old girl with LAS type B whose identical twin sister was unaffected. The diagnosis was confirmed with a painless hair pull test proven to contain anagen hairs with ruffled cuticles on trichoscopy, preventing the need for unnecessary referrals and diagnostic tests.

Keywords: loose anagen syndrome, hair analysis, hair disease, keratin 6hf, human, light microscopy

Introduction

Loose anagen syndrome (LAS) is a hair disorder involving insufficient anchoring of the hair follicle to the scalp owing to mutations in the gene encoding keratin 6 [1-3]. Loose anagen syndrome typically presents in young girls as thin, fine hair that can be easily and painlessly extracted [1-3]. Type A LAS includes decreased hair density with short and thin hair [3]. Type B includes unruly, uncombable hair, consistent with our patient's presentation [3]. Type C, the adult phenotype, includes normal appearing hair with excess shedding [3].

The diagnosis of LAS begins with a hair pull test; 40-60 hairs are pulled painlessly [1,2]. Examined using trichoscopy, the presence of more than 70% anagen hairs that lack inner and outer root sheaths and have a ruffled cuticle is diagnostic of LAS [1-3].

Case Synopsis

A 2-year-old identical twin girl presented to the clinic for evaluation of thinning and unruliness of her hair for 6 months. The patient's hair was previously normal and these changes began slowly and diffusely. Her identical twin sister had no hair abnormalities. The family denied any medication use or recent environmental changes, including any new products used on the face or scalp. There was no family history of similar findings. Her physical examination was remarkable for diffuse unruly and thin hair on the scalp that could be extracted easily and painlessly (**Figure 1A**) and was distinctly different from that of her twin (**Figure 1B**). The rest of the skin examination was unremarkable including normal nails; the patient was otherwise systemically well.

We performed a hair pull test. The hairs were extracted painlessly and more than 70 percent were determined to be anagen hairs under light microscopy (**Figure 2**). The hairs lacked inner and outer root sheaths and had ruffled cuticles (**Figure 2**). These findings, in the setting of the patient's clinical picture, are consistent with LAS, type B phenotype. We reassured the patient's family about this self-limited condition thereby relieving significant anxiety for the parents and preventing the need for unnecessary diagnostic tests.

Case Discussion

Loose anagen syndrome (LAS) is a hair disorder related to insufficient anchoring of the hair follicle to the scalp [1-3]. During the normal hair growth cycle, hair proceeds through phases of growth (anagen), apoptosis and regression (catagen), and rest (telogen) after which the hair falls out [4]. These



Figure 1. Clinical features of hair in the patient and her unaffected twin. **A)** Thin, unruly hair in patient. **B)** Unaffected hair in twin. Pictures taken in office by the patient's parent.

phases differ in the amount of nourishment via blood supply the follicle receives [4]. After telogen, the dermal papilla enters the various stages of anagen, including proanagen, in which the hair shaft grows inside the hair follicle, and metanagen, in which the hair shaft surfaces from the hair follicle and is present on the skin [3]. Metanagen is believed to have numerous substages that affect adhesion of hair, including formation of the inner root sheath (IRS) which anchors the hair to the follicle [3].

In LAS, premature keratinization of the inner root sheath leads to impaired adhesion among the hair shaft, inner root sheath, and outer root sheath [1-3]. This prematurely terminates anagen, causing failure of subsequent hair growth [3]. Loose anagen syndrome is attributed to an autosomal dominant or sporadic mutation in the gene encoding keratin 6 [2,3]. One mutation in *K6HF*, the companion layer keratin which represents the innermost layer of the outer root sheath, prematurely keratinizes the inner root sheath which impairs adhesion between the inner root sheath cuticle and the hair shaft cuticle [1,5,6]. An additional mutation in *K6IRS*, which is specific to the inner root sheath, impairs adhesion between the inner and outer root sheaths and prematurely terminates anagen [6]. This case of monozygotic twins presenting with one affected and one unaffected could be secondary to variable expressivity or a post-zygotic mutation.

Loose anagen syndrome is a self-limited condition that typically improves with time as many hairs begin to attain normal anagen appearance prior to adulthood; thus, reassurance is essential [2,3]. Often no treatment is necessary although patients may remain predisposed to thin hair or thick hair that easily sheds [2,3]. Topical minoxidil may be indicated in patients with severe symptoms leading to significant cosmetic concern [1,3,5]. It is believed that the presence of androgens may affect the follicle through the mesenchymal-epithelial interaction and that minoxidil may prolong the anagen phase and increase local blood flow, thus lengthening the hair [1,5]. Accurate and timely diagnosis can prevent the need for unnecessary referrals and diagnostic tests.

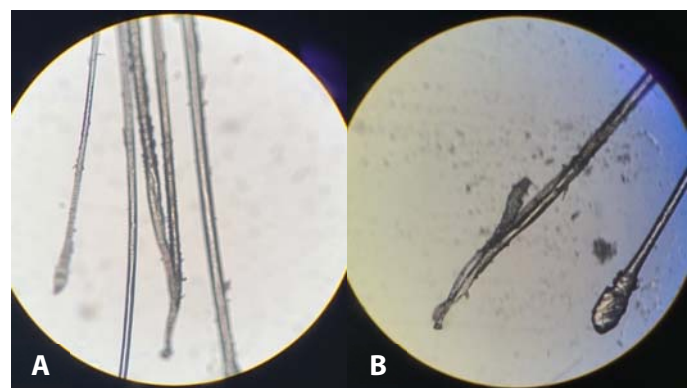


Figure 2. Hair under light microscopy. **A)** Anagen hairs with ruffled cuticles. **B)** An anagen hair with a detaching root sheath.

Conclusion

Loose anagen syndrome (LAS) is a hair disorder of inadequate anchoring of the hair follicle to the scalp because of mutations in the gene encoding keratin 6. Loose anagen syndrome type B presents as unruly, uncombable hair with diminished growth. We present a case of LAS type B in a 2-year-old girl whose identical twin sister was unaffected. The diagnosis

was confirmed with a painless hair pull test proven to contain anagen hairs with ruffled cuticles on trichoscopy, preventing the need for unnecessary referrals and diagnostic tests.

Potential conflicts of interest

The authors declare no conflicts of interests.

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