

Trichoscopy findings in loose anagen hair syndrome: rectangular granular structures and solitary yellow dots

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Key words:

alopecia, dermatoscopy, dermoscopy,
hair, hair loss

Abstract

Background: Loose anagen hair syndrome (LAHS) is typically diagnosed in girls older than 2 years who present with hair that "will not grow". Hair microscopic examination shows absent inner and outer root sheaths, ruffling of the cuticle on the proximal hair shaft and deformed pigmented anagen bulbs.

Objective: The aim of the study was to assess whether there are characteristic trichoscopic features favoring the diagnosis of LAHS.

Patients and methods: Eighty nine children patients were included into the study (24 girls with LAHS, 25 with alopecia areata, 20 with telogen effluvium and 20 healthy children). In all groups trichoscopy was performed. Trichoscopy images were analyzed for abnormalities in the hairs shafts, the hair follicle openings and the interfollicular area.

Results: Dirty dots were present in all groups. A unique feature of LAHS was the presence of rectangular black granular structures which differs from dense black dots seen in patients with alopecia areata. This feature was observed in 71% of patients with LAHS. Follicular units with single hairs constituted 92,9% of hair units in these patients (65,5% in telogen effluvium and 53% in the control group). Solitary yellow dots were found in 50% of patient with LAHS and in 24% of patients with alopecia areata, but was not found in control group or in patients with telogen effluvium.

Conclusion: The trichoscopy features favoring the diagnosis of LAHS are: rectangular black granular structures, solitary yellow dots and major predominance of follicular units with single hairs. (*J Dermatol Case Rep.* 2015; 9(1): 1-5)

Background

Loose anagen hair syndrome (LAHS) is an autosomal dominant trait, with incomplete penetrance and variable and age-dependent expression. It is typically diagnosed in young girls older than 2 years who present with hair that "will not grow" or hair that is thin and sparse but adequate to cover the scalp. Usually the hair color is blonde but in the recent years LAHS in brown and black hairs was described.¹

Clinical presentation is heterogeneous. Three primary LAHS phenotypes have been described based mainly on the clinical signs of reduced hair, hair length, increased hair shedding and altered hair texture: type A, with sparse hair that does not grow long; type B, characterized by diffuse or

patchy, unruly hair; and type C, characterized by normal-appearing hair with excessive shedding of loose anagen hairs and predominantly affects adults.² There is a tendency for the clinical presentation of patients with LAHS types A and B to evolve into LAHS type C with age.³

The diagnosis relies on the number and percentage of hairs in the pull test and loose anagen hairs in the trichogram. Pull test reveals more than 3 to 10 hairs which are easily and painlessly plucked.^{3,4} Loose anagen hairs have absent inner and outer root sheaths, ruffling of the cuticle on the proximal hair shaft (floppy sock appearance), and deformed pigmented anagen bulbs that may appear long and tapered, twisted, or positioned at an acute angle to the long axis of the hair shaft (Fig. 1).^{5,6}



Figure 1

Microscopic hair examination in loose anagen syndrome reveals absent inner and outer root sheaths, ruffling of the cuticle on the proximal hair shaft (floppy sock appearance), and deformed pigmented anagen bulbs. (x10)

Tosti *et al.*⁹ proposed diagnostic criteria for LAHS that include positive pull test results with painless extraction of at least 10 loose anagen hairs and the presence of more than 80% loose anagen hairs on trichogram, but these criteria may be considered too strict to identify individuals who are mildly affected to be diagnosed. Therefore revised criteria were suggested, which indicate that the diagnosis of LAHS should be established when the trichogram shows at least 70% loose anagen hairs.¹ Cantatore-Francis and Orlow proposed that LAHS should be diagnosed only when there are more than 50% loose anagen hairs in the trichogram.¹⁰

Trichoscopy (hair and scalp dermoscopy) is a rapid in-office technique, which has become a standard procedure in differential diagnosis of hair loss.^{11,13} However, there are no

specific trichoscopy features that favor the diagnosis of LAHS.¹⁴ The differential diagnosis includes alopecia areata (AA), trichotillomania, congenital hair shafts disorders and telogen effluvium (TE). Trichotillomania and hair shafts abnormalities have characteristic trichoscopy features and can be easily recognized by trichoscopy.¹⁴⁻¹⁸

The aim of the study was to assess whether there are any specific trichoscopy features which may facilitate the the diagnosis of LAHS.

Patients and methods

This retrospective study included 89 children patients, which were seen in our outpatient clinic between 2008-2015. Twenty four children (all girls) were diagnosed with LAHS (diagnosis confirmed by trichogram), 25 (11 girls and 14 boys) with alopecia areata (diagnosis confirmed by clinical examination and trichoscopy, histopathological and mycological examination only in doubtful cases), 20 (12 girls and 8 boys) with TE (diagnosis confirmed by trichogram) and 20 (11 girls and 9 boys) were healthy patients (control group). The groups were adjusted for age. The mean age of children was 5.25 (range 3-9) years in LAHS group, 6.2 (range 3-10) years in AA group, 6.5 (range 4-9) years in TE group and 6.55 (range 3-10) years in healthy control group.

Trichoscopy was performed with the use Fotofinder II (5-8 images at each, the 20- and 70-fold magnification). A total of 1126 images were analyzed by two independent blinded evaluators who evaluated abnormalities in hair shaft structure and skin surface. After the evaluation results were unblinded, the trichoscopic features were assigned to respective patients groups.

The occurrence of the scored trichoscopic criteria within each group was evaluated by Games-Howell multiple comparisons test and ANOVA. Significance level was set to $p < 0.001$.

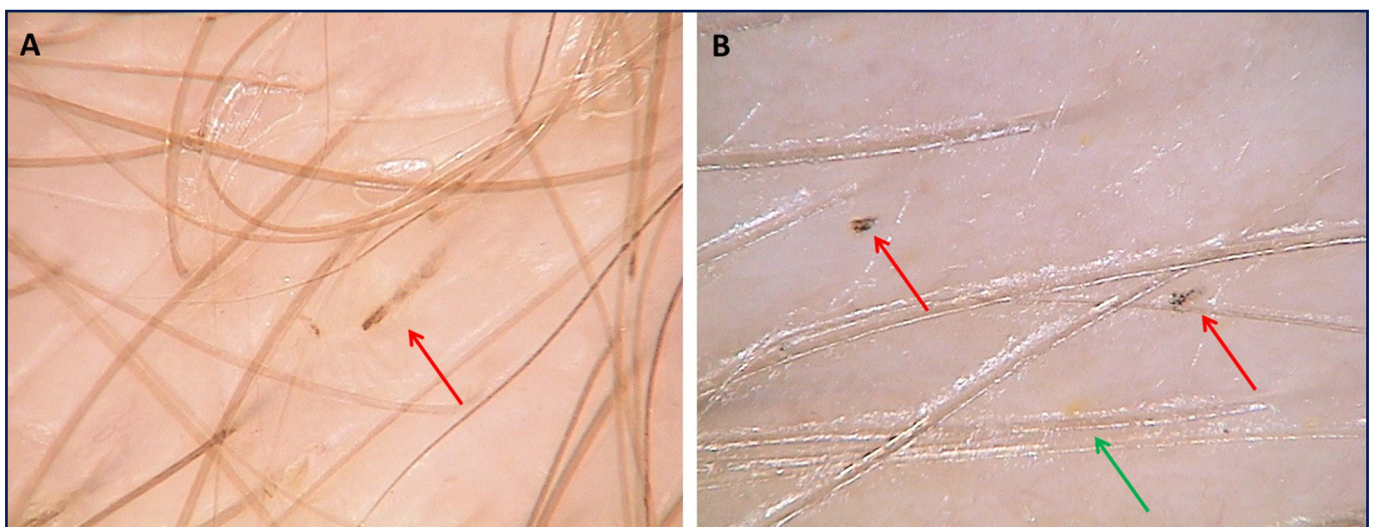


Figure 2

Trichoscopy in loose anagen hair syndrome shows black rectangular features with granular structure (A,B; red arrows) and solitary yellow dots seen by trichoscopy in loose anagen hair syndrome (B; green arrow). (x70)



Figure 3
In alopecia areata trichoscopy reveals black dots which are dense in structure (red arrows). (x70)

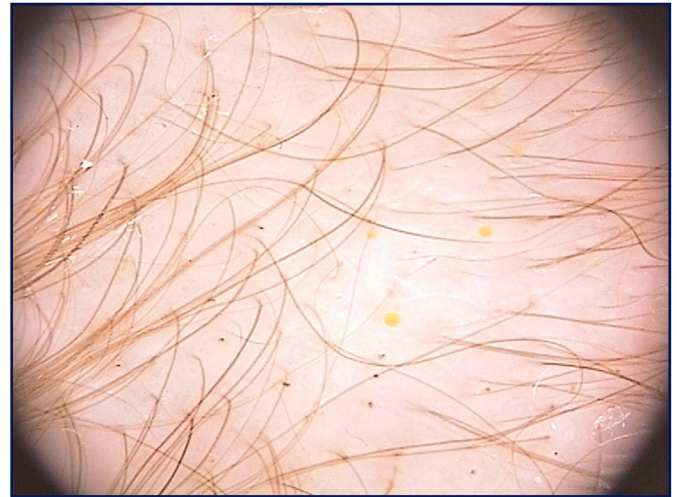


Figure 4
Black rectangular features and solitary yellow dots seen by trichoscopy in loose anagen hair syndrome. (x20)

Results

Trichoscopy features seen in analyzed images were: dirty dots, single rectangular black granular structures, groups of black dots dense in structure, single yellow dots, clustered yellow dots, exclamation-mark hairs, percentage of pilosebaceous units with single hair, percentage of pilosebaceous units with triple hairs.

Dirty dots were a common finding in all groups: 75% in LAHS group (18/24) vs 75% in control group (15/20) vs 72% in AA group (18/25) and 72% in TE group (15/20). Statistical differences between groups were not significant. A unique feature seen only in LAHS (71%; 17/24) was the presence of rectangular black granular structures (Fig. 2) different from black dots with uniform dense structure seen in patients with AA (44%; 11/25) (Fig. 3).

Follicular units with single hair constituted 92,9% of hair units in LAHS group, 53,5% in control group and 65,5% in TE



Figure 5
Easily pull out hair with rectangular anagen bulb in loose anagen syndrome seen by trichoscopy (red arrow). Pig-tail hairs are sometimes seen as a result of hair regrowth.

Table 1. Trichoscopy features in patients with loose anagen hair syndrome, alopecia areata, telogen effluvium and healthy controls.

	LAHS number of patients (%)	AA number of patients (%)	TE number of patients (%)	HC number of patients (%)	Statistical significance		
					LAHS vs AA	LAHS vs TE	LAHS vs HC
Dirty dots	18/24 (75%)	18/25 (72%)	15/20 (72%)	15/20 (75%)	NS	NS	NS
Rectangular granular black structures	17/24 (71%)	2/25 (8%)	0/25	0/25	p<0,001	p<0,001	p<0,001
Groups of dense black dots	0/24 (34%)	11/25 (44%)	0/25	0/20	p<0,001	P<0,001	P<0,001
Solitary yellow dots	12/24 (50%)	6/25 (24%)	1/25 (4%)	1/20 (5%)	p<0,001	p<0,001	p<0,001
Clustered yellow dots	0/24 (%)	17/25 (68%)	0/25	0/20	p<0,001	NS	NS
Exclamation-mark hair	0/24	17/25 (68%)	0/25	0/20	p<0,001	NS	NS
Percentage of follicular units with single hair	0,929	/	0,535	0,655	/	p<0,001	P<0,001
Percentage of follicular units with three hair	0	/	0,015	0,06	/	NS	p<0,001

* LAHS = loose anagen hair syndrome; AA = alopecia areata; TE = telogen effluvium; HC= healthy controls; NS = not significant;

group ($p < 0,001$). Follicular units with three or more hairs were seen in healthy controls (6%) and in TE (1,5%), not in LAHS. AA images were not searched for this parameter because of a different clinical presentation (focal). Solitary yellow dots (Fig. 4) were found in 50% of patient with LAHS (12/24) and this feature was not found in control group and in TE, although they were seen in 24% of patients with AA (6/25) ($p < 0,001$). Clustered yellow dots defined as three or more yellow dots in adjacent follicular units were seen only in AA group (68%; 17/25), not in others ($p < 0,001$). Exclamation mark hairs were seen only in children with AA (68%; 17/25, $p < 0,001$).

The numbers and frequency of trichoscopy features in patients with LAHS, AA, TE and healthy control are presented in detail in Table 1.

Discussion

On scanning electron microscopy of loose anagen hair, there are distortions in the shape and a pulling back of the cuticles of the proximal hair shaft, which correspond to the ruffling of the cuticle seen with light microscopy. The main defect is presumed to be abnormal adhesion between the cuticle of the inner root sheath and the cuticle of the hair shaft. The absent inner root sheath in a pulled anagen hair in the context of LAHS supports this concept. The major pathological changes of loose anagen hair in LAHS were found in the inner root sheath of anagen follicles. These consist of vacuolization and intercellular edema in Huxley cells, and dyskeratotic changes of Henle cells and cuticle cells of both the inner root sheath and hair shaft. These structural abnormalities conceivably could interfere with the normal interdigitation of the cuticle cells of the inner root sheath and hair shaft and disturb the normal anchoring function of the inner root sheath. Ultimately this would allow hairs to be easily pulled out.⁸

Black dots seen in trichoscopy are usually assumed as a "cadaverized" hairs or hair residues in the follicular opening. These are hairs that have undergone necrosis as a result of a severe inflammatory process but are still retained within a hair follicle. Black dots are most commonly found in alopecia areata, and they may be present in groups.¹⁹ Black structures seen in our study in patients with LAHS were solitary and differ from those seen in AA in a structure and shape. Black dots in AA usually have a dense structure, while the structures visible in LAHS are rather rectangular in shape and have a granular structure.

Our concept is that the rectangular shape of black structures corresponds to rectangular shape of an anagen follicle, which has become "rapidly empty" in the course of LAHS.

Yellow dots are follicular openings lacking hair shafts but filled with sebum or keratotic material. In cases of LAHS the solitary yellow dots are seen next to rectangular black structures. The chronology of events is unknown. We hypothesize that most probably the black structure is visible short after the anagen hair is lost and over time a yellow dot develops and is increasingly visible before a new upright regrowing hair or pig-tail hair (Fig. 5) appears.

Follicular units with single hair constituted 92,9% of hair units in LAHS group and 65,5% in control group and the difference was statistically significant ($p < 0,001$). Units with 3 hairs were found in healthy patients and in the TE group (AA group was not examined for this parameter because of focal hair loss). Triple hair units can be assumed as good hair health condition. A pulled hair with an anagen bulb is rarely seen in LAHS by trichoscopy (Fig. 5).

Conclusions

Trichoscopy may be helpful in differential diagnosis of LAHS. The characteristic trichoscopy features of the disease are: solitary rectangular black granular structures (which differ from densely structured black dots seen in alopecia areata), solitary yellow dots, and major predominance of follicular units with single hairs.

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