Isolated Linear Lichen Planopilaris: Extremely Rare When Limited to the Scalp

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Practice Points

- Linear lichen planopilaris (LPP) of the scalp can present as a linear patch or plaque associated with scaling alopecia followed by scarring.
- Classic LPP presents as a more diffuse, scaly patch or plaque associated with cicatricial alopecia.
- Systemic immunomodulatory therapy appears to be more effective in treating LPP than topical treatment alone.

Lichen planopilaris (LPP) is a primary cicatricial alopecia that rarely presents in a linear distribution. We present a case of linear LPP that was isolated to the frontal scalp with extension down the forehead. It is uncommon to see a linear distribution of LPP anywhere on the body, but it is particularly rare on the scalp. We also discuss the pathophysiology of disorders that present in unusual linear distributions.

Cutis. 2013;92:303-305.

Case Report

A 29-year-old man presented with a scaly linear plaque on the right side of the forehead of 4 months' duration with hair loss on the right frontal scalp that began 1 month following the appearance of the plaque. He had been treating the area with triamcinolone acetonide cream 0.1% for 3 weeks with no improvement. Physical examination revealed a sharply demarcated, erythematous, scaly, linear plaque on the right frontal scalp that extended down the forehead and was associated with a 35×20-mm patch of alopecia (Figure 1). Biopsy of the plaque on the forehead showed a moderate, superficial, perivascular, perifollicular, and perieccrine lymphocytic infiltrate with occasional plasma cells, rare eosinophils, mild spongiosis, and intermittent parakeratosis. Biopsy of the frontal scalp demonstrated a marked perifollicular infiltrate of lymphocytes with no involvement of the interfollicular epithelium and mild fibrosis consistent with scarring alopecia, more specifically lichen planopilaris (LPP)(Figure 2).

The patient was started on an oral dose of hydroxychloroquine 200 mg twice daily, clobetasol gel 0.05% twice daily, and salicylic acid shampoo 3% once daily. At 3 months' follow-up the patient showed remarkable improvement of LPP, with moderate hair regrowth at the margins of the original plaque and less scaling.

Comment

Lichen planopilaris is a primary cicatricial alopecia that is characterized by lymphocytic inflammation directed toward the infundibulum and isthmus portions of the hair follicle. Chronic inflammation leads to follicular scarring and irreversible hair loss, with an estimated annual incidence of 1.15% to 7.59%.[1,2]

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The authors report no conflict of interest.

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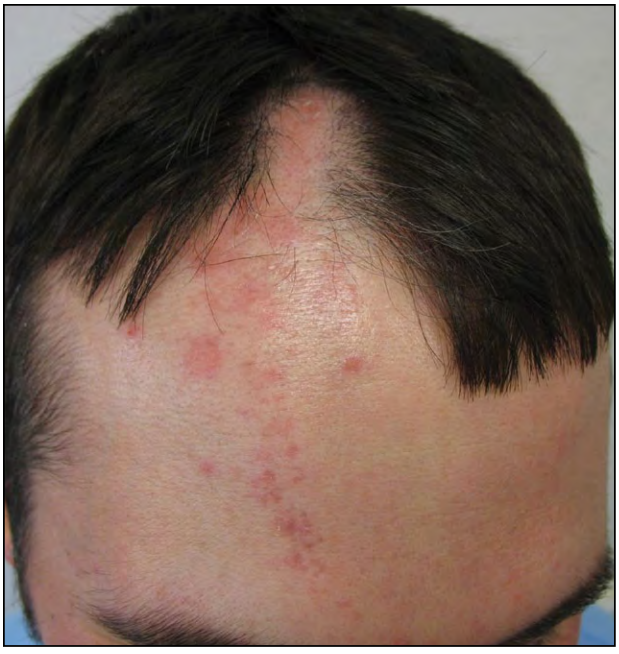

Figure 1. A scaly erythematous plaque with associated alopecia on the right frontal scalp and forehead.

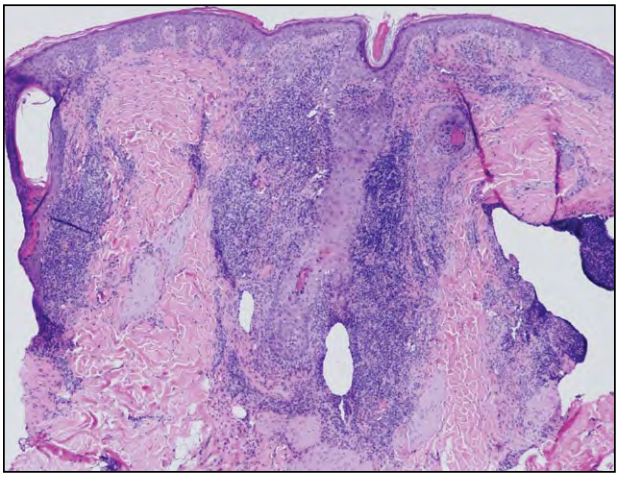

Figure 2. Punch biopsy of the frontal scalp demonstrated a dense lymphocytic infiltrate surrounding the superficial portions of the hair follicle (H&E, original magnification ×80).

A PubMed search of articles indexed for MEDLINE using the terms *lichen planopilaris* and *linear lichen planopilaris* revealed 9 reported cases of linear LPP,[3-8] with only 1 involving the scalp.[8]

Bussmann et al[8] described a phenomenon in which a polygenic cutaneous disorder can present with a superimposed segmental distribution following the lines of Blaschko. In these cases, patients present with a linearly distributed eruption somewhere on the body and a milder nonlinear presentation of the same eruption at another location on the body. The authors reported the first case of linear LPP presenting with this superimposed segmental manifestation.[8] This presentation is reminiscent of the type 2 segmental manifestation or loss of heterogeneity seen in some congenital disorders. As with most other cases of linear LPP, our patient presented with 1 linear plaque, which one might classify as a type 1 segmental manifestation or loss of heterogeneity. On the other hand, because LPP is considered a polygenic trait, Bussmann et al[8] reported that a type 1 segmental manifestation can never be recognized with certainty, and the use of types 1 and 2 segmental manifestations should be reserved for disorders with monogenic traits. For polygenic conditions or diseases, Happle[9] prefers the term *isolated* to describe segmental involvement similar to what was observed in our patient. The author stated that the pathophysiology of linear manifestations of inflammatory conditions is best explained by inherent etiologic factors in the affected skin and not simply by the inflammatory cells.[9]

The pathophysiology of LPP has long been poorly understood and is considered of autoimmune etiology. Some investigators have suggested that downregulation of the nuclear hormone receptor peroxisome proliferator–activated receptor γ within the epidermal cells of the isthmus of the hair follicle may be the pathogenesis of LPP.[10] Mirmirani and Karnik[11] treated a case of LPP refractory to all conventional therapies with the peroxisome proliferator–activated receptor γ agonist pioglitazone hydrochloride (15 mg daily) and reported that the patient showed dramatic improvement.

There are many treatment modalities for LPP, but the current first-line therapy includes high-potency topical and intralesional corticosteroids. Other modalities with varying reports of success include systemic corticosteroids, cyclosporine, mycophenolate mofetil, oral tetracyclines, oral retinoids, thalidomide, and hydroxychloroquine, though relapses after discontinuation of treatment frequently have been reported.[2] Hydroxychloroquine, an antimalarial and antilymphocytic drug, has been shown to be remarkably effective after 6 to 12 months of treatment, with most patients responding after 12 months.[12] A retrospective study comparing oral doxycycline, hydroxychloroquine, and mycophenolate mofetil found that only 27% (4/15) of patients showed improvement with doxycycline, 41% (9/22) showed improvement with hydroxychloroquine, and 30% (3/10) showed improvement with mycophenolate mofetil.[13] Most cases of linear LPP reported in

the literature regressed spontaneously with superficial scarring.[3] One case was treated with topical corticosteroids and topical retinoids with little success.[4] Another patient was treated with systemic and topical steroids with no improvement, which led to treatment with cyclosporine with notable improvement.[8] Our patient responded favorably to hydroxychloroquine therapy (200 mg twice daily) within 3 months.

Conclusion

The linear distribution of LPP with involvement of the face and scalp make our case unique. Most reports of linear LPP involve the face only, while cases of classic LPP usually involve the scalp.[3,4,6-7] These cases along with experimental findings of abnormal cellular activity within the epidermal cells of the isthmus of the hair follicle support Happle's[9] theory that the inherent etiologic factor for the disease is within the affected cutaneous structure, not simply the inflammatory cells.

Acknowledgment—The authors would like to thank Carol Daugherty for her help in locating references and coordinating communication between the authors.

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