

Dermoscopy as a Diagnostic and Evaluation Tools in Childhood Alopecia Totalis

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Abstract: Alopecia totalis is a manifestation of alopecia areata that is characterized by total scalp hair loss and have a higher risk for poor prognosis and treatment failure. Early-onset, nail involvement, history of atopy, and long duration of disease are associated with poor prognosis. Total scalp hair loss in alopecia totalis might have a sudden onset or following partial alopecia. In the case of diffuse scalp hair loss, the clinical finding might be similar to telogen effluvium and trichotillomania. Hence, a biopsy is required to establish the proper diagnosis. However, this invasive diagnostic method is not favorable for most patients, especially children. Several studies have been done to show the reliability of dermoscopy to diagnose alopecia areata. Some dermoscopic features, such as yellow dots, black dots, broken hairs, tapering hair (exclamation marks), and short vellus hairs, are known as characteristic findings in this disease. Sign of hair regrowth might be detected earlier with this technique, and even it is not yet visible with naked eyes. We are reporting an 8-year-old girl with a history of recurrent total scalp hair loss for four years and clinical findings of diffuse hair loss on smooth surface scalp skin, nail pitting, and transverse leukonychia. Initial dermoscopy evaluation revealed characteristic findings of alopecia areata and regrew of short vellus hair was detected earlier with this examination. Therefore, we would like to report the use of dermoscopy as a diagnostic and evaluation tools in childhood alopecia totalis.

1 INTRODUCTION

Alopecia areata is an autoimmune disease with nonscarring hair loss that affects any hair-bearing areas (Alkhalifah et al, 2010). However, the pathogenesis is not known, and prevalence of this disease varied from 0.7 – 3.8% around the globe with similar gender proportion (Wasserman et al, 2007). Most patients have the onset of the first lesion before 20 years old and 20% of cases found in childhood (Nanda et al, 2002). Clinical manifestation may vary from single patch to multiple or diffuse hair loss. In severe cases, the disease can extend to total scalp hair loss or known as alopecia totalis, and even including body hair in alopecia universalis (Alkhalifah et al, 2010). Around 5% of alopecia cases evolve into these long forms and associated with poor prognosis, treatment failure, and high relapse rate (Alkhalifah et al, 2010; Jang et al, 2017). Other prognostic factors such as early-onset

at a young age, long duration, ophiasis pattern, nail abnormalities, history of atopy, other autoimmune diseases, and family member are also related to a poor outcome (Alkhalifah et al, 2010; Olsen, 2011). In general, clinical diagnosis of alopecia areata is made based on the typical pattern of hair loss with the presence of characteristic exclamation mark hair (Alkhalifah et al, 2010). However, in some cases, the clinical diagnosis may not be straightforward where biopsy evaluation may be required to confirm the diagnosis, but this invasive technique is not favored by most patients, especially children. In the case of alopecia totalis with diffuse or total scalp hair loss, dermoscopy examination might be necessary to exclude some differential diagnosis such as telogen effluvium and trichotillomania.

Dermoscopy is a noninvasive diagnostic method that allows evaluation of microstructures of the epidermis, the dermo-epidermal junction, and the papillary dermis which are not visible to the naked

eye (Mane et al, 2011). Characteristic dermoscopic features of alopecia areata are yellow dots, black dots, broken hairs, tapering hair (exclamation marks), and short vellus hairs (Mane et al, 2011; Inui et al, 2008). In the remission phase, the white perihilar sign might be found, and short vellus hair becomes more prominent (Jha et al, 2017). However, the proportion of these diagnostic features might vary according to the literature. Therefore, we would like to report the use of dermoscopy as a diagnostic and evaluation tool in the case of childhood alopecia totalis.

2 CASE

An 8-year-old girl came with a chief complaint of diffuse scalp hair loss since the age of 4. Approximately four years ago, parents incidentally found a single hair loss patch on the scalp without any associated symptoms. The year after, the patient developed total scalp hair loss following an episode of high fever and upper respiratory tract infection. Eyebrows, eyelashes, and other hair-bearing areas were not involved. Eventually, hair regrowth observed after resolution of fever and infection. However, relapses were noted at least twice a year, following episodes of high heat and tonsillopharyngitis. The parents denied any application of topical or oral medications before the onset of disease. The patient has a history of rhinitis allergic (atopy). However, no prior history of autoimmune diseases and a family history of hair loss were noted. Parents brought her to a dermatologist, and unknown topical medications were given with good response. However, two months prior to a consultation, the patient developed a recurrent episode of total scalp hair loss three weeks after hospital admission due to typhoid fever. Even hair regrow already been noted, the frequent relapses and lack of self-confidence prompted this consult.

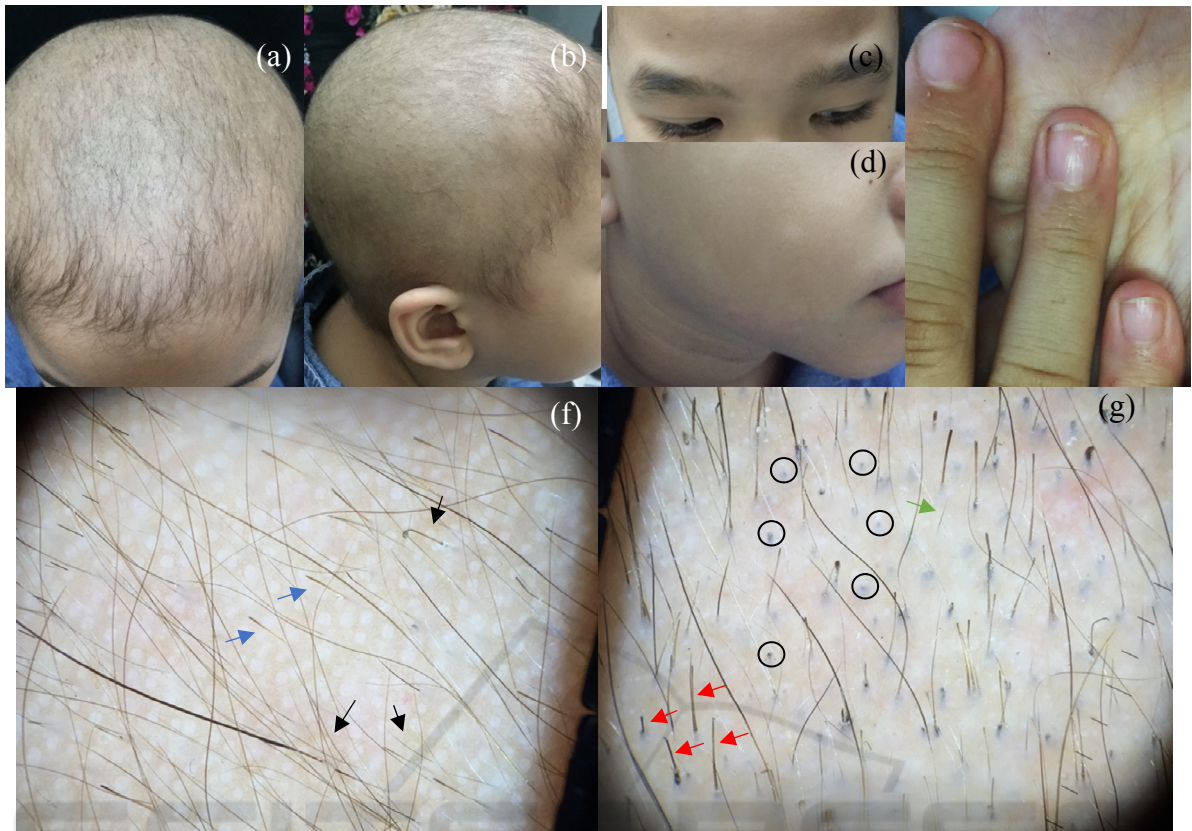
In physical examination, the patient was in good general condition and nutritional status. All of the vital signs were within the normal range. Dermatologic examination revealed skin-colored smooth surface macule with diffuse hair loss, decreased of terminal hairs, and predominance of vellus and broken hairs on the scalp. Furthermore, multiple discrete lenticular hypopigmented macules with indistinct borders and fine scales also observed on the face. Small superficial pitting and transverse leukonychia on fingernails were also noted. Eyebrows and eyelashes were intact (Figure 1a – e).

From the initial assessment, the differential diagnosis for the hair loss, in this case, are alopecia totalis, telogen effluvium, and trichotillomania. Wood's lamp examination on hypopigmented facial lesions did not show any enhancement and skin scraping with KOH 10% also failed to show fungal elements. These findings are compatible with pityriasis alba. Serology examination showed typical results for ANA, anti-dsDNA, free T3, T4, TSH, which ruled out possible associated autoimmune conditions such as lupus, and thyroid disease. Initial dermoscopy evaluation on the scalp revealed yellow dots, black dots, exclamation hairs, broken hairs, and short vellus hairs (Figures 1f – g). Based on these findings, the diagnosis of alopecia totalis was established.

Hence, the patient was given topical minoxidil 2% solution applied once a day. On follow up visit two months after treatment initiation, scalp hair has regrown with 3 – 4 cm length in most areas. The dermoscopic examination also showed signs of hair regrowth with an increasing proportion of terminal hairs and significant reduction of broken and short vellus hair. Exclamation hair, yellow dots, and black dots were not seen anymore (Figures 2). Therefore, the patient was advised to continue the medications until the scalp hair fully regrows with proper compliance, do regular monthly visit, and avoid triggering factors such as fever and infection.

3 DISCUSSION

Alopecia totalis is a manifestation of alopecia areata that is characterized by total scalp hair loss and associated poor prognosis and treatment failure with only less than 10% of patients achieved complete resolution. (Jang et al., 2017) Only limited data are available regarding alopecia areata in children. Prevalence of childhood alopecia is around 11.1% in a multiethnic community of Singapore. Even though there is a slight male predominance, the proportion of severe alopecia is found higher in female patients. In the case of extensive alopecia, such as alopecia totalis and universalis, the onset usually is more than six months, and most of them have recurrent episodes. (Tan et al., 2002) As an autoimmune disease with multifactorial etiologies, alopecia areata can be triggered by microtrauma or destruction of hair follicles, bacteria or virus infection, and emotional stress. (Ito, 2003) Clinical manifestation of alopecia totalis commonly found as sudden onset of asymptomatic total scalp hair loss on the healthy and smooth skin surface with the characteristic



Figures 1. Dermatologic examination revealed (a), (b) diffuse scalp hair loss with decreased of terminal hairs and predominance of vellus and broken hairs, (c) intact eyebrows and eyelashes, (d) multiple hypopigmented macules with fine scales on the face which are consistent with pityriasis alba, and (e) multiple small superficial pitting and transverse leukonychia on fingernails. (f), (g) Initial dermoscopic examination showed yellow dots (black arrow), exclamation hairs (blue arrow), black dots (black circle), broken hairs (red arrow), and short vellus hairs (green arrow).



Figures 2. Follow up visit two months after therapy initiation, (a), (b) hair regrowth was observed in most areas of the scalp. (c) The dermoscopic evaluation also revealed significantly increased of terminal hairs, reduction of broken and short vellus hairs, but exclamation hairs, yellow dots, and black dots not seen.

exclamation hairs and sparing of white hairs. In some cases, the onset might be following episodes of partial alopecia. (Alkhalifah et al, 2010; Otberg et al., 2012) Nail involvement found in 7 – 66% of cases, with small superficial pitting as the most common finding, followed by trachyonychia, Beau's lines, onychorrhexis, thinning or thickening of nails, onychomadesis, koilonychia, punctate or transverse leukonychia, and red lunulae. (Alkhalifah et al, 2010) In childhood alopecia, 8.4% of patients developed nail abnormalities, including pitting, trachyonychia, and longitudinal ridging, which correlated with the severity of the disease. (Tan et al., 2002) Furthermore, history of atopy, autoimmune diseases such as vitiligo, thyroid disease, lupus, and other conditions are also associated with a higher incidence of extensive alopecia (totalis and universalis). (Alkhalifah et al, 2010)

Diagnosis of alopecia areata usually mostly made based on clinical findings, and ancillary tests might not be needed to confirm the diagnosis. However, in some cases with diffuse hair loss, as seen in our patients, the clinical appearance might be similar to telogen effluvium and trichotillomania. (Alkhalifah et al, 2010) Hence, histopathology evaluation from a scalp biopsy is required to rule out other etiology. The common histopathological findings are generalized miniaturization, a marked increase in the catagen and telogen hair follicles and peribulbar lymphocytic infiltrate (a swarm of bees) as the hallmark of the acute phase. These features help the physician to confirm the diagnosis of alopecia areata.¹¹

However, this invasive diagnostic method is not favorable or routinely done, especially in children. Therefore, there are many studies have done lately to verify the diagnostic features of alopecia areata in dermoscopy. In the literature, characteristic dermoscopic findings of alopecia areata are yellow dots, black dots, broken hairs, exclamation mark hair, and short vellus hairs (Table 1). (Mane et al., 2011; Inui et al., 2008; Mahmoudi et al., 2018) Among these features, yellow dots can be regarded as a sensitive marker for alopecia areata due to its high frequency. (Jha et al., 2017) According to Inui et al., yellow dots and short vellus hairs were the most sensitive markers for the diagnosis, and black dots, exclamation mark hairs, and broken hairs were the most specific markers. (Inui et al., 2008) Some studies have tried to correlate these dermoscopic findings with disease activity and severity. Guttikonda et al. found that among those features, black dots, broken hairs, and exclamation mark hairs are correlated with disease activity. (Guttikonda et al., 2016) Furthermore, black dots and yellow dots correlated positively with the severity of the alopecia areata. (Inui et al., 2008; Guttikonda et al., 2016)

On the other hand, short vellus hairs correlated negatively with either disease activity or severity. This feature is commonly found in patients under treatment and indicates an early sign of disease remission. Regrowth of short vellus hair after surgery can be seen in dermoscopy even before they can be perceived by the naked eye. (Guttikonda et al., 2016)

Table 1. Dermoscopic features of alopecia areata

Findings	Definition	Frequency
Yellow dots	Round or polycyclic yellow to yellow-pink dots that represent distended follicular infundibula filled with sebum and keratin remnants	63,7 – 89,6%
Black dots	The remnant of broken hair shafts inside follicular ostia	40,9 – 78,4%
Exclamation mark hair	Broken hairs that tapered toward follicles (diameter of proximal hair follicle < distal)	31 – 66,6%
Short vellus hair	Thin, unpigmented hairs with length ≤10 mm may demonstrate early disease remission	12,1 – 31,7%
Broken hair	Fracture of dystrophic hair shafts or rapid regrowth of hairs that formerly manifested as black dots	9,5 – 55,4%

(references: Mahmoudi H, Salehi M, Moghadas S, Ghandi N, Teimourpour A, Daneshpazhooh M. Dermoscopic findings in 126 patients with alopecia areata: A cross-sectional study. International Journal of Trichology 2018;10(3):118-23.)

In contrast, other condition with prolonged diffuse hair loss such as chronic telogen effluvium did not show specific dermoscopic findings. Diagnosis of telogen effluvium may be suspected when empty hair follicles (sometimes appearing as yellow dots) and short, dark, regrowing hairs with standard thickness are present in the absence of the

characteristic features of other scalp disorders. In trichotillomania, the presence of broken hair shafts with different lengths and signs of plucking, such as a bleeding point on the scalp may help to establish the diagnosis. These findings are not usually seen in alopecia areata and remain as the hallmark of this psychiatric disorder. (Lacarrubba et al., 2015)

Treatment of childhood alopecia areata has to be pro-active due to its chronicity and high risk of extensive involvement. However, only limited data are available regarding treatment options in this population. Topical medications such as corticosteroid, minoxidil, anthralin, and immunotherapy (diphenylcyclopropenone (DPCP), squaric acid dibutyl ester (SADBE)) remains as first-line therapy. The physician might use a combination of 2 or 3 topicals as second-line therapy. Inadequate data are available to support the use of systemic treatment in childhood alopecia to prevent extensive alopecia. Therefore, systemic medications remain the last frontier in treatment options.(Cranwell et al., 2018).

In our case, we found an 8-year-old girl with a 4-year history of multiple episodes of sudden onset total scalp hair loss which were triggered by fever and infection. Nail abnormalities such as small superficial pitting, transverse leukonychia, and history of atopy (rhinitis allergic) and also a minor feature of atopic dermatitis (pityriasis alba) are found in this patient. Clinical findings of diffuse scalp hair loss can be found not only in alopecia totalis but also telogen effluvium and trichotillomania. However, diagnosis of alopecia totalis was established by characteristic dermoscopic findings of yellow dots, black dots, exclamation hairs, broken hairs, and short vellus hairs. Hence, telogen effluvium could be ruled out. Furthermore, trichotillomania was ruled out due to no signs of a bleeding point on the scalp, and all the broken hairs were within a similar length.

Sensitive markers for disease severity, black dots, and yellow dots, are found in the initial dermoscopy evaluation, which is compatible with the clinical manifestation of extensive alopecia totalis. Hence, our patient was immediately started on topical minoxidil solution as the first-line therapy in childhood alopecia. Within two months, scalp hair has regrown in most areas and dermoscopic markers for disease activity, including black dots, broken hairs and exclamation mark hairs, were not found anymore. Short vellus hairs, as the sign of remission phase in alopecia, were found as the dominant dermoscopic findings in the follow-up visit and indicated good treatment response. These findings are similar to several studies done previously and prove the reliability of dermoscopy, not only as a diagnostic tool but also for evaluation of treatment.

4 CONCLUSION

Alopecia totalis in children is associated with long duration, a severe progression of the disease, and inadequate response to treatment. In the case with diffuse scalp hair loss, the clinical findings might not lead to a straightforward diagnosis of alopecia totalis. Dermoscopy has been used to confirm a diagnosis in which biopsy may not be visible. Black dots, broken hairs, and exclamation mark hairs are correlated with disease activity. While short vellus hairs might be the early sign of remission, even the hair regrowth sign has not been visible with naked eyes. Therefore, dermoscopy is an advantageous and practical non-invasive diagnostic and evaluation method in childhood alopecia totalis.

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