Case Report

Linear Cutaneous Lupus Erythematosus Following Blaschko's Lines on the Scalp: Additional Cases and Review of the Literature

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Abstract

Description

Alopecia of the scalp has various causes and presentations. However, linear alopecia is unusual and lupus erythematosus presenting as linear alopecia is exceedingly rare. To date, there have been 16 documented cases of linear alopecia diagnosed as chronic cutaneous lupus erythematosus occurring in a linear configuration following Blaschko's lines. We report 2 additional cases and review the clinical and histologic features along with treatment. This Blaschkoid linear variant of cutaneous lupus erythematosus has distinct clinical and histologic characteristics that set it apart from other causes of alopecia and from classic forms of cutaneous lupus. These distinct features include a linear configuration, a younger age of presentation, a disproportionate number of Asians affected, and a paucity of cases with systemic involvement. Histologically, the lesions are characterized by prominent mucin in the dermis and subcutaneous tissues. Blaschkoid linear lupus of the scalp is sufficiently distinctive to suggest the diagnosis on histology alone, in the appropriate clinical context. The most common and successful treatments included systemic and/or combination treatment with oral hydroxychloroquine, oral steroids, and/or intralesional steroids.

Keywords

cutaneous lupus erythematosus; autoimmune diseases; skin/pathology; discoid lupus erythematosus; systemic lupus erythematosus; alopecia; scalp; Blaschkoid alopecia; Blaschkoid lupus; linear alopecia; linear lupus

Introduction

There are a variety of skin conditions that have been reported to occur in linear arrangements along Blaschko's lines. These include lichen striatus, linear morphea, linear psoriasis, linear granuloma annulare, and linear Darier disease, among many others.[1] While linear eruptions are not uncommon findings in dermatology, linear cutaneous lupus erythematosus resulting in alopecia is quite rare. To our knowledge, there have only been 16 cases reported in the English language of linear lupus involving the scalp. This condition appears to disproportionally affect young males of Asian descent, demographically distinguishing this condition from other forms of lupus erythematosus.[2] We report 2 additional cases of linear cutaneous lupus following Blaschko's lines on the scalp, and review the literature with an emphasis on this entity's distinct clinical and histopathologic findings.

Case Description

Patient 1

A 16-year-old Vietnamese male presented with a 1-year history of hair loss of the scalp. He was otherwise healthy with no family history of autoimmune diseases. The primary patch of alopecia presented as a linear, arc-shaped pattern involving his occipital and parietal scalp with faint erythema (**Figure 1**). There was a separate, 1.5 cm-round patch of alopecia on his right parietal scalp resembling alopecia areata. There was no induration, sclerosis, follicular plugging, or other changes. He denied scalp itching, pain,

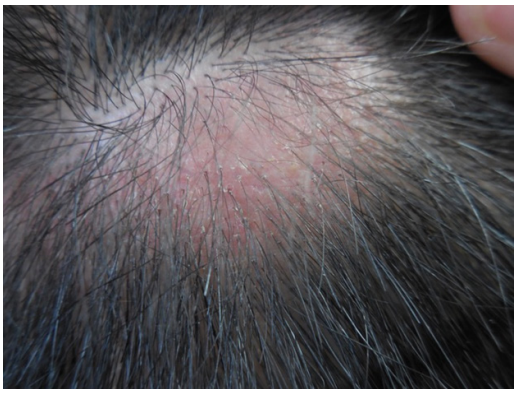

Figure 1. Patient 1 presented initially with linear, arc-shaped alopecia involving the occipital and parietal scalp. There was minimal erythema within the lesion.

photosensitivity, or systemic symptoms but reported prior patches of hair loss on his frontal scalp that had resolved spontaneously. Laboratory evaluation including blood count, metabolic panels, C-reactive protein, sedimentation rate, complement 3 (C3) and 4 (C4) were within the normal limits. Antinuclear antibodies (ANA) were positive with a titer of 1:160 and a homogeneous pattern. Additional workup including double-stranded DNA, anti-Ro, anti-La, and an Scl-70 antibody test was negative.

The patient was treated with 5 mg/ml of intralesional triamcinolone. At his 1 month follow-up, there was patchy regrowth of hair throughout the treated area and resolution of the erythema with no atrophy. The areas were again treated with 5 mg/ml of intralesional triamcinolone and a repeat follow-up revealed near resolution of the lesions (**Figure 2**). Follow-up was extended to 4 months and at that time there was recurrent alopecia in the arc-shaped area on the occipital scalp, which now had evident erythema and scale. There were new patches on his right parietal scalp with similar findings (**Figure 3**). The patient elected to repeat treatment with intralesional triamcinolone and 20 mg/ml was used for this purpose. At 6 weeks follow-up there was minimal improvement in the lesions. He was again treated with 20 mg/ml of intralesional triamcinolone and started on 200 mg of hydroxychloroquine daily. Additionally at 6 weeks follow-up there was patchy regrowth of hair with improvement in the associated erythema and scale with no new lesions. Biopsies were done with results described later in the article.

Patient 2

A 20-year-old male of Iranian descent presented for a 6-month history of progressive alopecia. The physical exam revealed a prominent, linear appearing non-scarring alopecia on the occipital scalp. He was otherwise healthy and denied systemic symptoms. ANA was positive but titers were not specified. Anti-SS-B was positive with a value of 7.6 U (range 0-0.9). The biopsy's histological features were very similar to those of the prior patient with linear alopecia, as described below. This patient was

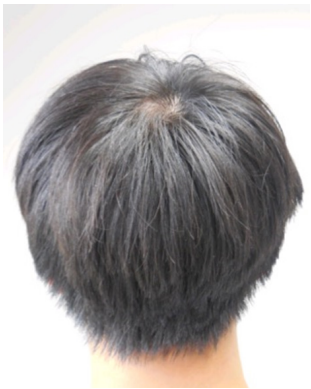

Figure 2. Patient 1 on follow-up revealed significant improvement in the patches of alopecia after intralesional triamcinolone.

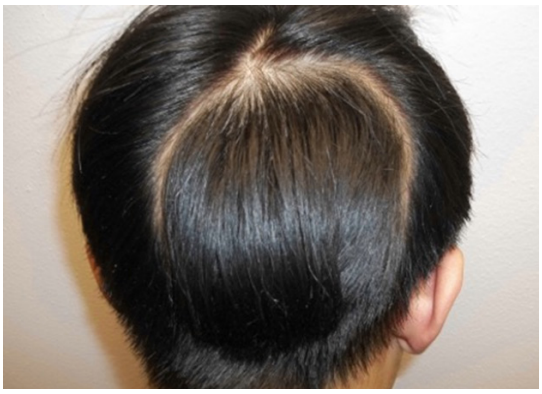

Figure 3. Patient 1 on second follow-up revealed recurrent and new lesions of alopecia, now with associated erythema and scale.

treated with a 3-week prednisone taper with improvement in the alopecia. He was then lost to further follow-up.

Histologic Findings

Two 4-mm punch biopsies were obtained on each patient. In both cases, 1 biopsy was oriented and cut in horizontal sections, and the other was cut in a vertical manner. The histologic findings within both cases were similar. There was increased mucin deposition throughout the reticular dermis, as well as prominent mucin deposition in the subcutaneous tissue (**Figure 4**). There was a mild perivascular and focally perifollicular lymphocytic infiltrate. Interface alteration of the dermal-epidermal junction, a lichenoid infiltrate, and basement membrane thickening were not present. There was miniaturization with an increased telogen/catagen ratio (approximately 18-20%) of the background hairs (**Figure 5**). A diagnosis of linear alopecia/Blaschkoid linear lupus erythematosus was made. Direct immunofluorescence testing for IgM, IgG, IgA, C3, and fibrinogen on both patient samples was negative.

Discussion

In 1901, Alfred Blaschko first reported epidermal nevi occurring on the body in linear or whorled patterns. Blaschko's lines are thought to result from the embryonic migration pathways of skin cells.[3] Skin conditions occurring in Blaschko's lines are believed to result from genetic mosaicism. The distribution of skin conditions along Blaschko's lines that involve dermal processes is thought to be due to epidermal mutations that lead to changes in dermal tissues.[4] Blaschko's original diagrams did not include the lines of the face or scalp; however, the pattern now accepted was later depicted by Happle in 2001 (**Figure 6**).[2]

The first report of cutaneous lupus occurring in Blaschko's lines was reported by Richarz et al. in 1986.[3] The term "linear cutaneous lupus erythematosus" was later proposed by Abe et al. in 1998 to reflect the linear nature of the disease and the lack of internal involvement.[4] In their review of 6 patients with linear discoid lupus erythematosus (DLE), they concluded that this entity presents with a linear configuration, en-

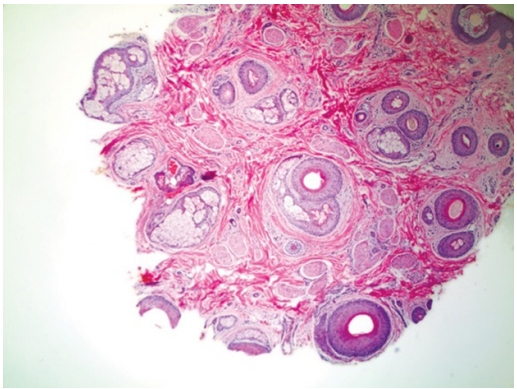

Figure 4. Patient 1 histopathology shows a vertical section stained with colloidal iron demonstrating prominent mucin.

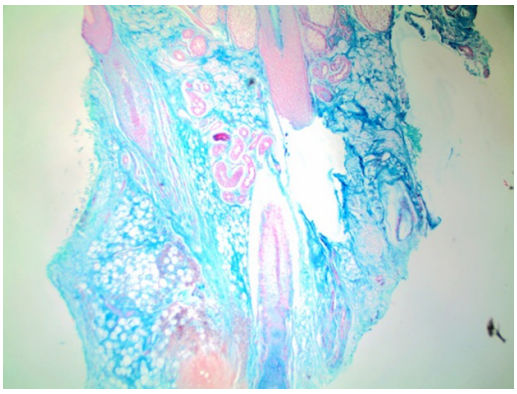

Figure 5. Patient 1 histopathology shows a horizontal section demonstrating the miniaturization of hairs with an increased telogen/catagen ratio.

compasses a younger patient population, lacks photosensitivity, and lacks systemic involvement.[4] The term "Blaschko linear lupus erythematosus" was recently proposed by Jin et al. to classify a distinct subtype of cutaneous lupus occurring in lines of Blaschko.[2] Blaschko linear lupus erythematosus encompasses all subtypes of cutaneous lupus erythematosus including DLE, subacute cutaneous lupus erythematosus, lupus erythematosus profundus, tumid lupus, and bullous lupus.[2] Jin's findings mirrored the earlier findings of Abe and showed that this involved a younger patient population, paucity of systemic involvement, and rare photosensitivity. In addition, both researchers found a disproportionate number of cases reported from Asian countries.[2]

Linear lupus of the scalp resulting in alopecia was first reported by Nagai et al. in 2003.[5] Since that time, an additional 18 cases have been reported, including the two present cases (**Table 1**). Of these 18 reported cases, 13 (72%) are from Asian countries or involve a subject of Asian descent, suggesting an ethnic predisposition. This is consistent with prior reports that 62% of Blaschko-linear lupus erythematosus cases were reported from Asian countries.[2] Twelve of the 18 (67%) cases involved male subjects. This is compared to women accounting for 64% to 82% of classic forms of cutaneous lupus erythematosus and over 90% of cases of systemic lupus erythematosus (SLE).[6-9] Although the pathogenesis of lupus is not fully elucidated, it is thought to be a multifactorial disease with environmental and genetic factors playing a role. In addition, it is believed that hormones play a role in the pathogenesis of lupus, contributing to the female predominance.[10] The relative sex equality seen in linear Blaschkoid lupus may suggest a different pathogenesis from classic forms of cutaneous lupus erythematosus with hormones playing a lesser role.

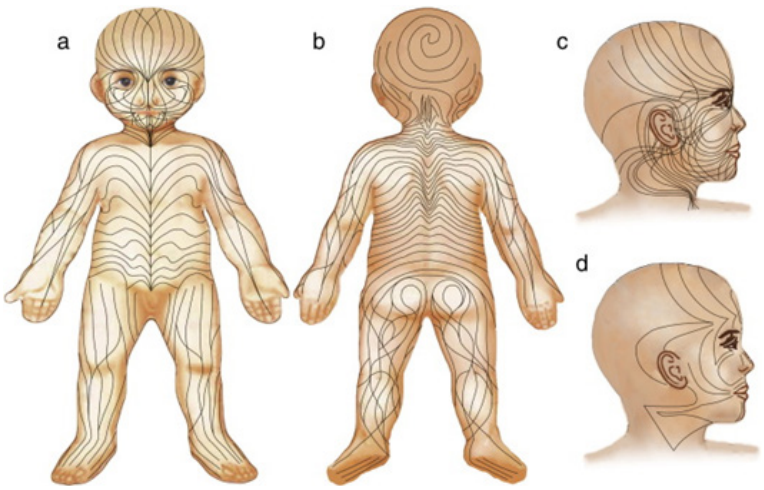

Figure 6. The illustration shows Blaschko's lines on an infant model. Note the lines on the posterior scalp in b. Reproduced from Jin et al. with permission from Elsevier.[15]

Table 1. Summary of Clinical Features of Linear Lupus Occurring on the Scalp

	Country (descent)	Age/ Gender	Abnormal labs	Classification	Length of follow-up	Treatment	Outcome
Case 1[15]	Japan (Japanese)	10/F	ANA 1:320	Lupus profundus	1.5 years	Topical steroids	Decreased erythema, no change in alopecia
Case 2[16]	Taiwan	21/F	SS-A, SS-B, Sm, Scl-70, RNP, ANA negative	Lupus profundus	1 year	Hydroxychloroquine 200 mg/day x 7 weeks	Complete resolution
Case 3[17]	Turkey	16/F	Leukopenia ANA 1:320	Lupus profundus	2 months	Topical mometasone furoate. hydroxychloroquine 200 mg/day	No change with topicals, significant improvement with hydroxychloroquine
Case 4[18]	Korea	20/M	ANA negative	Lupus profundus	1.3 years		Improvement with atrophy
Case 5[19]	Korea	14/M	No abnormalities reported	Lupus profundus	1 year	Hydroxychloroquine 400 mg/day x 5 weeks, prednisolone 20 mg/day x 2 weeks, IL triamcinolone 5 mg/ml x 3.	Complete resolution, facial hyperpigmentation thought to be related to hydroxychloroquine
Case 6[19]	Korea	32/M	No mention of labs	Lupus profundus	12 weeks	Dapsone 50 mg/day x 12 weeks	Regrowth
Case 7[20]	Taiwan (Taiwanese)	32/M	ANA negative, C3, C4 within normal limits	Lupus panniculitis	9 months	Hydroxychloroquine 200 mg/day and IL triamcinolone q2 weeks x 2 months.	Complete regrowth at 9 months with new lesions
Case 8[14]	Italy	21/F	ANA 1:1280, positive dsDNA, leukopenia, elevated ESR, low C3,	Tumid lupus	27 years	IL steroids	Healed with sclerotic atrophic plaque, no change in alopecia
Case 9[21]	Japan	26/F	ANA 1:1280, SS-A positive (12.3 IU/ml)	Lupus profundus	2 years	Prednisolone 20 mg/day x 2 years	Complete resolution
Case 10[22]	Spain	34/M	ANA 1:320, SS-A positive	Lupus panniculitis	1 year	Initial: hydroxychloroquine 400 mg/day x 5 months; Relapse: prednisone 30 mg/day tapered over 3 weeks	Improvement followed by relapse

Abbreviations: F=Female; M=Male; ANA=Antinuclear antibodies; IL=Intralesional

Table 1. Summary of Clinical Features of Linear Lupus Occurring on the Scalp (*continued*)

	Country (descent)	Age/ Gender	Abnormal labs	Classification	Length of follow-up	Treatment	Outcome
Case 11[23]	USA (Caucasian)	26/M	ANA, SS-A negative	Lupus panniculitis	3.5 years	Prednisone taper over 2 months, remission; Recurrence: hydroxychloroquine 400 mg/day x 6 months + prednisone x 1 month, IL triamcinolone Q2 months	Complete resolution
Case 12[28]	USA (African American)	53/F	Mild lymphopenia, ANA, C3, C4 within normal limits,	Lupus panniculitis	3 months	Prednisone, hydroxychloroquine, mycophenolate mofetil, and intralesional triamcinolone injections.	Ulcers healed with residual scarring
Case 13[29]	Germany	34/M	ANA 1:200, slightly increased C3	Lupus profundus	12 months	Hydroxychloroquine	Complete resolution
Case 14[30]	India	17/M	ANA negative	Lupus panniculitis	3 months	Oral prednisolone 20 mg, hydroxychloroquine 200 mg twice daily	Complete resolution
Case 15[31]	Thailand (Burmese)	28/M	ANA 1:320, speckled	Lupus panniculitis	3 months	Oral prednisolone 40 mg, hydroxychloroquine 400 mg daily, intralesional triamcinolone injections monthly, 5% minoxidil lotion twice daily	Improvement, increase hair count, decreased erythema
Case 16[32]	Korea	18/M	ANA 1:20 speckled	Lupus panniculitis	12 weeks	Oral hydroxychloroquine, topical corticosteroid	Regrowth of the terminal hairs
Present case 1	USA (Vietnamese)	16/M	ANA 1:160	Blaschko linear lupus	2 months	5 mg/ml IL triamcinolone	Improvement, patchy regrowth
Present case 2	USA (Iranian)	20/M	ANA positive, titer not specified, SS-B positive (7.6 IU/ml)	Blaschko linear lupus	3 months	3 week prednisone taper	Improvement

Abbreviations: F=Female; M=Male; ANA=Antinuclear antibodies; IL=Intralesional

Over half of the cases with linear Blaschkoid lupus of the scalp had positive autoantibodies. The most commonly detected autoantibody was ANA (positive in 10 of the 18 cases [56%]). This is in contrast to other forms of lupus with the ANA being positive in 95% to 100% of those with SLE, 70% to 80% of subacute cutaneous lupus erythematosus, and approximately 20% of DLE.[11,12] Six of the 10 patients with positive ANA titers had values of 1:320 or higher. ANA titers greater than or equal to 1:320 have been reported as a risk factor for progression from cutaneous lupus erythematosus to SLE. Thus, it is recommended that such patients be followed for the development of SLE.[13] None of the patients with linear lupus involving the scalp had systemic symptoms at the time of presentation, and only one developed systemic symptoms. This occurred 9 years after presentation and, at that time, she fulfilled the diagnostic criteria for SLE based on American Rheumatism Association criteria.[14]

The mean age of presentation with linear Blaschkoid lupus of the scalp was 24 years, which is younger than the age of 48.5 years reported for classic forms of cutaneous lupus erythematosus.[6] This was similar to the median age of 19 that was reported for patients with Blaschko linear lupus erythematosus elsewhere on the body.[2] This is also consistent with manifestations of polygenic skin disorders occurring in a segmental pattern, as they often present at a younger age.[15]

Blaschkoid lupus of the scalp and elsewhere on the body have a low incidence of photosensitivity.[2,4,5,14,16-23] This may be partly explained by the large proportion of Asian subjects affected, as it has been reported that people of Japanese descent have a lower incidence of photosensitivity in classic forms of cutaneous lupus erythematosus.[24]

The primary differential diagnoses for linear alopecia occurring in an adult are linear morphea, alopecia areata, and trichotillomania. These can be distinguished with clinicopathological correlation. Linear morphea clinically appears as an atrophic, sclerotic plaque that may feel indurated and tethered to underlying tissues. Lesions with active inflammation may have surrounding violaceous erythema.[25] Histological evaluation of early morphea lesions may have subtle findings or reveal a dense, perivascular, chronic inflammatory cell infiltrate; however, mucin is not a prominent finding.[25] Established lesions are characterized by thickened collagen bundles and absent or atrophic adnexal structures.[25] Alopecia areata typically presents as round, asymptomatic patches of alopecia, although an ophiasis pattern may be considered linear. There is no associated erythema or scale. Histological evaluation of early lesions reveals lymphocytes surrounding the hair bulb in the subcutaneous tissue. Late-stage alopecia areata has decreased inflammation and numerous miniaturized and telogen hair follicles.[26] Trichotillomania presents clinically as localized area(s) of incomplete hair loss with residual small non-pigmented broken hairs. Histological evaluation shows increased catagen follicles and pigment casts in the follicular canal with hyperkeratosis.[26] A vertically-oriented split within the hair shaft may be observed.[26] Hair follicle alteration may include hemorrhage, collapsed inner root sheath, or trichomalacia.[26]

In contrast, the clinical and histologic features of linear lupus are distinctive. More than half the cases presented with normal-appearing skin in the affected area. In addition, the lesions can present as a round patch prior to progression to a linear configuration. This was illustrated by Mitxelena et al., who reported a case that clinically resembled alopecia areata, which was also evident in our first patient's initial presentation.[22] Six out of 18 cases presented with mild erythema in the area of alopecia, which may serve as a subtle sign to lead the clinician away from a diagnosis of alopecia areata. Fourteen out of 18 cases of linear lupus occurring on the scalp had mucin deposition on histological evaluation and 10 out of 18 cases reported mucin as abundant, strong, or prominent. Abundant mucin deposition in the scalp and subcutaneous tissue is not a histologic feature of other causes of alopecia, and its presence makes it an important feature to differentiate from other causes of hair loss and allows the dermatopathologist to render the correct diagnosis (**Table 2**). Nine patients with linear lupus erythematosus on the scalp had direct immunofluorescent antibody testing performed. Of these, 4 (44%) were positive (**Table 2**). This is in contrast to a positive result in the nonlinear form of DLE and SLE reported as 69% and 72%, respectively.[27] Consistent with other forms of cutaneous

Table 2. Histological Findings

	Histological findings	DIF
Case 1[5]	Fat degeneration, abundant mucin deposit, slight perivascular and periappendegeal infiltrate, normal epidermis	Not performed
Case 2[16]	No vacuolar degeneration of the BMZ, perieccrine plasma cell and lymphocytic infiltrate, fat degeneration, slight lymphocytic infiltrate in subQ, abundant mucin	Not performed at initial presentation; negative 1 year after treatment
Case 3[17]	Scalp: perifollicular lymphocyte inflammation; axilla: prominent dermal mucin, basal vacuolar degeneration, lymphocytic nodules in the deep dermis, hyaline degeneration of fat	C3 and IgM in the follicular epithelium
Case 4[18]	Lymphocytic panniculitis with fat degeneration and mucin deposit, focal hydropic degeneration of basal cells	Peribulbar granular deposits of IgM, IgA, and C3
Case 5[19]	Follicular plugging, perifollicular lymphoid cell infiltrate and frequent catagen hairs (2/3 were in catagen), perieccrine inflammation, abundant mucin	IgM, IgG, IgA, C3, fibrinogen negative
Case 6[19]	Deep dermal perivascular lymphoid infiltrate, mucin deposition, frequent catagen stage, no significant perifollicular infiltrate.	IgM, IgG, IgA, C3, fibrinogen negative
Case 7[20]	Unremarkable epidermis, shrinking hair follicles, prominent peribulbar lymphocytic and plasma cell infiltration, abundant mucin	IgM, IgG, IgA, C3 negative
Case 8[14]	No histologic description provided	Not performed
Case 9[21]	Perivascular lymphocytic infiltrate with lobular and septal panniculitis, mucin	Granular IgG in BMZ
Case 10[22]	Dense lymphocytic infiltrate affecting the deep dermis and subQ, vacuolar damage of the basal layer, nodular lymphoid aggregates with plasma cells	Granular IgM in BMZ
Case 11[23]	Superficial and deep lymphocytic infiltrate, mucin deposition in dermis and subQ	Not performed
Case 12[28]	Mildly atrophic epidermis, basal vacuolization, dermal melanophages, BMZ thickening, superficial and deep dermal perivascular lymphohistiocytic inflammation, subQ with hyaline fat necrosis, septal fibrosis and dense lobular lymphocytic inflammation	Not performed
Case 13[29]	Normal epidermis, strong dermal and subQ mucin accumulation, lobular lymphocytic panniculitis	Not performed
Case 14[30]	Moderate pericapillary lymphocytic infiltrate, fat necrosis, myxoid degeneration, interlobular septa hyaline deposits	Not performed
Case 15[31]	Dense perifollicular lymphoid infiltration at the infudibuloisthmic hair follicle, vacuolar interface at the infundibular epithelium, epidermal atrophy, follicular plugging, apoptotic, keratinocytes, mild melanin incontinence, superficial and deep perivascular infiltrates, necrosis of fat lobules, interstitial mucin in the dermis and subQ fat	Not performed
Case 16[32]	Dense lymphocytic infiltrates in deep dermis subq perivascular and periadnexal lymphocytic infiltrate, abundant mucin deposition	Not performed
Present case 1	Elevated telogen/catagen hair ratio (45%), abundant mucin particularly in subcutaneous fat, perivascular lymphocytic infiltrate	IgM, IgG, IgA, C3, fibrinogen negative
Present case 2	Elevated telogen/catagen hair ratio, abundant mucin particularly in subcutaneous fat, perivascular lymphocytic infiltrate	IgM, IgG, IgA, C3, fibrinogen negative

Abbreviations: DIF=Direct immunofluorescence; BMZ=Basement membrane zone; subQ=subcutaneous

Table 3. Case Series Summary of Treatment Modalities

	Monotherapy versus combination therapy	Topical steroids	Intralesional (IL) steroids	Oral steroids	Hydroxychloroquine	Mycophenolate mofetil	Dapsone	Topical minoxidil	Response to treatment
Case 1[5]	Monotherapy	Topical steroids							No change
Case 2[16]	Monotherapy				Hydroxychloroquine 200 mg/day x 7 weeks				Complete resolution
Case 3[17]	Combination	Topical mometasone furoate			Hydroxychloroquine 200 mg/day				Partial response
Case 4[18]	Unknown								Partial response
Case 5[19]	Combination		IL triamcinolone 5 mg/ml x 3	Prednisolone 20 mg/day x 2 weeks	Hydroxychloroquine 400 mg/day x 5 weeks				Complete resolution
Case 6[19]	Monotherapy						Dapsone 50 mg/day x 12 weeks		Partial response
Case 7[20]	Combination		IL triamcinolone q2 weeks x 2 months		Hydroxychloroquine 200 mg/day				Partial response
Case 8[14]	Monotherapy		IL steroids						No change
Case 9[21]	Monotherapy			Prednisolone 20 mg/day x 2 years					Complete resolution
Case 10[22]	Combination			Prednisone 30 mg/day tapered over 3 weeks	Hydroxychloroquine 400 mg/day x 5 months				Partial response
Case 11[23]	Combination		IL triamcinolone Q2 months	Prednisone taper over 2 months, remission. Recurrence: prednisone x 1 month	Hydroxychloroquine 400 mg/day x 6 months +				Complete resolution
Case 12[28]	Combination		IL triamcinolone injections	Prednisone, hydroxychloroquine	Hydroxychloroquine	Mycophenolate mofetil			Partial response
Case 13[29]	Monotherapy				Hydroxychloroquine				Complete resolution
Case 14[30]	Combination			Oral prednisolone 20 mg	Hydroxychloroquine 200 mg twice daily				Complete resolution
Case 15[31]	Combination		IL triamcinolone injections monthly	Oral prednisolone 40 mg	Hydroxychloroquine 400 mg daily			5% minoxidil lotion twice daily	Partial response
Case 16[32]	Combination	Topical corticosteroid			Oral hydroxychloroquine				Partial response
Present case 1	Monotherapy		5 mg/ml IL triamcinolone						Partial response
Present case 2	Monotherapy			3-week prednisone taper					Partial response
Total	9 Monotherapy, 9 Combination	3 (17%)	7 (39%)	8 (44%)	11 (61%)	1 (6%)	1 (6%)	1 (6%)	2 no change, 10 partial responses, 6 complete resolution

lupus, IgM, and C3 were the deposits most frequently detected.

The clinical course of linear cutaneous lupus of the scalp is unpredictable. Similar to DLE and lupus erythematosus profundus, Blaschko linear lupus of the scalp may result in scarring alopecia.[2,5,14] This occurred in 2 of the 18 documented patients. In addition, Blaschkoid linear lupus of the scalp can also be a self-limited process. This was illustrated by both our patients, who reported prior areas of alopecia that had spontaneously resolved without sequela prior to presentation. Given that lupus profundus and lupus panniculitis involve the subcutaneous tissues and deep dermis, it is unlikely that topical medications penetrate sufficiently for efficacy. This is consistent with the lack of improvement in prior cases that used only topical therapy.[5,17]

It is difficult to make treatment recommendations from the previous case reports. Treatment was similar to standard treatment for cutaneous lupus. Therapy generally consisted of mid- and high-potency topical steroids and immunosuppressive agents. Oral hydroxychloroquine was the most frequently used medication (61%), followed by oral steroids (44%). Intralesional steroid injections were used less often (39%), while it is worthwhile to note care should be given due to the risk of atrophy. Topical steroids as monotherapy were seldom used (17%). It can be inferred that the treating clinicians considered topical treatment to have limited efficacy due to the level of tissue involvement. Mycophenolate mofetil and topical minoxidil were each used in separate cases, both in combination with other treatments, while dapsone was used in a single case as monotherapy.

Response to treatment(s) was reported as either no change, improved, or complete resolution of alopecia (**Table 3**). Topical and intralesional steroids as monotherapy were associated with higher treatment failure and were the only 2 cases showing no change. The other cases reviewed used a combination of treatments, either systemic, topical, and/or intralesional, with 100% of cases showing partial improvement or complete resolution. Monotherapy using oral hydroxychloroquine in 2 cases and oral prednisolone in another showed complete resolution. In a fourth case showing complete resolution, the combination of the two aforementioned therapies was used. Complete resolution was also achieved in a fifth and sixth cases when intralesional triamcinolone was added to oral hydroxychloroquine and systemic steroids. Early systemic and/or combination treatment is recommended due to the depth of subcutaneous involvement, with the hope of preventing scarring alopecia and a higher likelihood to have a positive response.

Conclusion

Linear Blaschkoid lupus of the scalp resulting in alopecia is extremely rare, with only 18 reported cases. It disproportionally affects young adult Asian males, distinguishing it from other forms of lupus, including DLE. The histology is characterized by a dermal and subcutaneous lymphoid infiltrate with abundant subcutaneous mucin. Dermatologists and dermatopathologists should be aware of this rare entity. Abundant and prominent mucin should raise suspicion for this entity, and in the correct clinical context, a specific diagnosis of linear cutaneous lupus following Blaschko's lines can be rendered.

Conflicts of Interest

The authors declare they have no conflicts of interest.

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