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## Case and Review

# Linear and Annular Lupus Panniculitis of the Scalp: Case Report with Emphasis on Trichoscopic Findings and Review of the Literature

Siriorn Udompanich Kumutnart Chanprapaph Poonkiat Suchonwanit

Division of Dermatology, Faculty of Medicine, Ramathibodi Hospital, Mahidol University, Bangkok, Thailand

## Keywords

Alopecia · Blaschko's line · Hair loss · Linear lupus · Lupus panniculitis

## Abstract

Linear and annular lupus panniculitis of the scalp (LALPS) is a unique subset of lupus panniculitis, which results in non-scarring alopecia along the Blaschko line of the scalp in an otherwise healthy young patient. Numerous cases have been reported around the world, but data on their trichoscopic findings and correlations with the underlying pathology is sparse. We hereby present a case of 23-year-old male with LALPS and further describe his trichoscopic findings as well as their correlations with histopathological features.

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## Introduction

Lupus panniculitis contributes to 1–3% of all specific cutaneous lupus erythematosus (LE) cases and presents with indurated erythematous subcutaneous nodule or plaque. It

typically appears on the face, proximal extremities, and on the trunk [1–3]. Histopathological sections reveal lobular panniculitis with predominant lymphocytic infiltration, accompanied by hyaline fat necrosis [4]. The scalp can be involved in up to 16.7% of the cases [2]. Lesions on the scalp either share similar characteristics with classic lupus panniculitis, with indurated erythematous plaque resulting in scarring alopecia, or they take a distinct form that is non-scarring and follows the Blaschko lines of the scalp [5]. The latter was first described by Nagai et al. [6] in 2003. Since then, similar cases have been increasingly recognized, confirming its unique presentation and clinical course. The condition was later termed linear lupus panniculitis of the scalp or linear and annular lupus panniculitis of the scalp (LALPS) [5]. Nevertheless, there are limited data on the trichoscopic findings and correlations with the underlying pathology. We hereby present a case of 23-year-old male with LALPS and further describe his trichoscopic findings as well as their correlations with histopathological features.

### Case Presentation

A 23-year-old male presented with localized hair loss on his right parietal scalp. The lesion was asymptomatic and gradually expanded over the course of 5 months. The patient was otherwise well with no known underlying pathology or any history suggesting systemic involvement, such as joint pain and photosensitive rash. On examination, there was a non-scarring alopecia patch, approximately 8 cm in diameter, with bizarre configuration and faint erythema on the right parietal scalp and without any induration or atrophy (Fig. 1). The hair appeared normal; both hair pull and rolling tests were negative. His complete blood count, blood urea nitrogen, serum creatinine, liver function tests, urinalysis, and venereal disease research laboratory test were all within normal limits, while a speckled-pattern antinuclear antibody was detected with a titer of 1:160.

Trichoscopic examination revealed several hair shaft abnormalities, including broken hair, exclamation mark hair, angulated hair, and short regrowing hair (Fig. 2a). Scalp changes were also evident. The erythema on the interfollicular areas turned out to be prominent blood vessels (Fig. 2b). Some hairs were broken inside the follicles at the scalp level and appeared as black dots. The sparse yellow dots were displayed in variable sizes, with some larger than others (Fig. 2b). On histopathology, focal interface changes with vacuolar alteration and occasional necrosis of basal keratinocytes were detected. The dense perivascular, perifollicular, and interstitial inflammatory cell infiltrates of mainly lymphocytes were found within the dermis, with dense nodular infiltration in the fat lobules of the subcutaneous tissue, along with some hyalinized necrosis of fat cells (Fig. 3a). A close examination at the uppermost level of the horizontal section showed perifollicular infiltration, fibroplasia, as well as dilatation of the follicles and keratin plugging (Fig. 3b).

From his clinical presentation and supporting histopathological findings, we arrived at the diagnosis of LALPS. In accordance with previously reported cases of LALPS, several treatments, including hydroxychloroquine, corticosteroids (systemic, intralesional, or topical), methotrexate, mycophenolate mofetil, cyclophosphamide, dapsone, gold therapy, and topical minoxidil were found to be effective. The patient was given oral hydroxychloroquine 200 mg once daily and 0.05% clobetasol propionate solution to apply on the lesion twice daily. Almost

complete hair regrowth was achieved within 3 months. There has been no recurrence during the follow-up period of 1 year.

## Discussion

LALPS is a distinct type of lupus panniculitis that follows the Blaschko line. [Table 1](#) and [Table 2](#) summarize all case reports that are available in full text. A majority of patients were young East Asian males, but females and Caucasians were also affected. All cases achieved significant improvement or complete hair regrowth after the administration of oral prednisolone or hydroxychloroquine, with very few recurrences. While antinuclear antibodies were positive in almost half of the patients, only 1 fulfilled the criteria for systemic LE (SLE). Chen et al. [7] compared LALPS to classic lupus panniculitis and concluded that LALPS occurs at a younger age, predominantly in East Asian males, has a reversible clinical course, and fewer associations with SLE.

The true pathomechanism of LALPS is largely unknown. The most striking features that discriminate this condition from classic lupus panniculitis are its non-scarring nature and distribution along the Blaschko line. It was speculated that the inflammation is confined to the subcutaneous layer; hence, the stem cells in the hair bulge are well preserved, allowing spontaneous hair regrowth once the inflammation has subsided. Alternatively, the condition may have appeared to be non-scarring because it was treated early. We suspect that if the inflammation persists and expands upward, the hair bulge can be involved, leading to subsequent scarring alopecia. The horizontal section from our patient revealed signs of early fibrosis at the hair follicles, which supports this hypothesis. The Blaschko line predilection is more challenging to explain. Some believe the lesion is caused by genetically programmed cloning of cells along the Blaschko lines from early embryogenesis [8]. A recent review by Luengarun et al. [9] suggested that the disease mostly occurs on the parietal scalp (70%) and does not always follow the Blaschko line distribution. However, the exact pattern of Blaschko's line on the head and neck remains to be established and may vary between individuals.

Our case offers further insight into the trichoscopic features of LALPS and demonstrates some trichoscopic and histopathological correlations. Numerous signs have indicated insult to the hair shaft. Angulated hairs are fracture hairs with a sharp angle along the hair shaft [10]. Complete fracture of the hair above the follicles is seen as broken hair and at the follicular level as black dots, while exclamation mark hair suggests a slow progressive insult at the level of hair formation. All these signs may be explained by the nodular perifollicular infiltration with lymphocytic predominance on histopathology, leading to destruction of the lower part of the hair. The broken hair, black dots, angulated hair, and especially exclamation mark hair, are hallmarks of alopecia areata (AA), in which dense nodular follicular infiltration or "swarm of bees" is also the main histopathological feature [10–12]. Moreover, the keratin plugs, which are follicular hyperkeratosis on histopathology, are consistent with large yellow dots on trichoscopy.

Differential diagnosis of LALPS includes conditions with localized non-scarring alopecia such as AA, trichotillomania, and syphilitic alopecia [5]. Broken hair, black dots, and yellow dots are all features of these disorders [11]; however, certain clues could point towards LALPS. The size of the blood vessels was noted to be larger in cutaneous LE when compared

to those of AA. The yellow dots also appeared to be larger and darker [13]. Thorough history taking and laboratory investigations may help exclude trichotillomania and syphilitic alopecia, and histopathology can always give a definite diagnosis. Another differential diagnosis is a further subtype of cutaneous LE on the scalp termed “non-scarring patchy alopecia in SLE.” It presents with localized non-scarring patches with similar trichoscopic features, especially regarding predominant blood vessels. However, the disorder is restricted to patients with SLE, often in cases with severe exacerbations, unlike LALPS where the risk of SLE is extremely low. The patches resemble AA, do not follow a bizarre distribution, or involve the subcutaneous tissue on histopathology [5, 14, 15].

In conclusion, we report a case of localized non-scarring alopecia along the suspected Blaschko line and biopsy results consistent with lupus panniculitis. Trichoscopic examination displayed hair shaft abnormalities, including broken hair, exclamation mark hair, angulated hair, and short regrowing hair. Furthermore, scalp changes such as prominent blood vessels and yellow dots were present. While most signs can be found in other conditions with non-scarring alopecia, prominent blood vessels should alert clinicians to the possibility of LALPS. Furthermore, continuing follow-up is recommended due to the possibility of LALPS recurrence and the development of SLE.

### Statement of Ethics

The patient provided written informed consent to perform all necessary investigations, to take clinical photographs, and to use them for research purposes and publication.

### Disclosure Statement

The authors have no conflicts of interest to declare.

### Author Contributions

All named authors meet the International Committee of Medical Journal Editors (ICMJE) criteria for authorship for the manuscript, take responsibility for the integrity of the work as a whole, and gave final approval to the version to be published.

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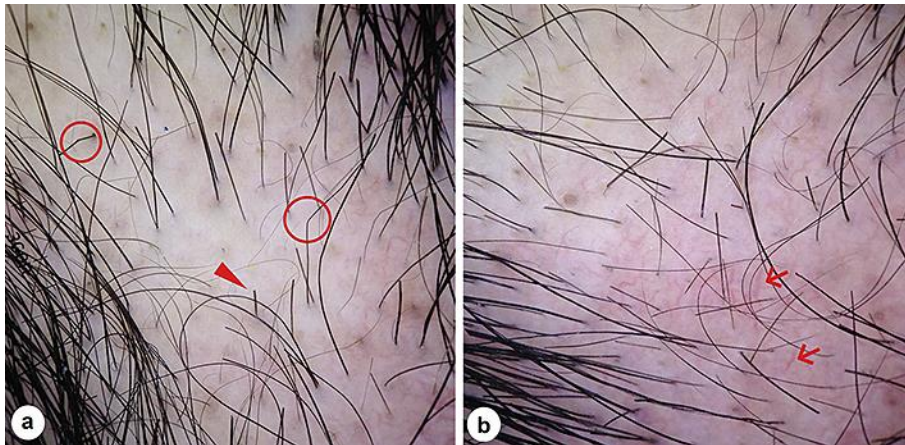
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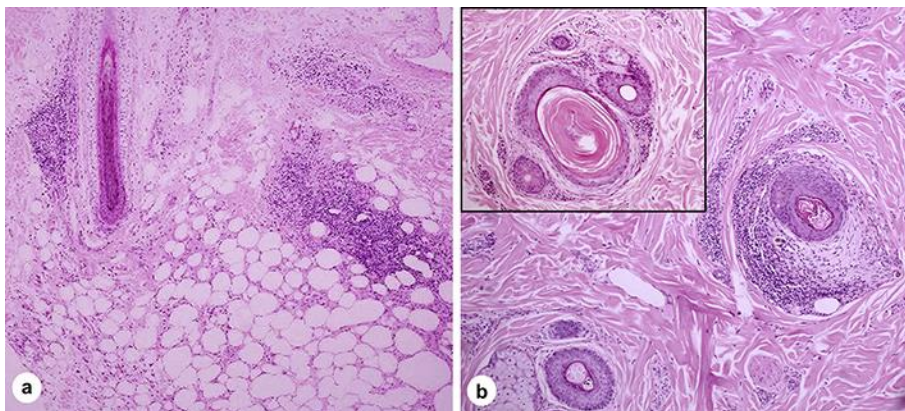


**Fig. 1.** Localized non-scarring alopecia with bizarre distribution on the right parietal scalp.





**Fig. 2.** Trichoscopic features of LALPS (magnification  $\times 20$ ). **a** Hair shaft abnormalities: broken hair, exclamation mark hair (arrowhead), angulated hair (circles), and short regrowing hair. **b** Scalp changes: prominent blood vessels (arrows) and yellow dots of variable sizes.



**Fig. 3.** Histopathologic features of LALPS. **a** Dense perivascular, perifollicular, and interstitial lymphocytic infiltration in the dermis and lobular panniculitis in the subcutaneous tissue (hematoxylin-eosin,  $\times 40$ ), with some hyalinized necrosis of fat cells (hematoxylin-eosin,  $\times 100$ ). **b** Close examination at the uppermost level of the horizontal section (hematoxylin-eosin,  $\times 100$ ) showing perifollicular infiltration, fibroplasia, dilatation of the follicles, and keratin plugging (inset).

**Table 1.** Available case reports of linear and annular lupus panniculitis of the scalp

First author, year [ref.]	Age of onset, years/sex	Race/ethnicity	Clinical presentation	Duration of disease	Histology	Direct immunofluorescence	Evidence of systemic involvement	Treatment and outcome
Nagai, 2003 [6]	10/female	Japanese	Linear and arc-shaped alopecia on right and left temporal scalp with band-like erythema	2 months	Normal epidermis Perivascular and periadnexal lymphocytic infiltration Fat degeneration Abundant mucin	n.a.	ANA positive 1:320 homogenous and speckled pattern	Topical steroid No improvement
Wu, 2004 [16]	21/female	Taiwanese	Spiral-shaped non-scarring alopecia on the frontal and parietal areas along the Blaschko line	1 year	Normal epidermis Perieccrine aggregation of plasma cells and lymphocytes in the dermosubcutaneous junction Fat degeneration, abundant deposition of mucoid material through the dermis and the fat tissue	n.a.	Blood tests all normal	HCQ Complete resolution in one year
Bacanli, 2005 [17]	16/female	n.a.	Annular erythematous plaque right side of scalp, scarring alopecia observed on the inner side of the plaque	2 years	Follicular hyalinization and perifollicular lymphocyte infiltration Lymphocytic panniculitis, hyaline degeneration of fat, hyaline papillary bodies, lymphocytic nodule	Positive C3 and IgM in follicular epithelium	ANA positive 1:320 homogenous and nucleolar pattern	HCQ Significant improvement of the lesion
Rhee, 2009 [18]	14/male	Korean	Linear non-scarring alopecia patch on occipital scalp	9 months	Epidermis: hyperkeratosis, follicular plugging Dermis: perifollicular lymphoid infiltration, increased catagen hair Subcutaneous tissue: dense mucin deposition, lymphoid cell infiltration	Negative for IgM, IgG, IgA, C3 and fibrinogen	n.a.	HCQ and prednisolone Hair regrowth from 8th week, complete remission
	32/male	Korean	Linear non-scarring alopecia patch on occipital scalp	2 years	Similar to case one but less perifollicular infiltration	Negative for IgM, IgG, IgA, C3 and fibrinogen	n.a.	Dapsone Hair regrowth (no F/U)
Chen, 2012 [7]	32/male	Taiwanese	Linear and wavy non-scarring alopecia on occipital scalp along Blaschko's line, faint interfollicular erythema and follicular plugging	6 months	Normal epidermis Dermis: peribulbar infiltration of lymphocytes and plasma cells Subcutaneous tissue: mucin deposition, lymphoplasmocytic infiltration	Negative for IgG, IgA, IgM and C3	Blood tests all normal	HCQ, IL steroid Complete hair regrowth but recurred after 9 months
Tsuzaka, 2012 [19]	26/male	Japanese	Multiple arc-shaped erythematous plaques with non-scarring alopecia	2 years	Dermis: perivascular lymphocytic infiltration Subcutaneous tissue: lobular and septal panniculitis, mucin deposition	Granular deposits of IgG along DEJ	ANA positive 1:1,280 speckled pattern Anti-Ro positive	Prednisolone Complete hair regrowth

n.a., data not available; ANA, antinuclear antibodies; HCQ, hydroxychloroquine; IL, intralesional; DEJ, dermo-epidermal junction; SLE, systemic lupus erythematosus.



**Table 2.** Available case reports of linear and annular lupus panniculitis of the scalp (continued)

First author, year [ref.]	Age of onset, years/sex	Race/ethnicity	Clinical presentation	Duration of disease	Histology	Direct immunofluorescence	Evidence of systemic involvement	Treatment and outcome
Pandhi, 2012 [20]	25/male	Indian	Annular non-scarring alopecic patch on frontal scalp	n.a.	Hyperkeratosis, follicular plugging, basal vacuolization Superficial and deep perivascular and perifollicular lymphocytic infiltration lobular panniculitis	Granular deposition of IgG, IgM, IgA and C3 along DEJ	Developed SLE 2 years later	Prednisolone, HCQ Improved but with partial scarring after 8 weeks of therapy
Chiesa-Fuxench, 2013 [21]	26/male	Caucasian	Arc-shaped linear patches of non-scarring alopecia over right parietal and occipital scalp along the line of Blaschko	n.a.	Superficial and deep lymphocytic infiltration with mucin deposition within the dermis and extension into the subcutaneous fat	n.a.	Blood tests all normal	Prednisolone, HCQ, IL steroid Complete hair regrowth
Mitxelena, 2013 [22]	34/male	Caucasian	Erythematous edematous annular and linear plaques of non-scarring alopecia on vertex and parietal scalp	1 year	Dense lymphocytic infiltrate affecting the deep dermis and adipose tissue, fat necrosis and vacuolar damage of the basal layer of the epidermis	Granular deposits of IgM along DEJ	ANA 1:320 Anti-Ro positive	HCQ, prednisolone Resolved but recurred after 1 year
Kiritsi, 2014 [23]	34/male	Caucasian	Linear non-scarring alopecia along Blaschko's line	1 year	Normal epidermis Accumulation of mucin in the dermis lymphocytic panniculitis	n.a.	ANA 1:200	HCQ Complete hair regrowth in 12 weeks
Kshetrimayum, 2016 [24]	17/male	Indian	Two linear non-scarring alopecic patches with mild erythema and tenderness on temporal and frontal scalp	7 years	Dermis: moderate perivascular lymphocytic infiltration Subcutaneous tissue: fat necrosis, myxoid degeneration and hyaline deposits in the interlobular septa	n.a.	Blood tests all normal	Prednisolone, HCQ Complete hair regrowth
Luengaran, 2017 [25]	28/male	Burmese	Linear non-scarring alopecia on vertex and parietal areas Hair pull negative	1 year	Epidermis and dermis: dense perifollicular lymphoid infiltration, vacuolar interface along infundibular epithelium, epidermal atrophy, follicular plugging Subcutaneous tissue: fat necrosis, mucin infiltration	n.a.	ANA 1:320 speckled pattern	Prednisolone, HCQ, IL steroid, topical 5% minoxidil lotion Remarkable improvement
Park, 2017 [26]	18/male	Korean	Arc-shaped, non-scarring alopecic patches on right frontal-parietal and left temporoparietal scalp	1 year	Dense lymphocytic infiltrates mainly in the peribulbar and interfollicular areas in lower dermis and subcutaneous tissue	n.a.	ANA 1:20 speckled pattern	HCQ Improved in 12 weeks
Present case	23/male	Thai	Non-scarring alopecic patch with bizarre configuration on the right parietal scalp	5 months	Epidermis: focal interface changes with vacuolar alteration and occasional necrosis of basal keratinocytes Dermis and subcutaneous tissue: dense perivascular, perifollicular, interstitial and subcutaneous tissue with some hyalinized necrosis of fat cells	n.a.	ANA 1:160 speckled pattern	HCQ, topical steroid Complete hair regrowth in 3 months

n.a., data not available; ANA, antinuclear antibodies; HCQ, hydroxychloroquine; IL, intralesional; DEJ, dermo-epidermal junction; SLE, systemic lupus erythematosus.