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 disproportionately 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,
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
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/Blaschkooid 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. 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. 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). The first report of cutaneous lupus occurring in Blaschko’s lines was reported by Richarz et al. in 1986. 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. In their review of 6 patients with linear discoid lupus erythematosus (DLE), they concluded that this entity presents with a linear configuration, en-
compasses a younger patient population, lacks photosensitivity, and lacks systemic involvement. The term "Blaschko linear lupus erythematous" was recently proposed by Jin et al. to classify a distinct subtype of cutaneous lupus occurring in lines of Blaschko. Blaschko linear lupus erythematous encompasses all subtypes of cutaneous lupus erythematous including DLE, subacute cutaneous lupus erythematous, lupus erythematous profundus, tumid lupus, and bullous lupus. 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.

Linear lupus of the scalp resulting in alopecia was first reported by Nagai et al. in 2003. 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 erythematous cases were reported from Asian countries. 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 erythematous and over 90% of cases of systemic lupus erythematous (SLE).

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. The relative sex equality seen in linear Blaschko-linear lupus may suggest a different pathogenesis from classic forms of cutaneous lupus erythematous with hormones playing a lesser role.

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

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.
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* Japan (Japanese) | 10/F | ANA 1:320 | Lupus profundus | 1.5 years | Topical steroids | Decreased erythema, no change in alopecia |
| Case 2* 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* 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 Korea | 20/M | ANA negative | Lupus profundus | 1.3 years | Improvement with atrophy |
| Case 5 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 Korea | 32/M | No mention of labs | Lupus profundus | 12 weeks | Dapsone 50 mg/day x 12 weeks | Regrowth |
| Case 7 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 resolution at 9 months with new lesions |
| Case 8 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 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 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
| Case Number | Country (descent) | Age/Gender | Abnormal labs | Classification | Length of follow-up | Treatment | Outcome |
|-------------|------------------|------------|---------------|----------------|---------------------|-----------|---------|
| Case 1123  | 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 1228  | 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 1329  | Germany          | 34/M       | ANA 1:200, slightly increased C3 | Lupus profundus | 12 months | Hydroxychloroquine | Complete resolution |
| Case 1430  | India            | 17/M       | ANA negative | Lupus panniculitis | 3 months | Oral prednisolone 20 mg, hydroxychloroquine 200 mg twice daily | Complete resolution |
| Case 1531  | Thailand (Burmese) | 28/M | ANA 1:320, speckled | Lupus panniculitis | 3 months | Oral prednisolone 40 mg, hydroxychloroquine 400 mg daily, intrale- sional triamcinolone injections monthly, 5% minoxidil lotion twice daily | Improvement, increase hair count, decreased erythe- ma |
| Case 1632  | Korea            | 18/M       | ANA 1:20 speckled | Lupus panniculitis | 12 weeks | Oral hydroxychloroquine, topical cortico- steroid | 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. 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. 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.

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. This was similar to the median age of 19 that was reported for patients with Blaschko linear lupus erythematosus elsewhere on the body. This is also consistent with manifestations of polygenic skin disorders occurring in a segmental pattern, as they often present at a younger age.

Blaschkoid lupus of the scalp and elsewhere on the body have a low incidence of photosensitivity. 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.

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. 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. Established lesions are characterized by thickened collagen bundles and absent or atrophic adnexal structures. 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. 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. A vertically-oriented split within the hair shaft may be observed. Hair follicle alteration may include hemorrhage, collapsed inner root sheath, or trichomalcia.

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. 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. Consistent with other forms of cutaneous
Table 2. Histological Findings

| Case  | Histological findings                                                                                                                                                                                                 | DIF                                                                 |
|-------|-----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|----------------------------------------------------------------------|
| 1³    | Fat degeneration, abundant mucin deposit, slight perivascular and periappendageal infiltrate, normal epidermis                                                                                                | Not performed                                                        |
| 2⁶    | 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 |
| 3⁷    | 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                                |
| 4⁸    | Lymphocytic panniculitis with fat degeneration and mucin deposition, focal hydropic degeneration of basal cells                                                                                                     | Perilobular granular deposits of IgM, IgA, and C3                     |
| 5⁹    | 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                                 |
| 6⁹    | Deep dermal perivascular lymphoid infiltrate, mucin deposition, frequent catagen stage, no significant perifollicular infiltrate.                                                                               | IgM, IgG, IgA, C3, fibrinogen negative                                 |
| 7¹⁰   | Unremarkable epidermis, shrinking hair follicles, prominent perilobular lymphocytic and plasma cell infiltration, abundant mucin                                                                               | IgM, IgG, IgA, C3 negative                                           |
| 8¹⁴   | No histologic description provided                                                                                                                                                                               | Not performed                                                        |
| 9¹⁴   | Perivascular lymphocytic infiltrate with lobular and septal panniculitis, mucin                                                                                                                                   | Granular IgG in BMZ                                                  |
| 10²²  | 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                                                  |
| 11²³  | Superficial and deep lymphocytic infiltrate, mucin deposition in dermis and subQ                                                                                                                                  | Not performed                                                        |
| 12²⁸  | 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                                                        |
| 13²⁹  | Normal epidermis, strong dermal and subQ mucin accumulation, lobular lymphocytic panniculitis                                                                                                                     | Not performed                                                        |
| 14³⁰  | Moderate pericapillary lymphocytic infiltrate, fat necrosis, myxoid degeneration, interlobular septa hyaline deposits                                                                                             | Not performed                                                        |
| 15³¹  | Dense perifollicular lymphoid infiltrate at the infundibuloisthmic hair follicle, vascular 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                                                        |
| 16³²  | 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
| Case | Monotherapy versus combination therapy | Monotherapy | Combination | Intralesional (IL) steroids | Oral steroids | Hydroxychloroquine | Mycophenolate mofetil | Dapsone | Topical minoxidil | Response to treatment |
|------|----------------------------------------|-------------|-------------|-----------------------------|--------------|--------------------|----------------------|---------|-----------------|----------------------|
| 1    | Monotherapy                            | Topical steroids | Topical mometasone furoate | Hydroxychloroquine 200 mg/day | 3 (17%) | 8 (44%) | 11 (61%) | 1 (6%) | 1 (6%) | No change |
| 2    | Monotherapy                            | Topical steroids | Intralesional (IL) steroids | Hydroxychloroquine 200 mg/day x 7 weeks | 7 (39%) | 11 (61%) | 1 (6%) | 1 (6%) | 1 (6%) | Complete resolution |
| 3    | Unknown                                | Hydroxychloroquine 400 mg/day x 5 weeks | Prednisone taper over 2 months, remission. Recurrence: prednisone 1 month | Complete resolution |
| 4    | Unknown                                | IL triamcinolone 5 mg/ml x 3 | Prednisolone 20 mg/day x 2 weeks | Partial response |
| 5    | Combination                            | IL triamcinolone q2 weeks x 2 months | Hydroxychloroquine 200 mg/day | Complete resolution |
| 6    | Monotherapy                            | IL steroids | Hydroxychloroquine 200 mg/day | Partial response |
| 7    | Combination                            | IL triamcinolone Q2 months | Hydroxychloroquine 400 mg/day x 6 months + | Complete resolution |
| 8    | Monotherapy                            | IL triamcinolone injections | Hydroxychloroquine | Mycophenolate mofetil | Partial response |
| 9    | Monotherapy                            | Prednisolone 20 mg/day x 2 years | Complete resolution |
| 10   | Combination                            | Prednisone 30 mg/day tapered over 3 weeks | Hydroxychloroquine 400 mg/day x 5 months | Partial response |
| 11   | Combination                            | IL triamcinolone Q2 months | Prednisone taper over 2 months, remission. Recurrence: prednisone 1 month | Complete resolution |
| 12   | Combination                            | IL triamcinolone injections | Hydroxychloroquine | Complete resolution |
| 13   | Monotherapy                            | Hydroxychloroquine | Complete resolution |
| 14   | Combination                            | Oral prednisolone 20 mg | Hydroxychloroquine 200 mg twice daily | Complete resolution |
| 15   | Combination                            | IL triamcinolone injections monthly | Oral prednisolone 40 mg | 5% minoxidil lotion twice daily | Partial response |
| 16   | Combination                            | Topical corticosteroid | Oral hydroxychloroquine | Partial response |
| Present case 1 | Monotherapy | 5 mg/ml IL triamcinolone | Complete resolution |
| 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.\textsuperscript{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.\textsuperscript{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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