Trichoscopy of Focal Alopecia in Children – New Trichoscopic Findings: Hair Bulbs Arranged Radially along Hair-Bearing Margins in Aplasia Cutis Congenita

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Key Words

Alopecia areata · Aplasia cutis congenital · Alopecia trichoscopy · Dermoscopy

indicates that radially arranged hair bulbs visible through the translucent epidermis are characteristic of nonbullous type aplasia cutis congenita.

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Abstract

Purpose: To establish whether trichoscopy can be useful in the differential diagnosis of patchy alopecia in children. **Procedures:** The study was a retrospective analysis (2012–2015) and included 68 patients under 6 years of age. The inclusion criteria were age and the presence of 1–3 alopecia patches. A total of 124 alopecia patches were examined with the use of a videodermoscope: 102 alopecia areata, 8 tinea capitis, 6 trichotillomania, 3 temporal triangular alopecia and 5 aplasia cutis congenita. **Results:** In all aplasia cutis congenita lesions, trichoscopy revealed elongated hair bulbs visible through the semitranslucent epidermis, seen at the hair-bearing margin and radially arranged. Hair regrowth [upright regrowing hairs (44%), circular hairs (23%) and vellus hairs (20%)] was observed in the majority of alopecia areata patches. For triangular alopecia, upright regrowing hairs (100%; 3/3), vellus hairs (100%; 3/3) and circle hairs (33%; 1/3) were seen inside the alopecia patch. **Conclusion:** Trichoscopy is a useful technique for the differential diagnosis of patchy alopecia in children. A novel finding in this study in-

Background

The majority of alopecia in children is presented as patchy alopecia, which is most commonly diagnosed as alopecia areata. However, other causes of patchy alopecia such as tinea capitis, trichotillomania, temporal triangular alopecia (TTA), nevus sebaceous and aplasia cutis congenita (ACC) can be easily missed [1].

Trichoscopy (hair and scalp dermoscopy) is a rapid in-office technique, which has become a standard procedure in the differential diagnosis of hair loss [2]. The procedure can easily confirm alopecia areata due to its particular trichoscopic features: exclamation mark hairs, yellow and black dots, and cadaver hairs [3]. The trichoscopic features of trichotillomania in adult patients (flame hairs, tulip hairs, coiled hairs, hook hairs, v-sign and irregularly broken hairs) have been described [4, 5], while the characteristic findings in tinea capitis (comma hairs, zigzag hairs, corkscrew hairs and block hairs) have also been

documented [6, 7]. To the best of our knowledge, there is only one description referring to the trichoscopy of ACC [8]. Trichoscopic findings in TTA have been described as normal follicular openings with vellus hairs covering the area of alopecia, and terminal hairs on the outskirts of the lesion [9].

The aim of the study was to establish the usefulness of trichoscopy in the differential diagnosis of the abovementioned entities.

Procedures

The study was a retrospective analysis (2012–2015) and included 68 patients under 6 years of age (39 girls and 29 boys; median age 4.5 years; range 14 months to 6 years) and was approved by the local ethics committee. The inclusion criteria were age and the presence of 1–3 alopecia patches. A total of 124 alopecia patches were examined with the use of a videodermoscope (Fotofinder II) as a routine procedure, of which 102 were recognized as alopecia areata, 8 as tinea capitis, 6 as trichotillomania, 3 as TTA and 5 as ACC. Other procedures performed to confirm the diagnosis included trichograms, fungal cultures and histopathological examinations. In 1 patient, ACC was recognized as a spectrum of Kabuki syndrome. This diagnosis was based on the presence of typical facial abnormalities, short stature and mild mental retardation. Three small patches of alopecia were also present (fig. 1). The 2 remaining patients had a sporadic type without other abnormalities. In all cases, the patches of alopecia were present from birth (in utero healing).

A total of 453 images were analyzed by two independent blinded evaluators (abnormalities in hair shaft structure and skin surface were to be described). The results were unblinded, and the evaluated trichoscopic features were assigned to the appropriate patient group.

The occurrence of the scored trichoscopic criteria within each group (alopecia areata, trichotillomania, tinea capitis and TTA) was evaluated by respective Z tests. To prevent alpha inflation in repeated tests, the significance level was adjusted according to Bonferroni ($\alpha = 0.05/n$) and set to $p < 0.001$.

Results

Identified trichoscopic abnormalities in the evaluated patient groups included broken hairs, coiled hairs, upright regrowing hairs, exclamation mark hairs, tapered hairs, flame hairs, tulip hairs, v-sign, hook hairs, black dots, yellow dots, regrowing pigtail hairs (circular or oval), hypopigmented vellus hairs, comma hairs, corkscrew hairs, zigzag hairs, upright regrowing hairs and radially arranged hair bulbs at the border of alopecia lesions.

Color version available online

Fig. 1. ACC (×20). At the edge of patchy alopecia radially arranged hair bulbs are visible. These hair bulbs are directed into the middle of the lesion, with dark pigmented proximal ends (typical for the anagen phase). There are no follicular openings at the center of the lesion, but a vascular network resembling a spider's web is seen.

In all ACC lesions ($n = 5$), trichoscopy revealed hair bulbs visible through the semitranslucent epidermis. All were elongated, with very dark proximal ends (typical for anagen bulbs observed under the microscope). All bulbs were seen at the hair-bearing margin and were arranged radially. The central parts of the lesions showed no follicular openings. However, prominent vasculature was observed, which corresponds to skin atrophy in these areas (fig. 1). This finding was highly specific for ACC and was not found in other diseases.

Broken hairs as a feature of trichotillomania were found in 100% (6/6) of cases. However, they were also observed in 5% (5/102) of alopecia areata and 87% (7/8) of tinea capitis lesions. Coiled hairs were observed in 33% (2/6) of trichotillomania and 12.5% (1/8) of tinea capitis lesions. There were no cases of coiled hairs found in other groups ($p = 0.005$). Flame hairs (wavy and cone-shaped hair residues) were specific for trichotillomania (67%). Other characteristic features of trichotillomania included v-sign (33%) and tulip hairs (50%). V-sign (2%) and tulip hairs (2%) were also found in alopecia areata patches ($p < 0.001$) (fig. 2).

Yellow dots (37%; 37/102), black dots (42%; 43/102) and exclamation mark hairs (36%; 37/102) were all prevalent in alopecia areata lesions. Micro-exclamation mark hairs were also found in trichotillomania lesions (17%; 1/6), but were not present in tinea capitis, triangular alopecia and ACC. Tapered hairs were found only in alopecia areata patches (10%; 10/102). Hair regrowth was observed in the majority of alopecia areata patches in children – upright regrowing hairs (44%), circular hairs (23%) and vellus hairs (20%) (fig. 3).

Fig. 2. Trichotillomania (×20). Specific features of trichotillomania can be seen: irregularly broken hairs, coiled hairs, v-sign and circle hairs.

Fig. 4. TTA (×20). Short upright regrowing hairs cover the area of alopecia. Terminal hairs on the outskirts of the lesion can be seen.

Fig. 3. Alopecia areata (×20). Exclamation mark hairs and coiled hairs are visible.

Tinea capitis lesions showed specific trichoscopic features – comma hairs (50%), corkscrew hairs (12.5%) and zigzag hairs (50%). As mentioned above, broken hairs (87%), coiled hairs (12.5%) and regrowing pigtail hairs (12.5%) were also observed, but were not specific for this diagnosis.

In cases of TTA, upright regrowing hairs (100%; 3/3), vellus hairs (100%; 3/3) and regrowing pigtail hairs (33%; 1/3) were observed inside the patch and surrounded by terminal hairs on the outskirts of the lesions (fig. 4).

Detailed results are shown in table 1. Key trichoscopic findings are summarized in figure 5.

Discussion

Focal hair loss in children is one of the most common types of hair loss, yet establishing the correct diagnosis may be difficult. Alopecia areata is the most common diagnosis in such cases, though other rare diseases leading to patchy alopecia may be easily missed. Differential diagnosis includes nevus sebaceous (slight erythematous appearance at birth, mimicking superficial erosion), ACC, tinea capitis, triangular alopecia and trichotillomania. Trichoscopy as an easy and noninvasive method can be readily applied to diagnose patchy hair loss during childhood [1].

The limitation of this study lies in the absence of patients with nevus sebaceous.

Alopecia areata patches in children show the same trichoscopic features as those found in adult patients (black dots, exclamation mark hairs and yellow dots) [1, 10]. The study shows a prevalence of regrowing hairs in alopecia areata patches in children – upright regrowing hairs (44%), circle hairs (23%) and vellus hairs (20%). At the time of writing this paper, there are no data on the prevalence of regrowing hairs in adults, although it does seem that regrowing hairs are more extensive in children (own unpublished observations).

Although not as prevalent as in adolescents, trichotillomania does occur in children under 6 years of age. Previously published trichoscopic characteristic features (flame hairs, coiled hairs, hook hairs, irregularly broken

Table 1. Trichoscopic features found in trichotillomania, alopecia areata and tinea capitis

	TM (n = 6)	AA (n = 102)	TC (n = 8)	TTA (n = 3)	ACC (n = 5)
Broken hairs	6/6 (100%)	5/102 (5%)	7/8 (87%)	0	0
Coiled hairs	2/6 (33%)	0/0	1/8 (12.5%)	0	0
Upright regrowing hairs (<3 mm)	1/6 (17%)	45/102 (44%)	0	3/3 (100%)	0
Micro-exclamation mark hairs	1/6 (17%)	37/102 (36%)	0	0	0
Tapered hairs	0	10/102 (10%)	0	0	0
Flame hairs	4/6 (67%)	0	0	0	0
Tulip hairs	3/6 (50%)	2/102 (2%)	0	0	0
V-sign	2/6 (33%)	2/102 (2%)	0	0	0
Comma hairs	0	0	4/8 (50%)	0	0
Zigzag hairs	0	0	4/8 (50%)	0	0
Vellus hairs	0	20/102 (20%)	0	3/3 (100%)	0
Upright regrowing hairs	2/6 (33%)	25/102 (25%)	0	0	0
Regrowing pigtail hairs (circular or oval)	2/6 (33%)	23/102 (23%)	1/8 (12.5%)	1/3 (33%)	0
Black dots	2/6 (33%)	43/102 (42%)	2/8 (25%)	0	0
Yellow dots	0/6	37/102 (37%)	0/0	0	0
Corkscrew hairs	0	0	1/8 (12.5%)	0	0
Radially arranged hair bulbs	0	0	0	0	5/5 (100%)

TM = Trichotillomania; AA = alopecia areata; TC = tinea capitis.

hairs, v-sign and tulip hairs) were found in our study. The same is true for adult patients with trichotillomania, although flame hairs are more common in early childhood (67 vs. 25–30%) [4, 5].

Tinea capitis (with the presence of comma hairs, zigzag hairs or corkscrew hairs) is easily recognized by trichoscopy. Our findings are consistent with previous papers [1, 6, 11, 12].

TTA is of unknown origin. Its incidence has been estimated at 0.11% [13]. The majority of cases present between 2 and 6 years of age with a unilateral alopecia patch in the frontotemporal region. It is suspected that one third of TTA patients are born with this condition. The other two thirds develop it in the first 2–3 years of life. Histopathology shows a normal number of follicles, with a predominance of vellus hairs and rare terminal hairs on the superficial dermis [14].

In agreement with previous papers, our dermoscopic findings included short hairs covering the whole area of alopecia, as well as terminal hairs on the outskirts of the lesion [9]. The short hairs were recognized as vellus hypopigmented hairs, short upright regrowing hairs and circle hairs. The diversity of thin short hairs in TTA cases has been recently described [15].

ACC is the congenital focal absence of the epidermis and dermis. Clinically, one or more sharply circumscribed ulcers or scars can be found, most commonly on

the scalp [16]. Solitary ACC is the most common presentation, and 86% of all solitary ACC cases involve the scalp. At birth, ulcerations can be shallow or deep with a complete absence of all layers of the skin, in some cases extending to the dura or bone. In other cases, a healed scar might be the only finding. Over the course of a few months, the ulcerated lesions heal spontaneously from the periphery, leaving a smooth, yellowish atrophic area of cicatricial alopecia [16, 17].

The incidence of ACC is estimated to be between 0.5 and 1/100,000 newborns [18]. Most ACC lesions are small, superficial and clinically present as patches of focal alopecia. In the event of any accompanying abnormalities, a proper diagnosis may be easily missed. Clinically, two different types of ACC may be distinguished. It has been proposed that membranous ACC of the scalp is due to an incomplete closure of ectodermal fusion lines. Lesions in these cases have a membranous covering that may be filled with fluid, giving the lesion a bullous appearance. The existence of a hair collar suggests the presence of ectopic neural tissue [19]. Histologic evaluation of the bullous or membranous ACC reveals fibrovascular and/or edematous stroma, similar to the histopathological appearance of encephaloceles or meningoceles [20]. A second type of ACC, nonmembranous scalp ACC, has been hypothesized to be due to a tension-induced disruption of the skin where tensile forces are greatest during

Diagnosis	Key trichoscopic findings
Alopecia areata	Exclamation mark hairs Black dots Yellow dots Circle hairs
Tinea capitis	Comma hairs Corkscrew hairs Zigzag hairs
TTA	Upright regrowing hairs, vellus hairs and regrowing pigtail hairs inside the patch, surrounded by terminal hairs on the outskirts of the lesion
Trichotillomania	Flame hairs Broken hairs V-sign Tulip hairs
ACC	Elongated hair bulbs visible through the semitranslucent epidermis, seen at the hair-bearing margin and radially arranged

Fig. 5. Key trichoscopic findings for patchy alopecia (alopecia areata, tinea capitis, ACC, TTA and trichotillomania) in pediatric trichology.

brain development. This may explain why it occurs in the vertex area. The defect may be secondary to vascular disruption or biomechanical stretch. Familial ACC is generally of the nonmembranous type, whereas membranous ACC is usually sporadic [19].

Histologic examination of nonbullous ACC of the scalp shows a layer of thin dermal collagen without overlying epithelium or adnexal structures. Histology of the involved areas shows a gradual progression from normal skin to rudimentary and small dermal appendages such as hair papillae, sebaceous glands and sweat glands. Within the defective area itself, there are no elastic fibers, no normal blood vessels and no dermal papillae. After several weeks, the epidermis may appear flattened, and there may be a proliferation of fibroblasts within the connective tissue stroma [19, 21].

In this article, we present a trichoscopic picture of nonbullous type ACC, with erosions healed in utero. In all ACC patches, a unique trichoscopic picture was shown, with visible hair bulbs arranged radially along the hair-bearing margins of patchy alopecia. Although one report does indicate that ACC trichoscopy is characterized by a complete lack of skin appendages and translucent appearance, it does not describe the presence of dark anagen bulbs radially distributed around the alopecia patch [8].

Conclusion and Message of the Paper

Trichoscopy is a useful technique for the differential diagnosis of patchy alopecia in children. A novel finding in this study indicates that radially arranged hair bulbs visible through the translucent epidermis are characteristic of nonbullous type ACC. This knowledge can help clinicians perform a quick and painless diagnosis of ACC in newborns. In such cases, ocular and neurological anomalies should be investigated. In alopecia areata, regrowing hairs (vellus hairs, upright regrowing hairs and circle hairs) appear to be more prevalent in children than in adults. TTA lacks specific trichoscopic features, but the presence of short hairs (vellus hairs, upright regrowing hairs and coiled hairs) on the patch are sufficient to establish a proper diagnosis.

Statement of Ethics

The study was approved by the local ethics committee.

Disclosure Statement

The authors have no conflicts of interest to disclose.

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