

## Loose Anagen Hair Syndrome

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### CASE SUMMARY

#### History

A nine-year-old girl complained of sparse and unmanageable scalp hair since birth. She seldom required haircuts, as her hair did not grow sufficiently long. She also reported that her scalp hair could be pulled out easily and painlessly. She denied habit of hair pulling. Her past history was unremarkable and there was no history of long-term drug taking. Increased hair loss was occasionally noted after bouts of fever. There was no significant family history of hair loss.

#### Physical examination

There was diffuse thinning of scalp hair (Figures 1 and 2). Most hair was short and curved (from 4 to 9 cm). Hair pull test was positive. There were no inflammation, scarring, bald patches, unevenness of hair

color, exclamation hair or keratosis pilaris. There was no eyebrow or eyelash involvement. Her nails and teeth were normal. No other physical or mental abnormality was found.

#### Differential diagnosis

The differential diagnoses include conditions that cause non-scarring alopecia: alopecia areata, trichotillomania, anagen effluvium, telogen effluvium, anaemia or systemic illness, hair shaft disorders, loose anagen hair syndrome and androgenetic alopecia.

#### Investigations

She had normal complete blood counts, renal and liver function tests, serum iron and total iron binding capacity. Scalp biopsy was non-diagnostic with unremarkable anagen follicles, no peri-follicular inflammatory cell infiltration and negative immunofluorescence. Light microscopy of hair from pull test showed that over 90% were anagen hair. In longer hairs, alternating thick elliptical nodes and narrow internodes that lack medulla were also seen. There was no twisting or fracture. Polarized light microscopy revealed no alternating light and dark bands. Scanning electron microscope showed normal to slight weathering of



Figure 1: Diffuse non-scarring alopecia (occipital view)



Figure 2: Diffuse non-scarring alopecia (right parietal view). Hair was short and curved. Hair pull test was positive

cuticle covering the internodal areas. Excessive weathering with marked disruption of cuticle was seen in the thickened and flattened nodal areas.

### Diagnosis

Despite the unusual elliptical nodes seen at the hair shaft, the patient did not have other supportive associated features of monilethrix, such as follicular keratosis at scalp, keratosis pilaris and family history. The positive hair pull test was also against the diagnosis of monilethrix. The clinical and pathological findings therefore best fit the diagnosis of loose anagen hair syndrome with some hair shafts showing features of monilethrix.

### Management

The diagnosis and natural history of loose anagen hair syndrome were explained to both patient and mother who agreed to be treated conservatively.

## REVIEW ON LOOSE ANAGEN HAIR SYNDROME

### Normal anatomy of scalp hair<sup>1</sup>

The vast majority of scalp hairs are anagen or actively growing hairs. Most of the remaining are telogen, or "resting hairs" that are about to be shed. The normal range for numbers of telogen hair is 4-25% with a mean of about 15%. Telogen counts above this range are presumptively abnormal. Hair pull test performed on normal scalp may yield several hairs and they are always telogen. To obtain anagen hair, it must be forcibly plucked as the cuticular cells of inner root sheath (IRS) and hair shaft interlock with each other. Hairs that are forcibly plucked are usually removed with intact inner and outer root sheath.

### Definition

A disorder of anagen hair anchorage to the hair follicle, characterized by ability to easily and painlessly pull out large numbers of anagen hairs from the scalp.

### Epidemiology

First described in 1986, the true incidence is unknown. The majority of cases are fair-haired girls between two to nine years old.<sup>2</sup>

### Aetiology

Familial cases are frequently observed suggesting autosomal dominant inheritance.<sup>2</sup>

### Pathogenesis

This is thought to be due to a generalized disturbance of cellular adhesion, most pronounced between the inner root sheath and the hair cuticle. Aberrant keratinization of IRS has been observed. Traction causes separation of the hair from IRS, which remains attached to the rest of the follicle and stays in the dermis.

### Clinical features

There is considerable clinical variation from diffuse thinning of hair of uneven length to focal areas of alopecia. Hair is dry and lusterless and is frequently "unruly".

### Pathology

Trichogram shows 98-100% plucked hairs to be anagen hairs. Light microscopy may demonstrate ruffling of the cuticle adjacent to the anagen bulb giving a 'floppy sock' appearance.<sup>2</sup> There are no inner and outer root sheaths. Twist and grooves may be seen in shaft. Electron microscopy reveals hairs with abnormal shapes: triangular, quadrangular, heart or kidney shaped. Longitudinal grooves with slight twisting about their axis can sometimes be seen. Scalp histology shows a cleft between the inner and outer root sheaths,<sup>3</sup> and the IRS appears homogenized due to premature keratinization of the Huxley and Henle layers.<sup>4</sup> There is no peri-follicular inflammation, and some hairs are not involved in the process.

### Differential diagnosis

This condition should be distinguished from alopecia areata (exclamation mark hairs) and from telogen effluvium (clubbed hairs). The unruly hair may be confused with pili torti, uncombable hair syndrome or woolly hair.<sup>5</sup>

### Prognosis and management

In most patients the condition improves spontaneously in mid to late teens and reassurance is all that is required. It is important to look out for associated

features including hypohidrotic ectodermal dysplasia and ocular coloboma.<sup>2</sup>

***Learning points:***

***Loose anagen hair syndrome is a disorder of anagen hair anchorage to the hair follicle, characterized by ability to easily and painlessly pull out large numbers of anagen hairs from the scalp. Spontaneous improvement is expected in most cases.***

**References**

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