# Lichen Planopilaris Presenting as Truncal Alopecia: A Case Presentation and Review of the Literature

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*Lichen planopilaris is an inflammatory condition of unknown etiology, characterized by violaceous folliculocentric papules and hair loss. Some clinicians consider lichen planopilaris to be a variant of lichen planus, but others believe it to be a separate disease entity. Many treatment modalities have been utilized, with varying degrees of success. We describe the case of a 63-year-old man who presented with widespread alopecia of the trunk and extremities and was subsequently diagnosed with lichen planopilaris.*
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Lichen planopilaris is an inflammatory skin disorder involving hair follicles of the scalp, trunk, and extremities. It is a chronic, often progressive disorder resulting in cicatricial alopecia that frequently presents in conjunction with lichen planus.[1] We present a case of lichen planopilaris presenting with alopecia of the trunk and a review of the literature.

## Case Report

A 63-year-old man presented with a 6-month history of hair loss over his trunk and extremities associated with pruritic purple lesions in the same distribution. Many years ago, he was treated for dermatosis of the scalp, which has remained quiescent. He has not had any treatment for his current skin condition. His medical history is significant for diabetes, hyperlipidemia, and hypertension. His current medications include metformin, simvastatin, and amlodipine. Findings from the physical examination revealed scattered, violaceous follicular papules diffusely distributed over the trunk (Figure 1) and extremities and patches of alopecia in a similar pattern. His scalp revealed areas of scarring alopecia. There were no mucous membrane or nail changes.

A punch biopsy of a follicular papule was performed, and results revealed a dilated, keratin-filled channel in the infundibular portion of the hair follicle, with an underlying dense lymphocytic infiltrate. A second biopsy of an area of alopecia demonstrated epidermal vacuolization of the basal layer, with a dense surrounding lymphocytic infiltrate, and a fibrotic tract inferior to the infundibular portion of the hair follicle (Figure 2). Direct immunofluorescence microscopy was negative for IgG, IgA, and C3. IgM was present in cytoid bodies at the dermal-epidermal junction. A diagnosis of lichen planopilaris was made. The patient refused treatment at the time.

## Comment

Lichen planopilaris was described first by Pringle[2] in 1895 as classic lichen planus in conjunction with spinous follicular papules. Today, lichen planopilaris or lichen planus follicularis[3] is considered a clinical syndrome of acuminate follicular papules, cicatricial (scalp) alopecia, and classic lichen planus; the latter is not necessarily always present.[4]

Lichen planopilaris usually affects adults, and, in contrast to lichen planus, is more common in women. Mean age of onset is 52 years, and, although the disorder can be chronic and progressive, it generally lasts from 6 months to 2 years.[3,4] The disorder evolves as a progressive series of clinical changes accompanied by characteristic histopathologic features.[5] Clinically, lichen planopilaris begins as a pruritic perifollicular erythema, often followed by the development of hyperkeratotic acuminate papules that are often purple. The

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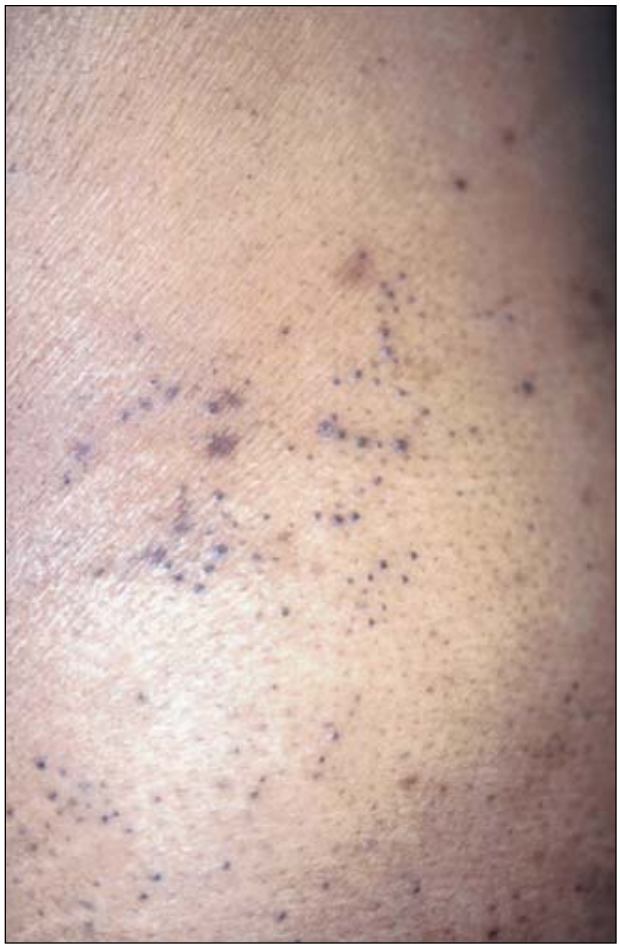

**Figure 1.** Violaceous folliculocentric papules over the right lower back.

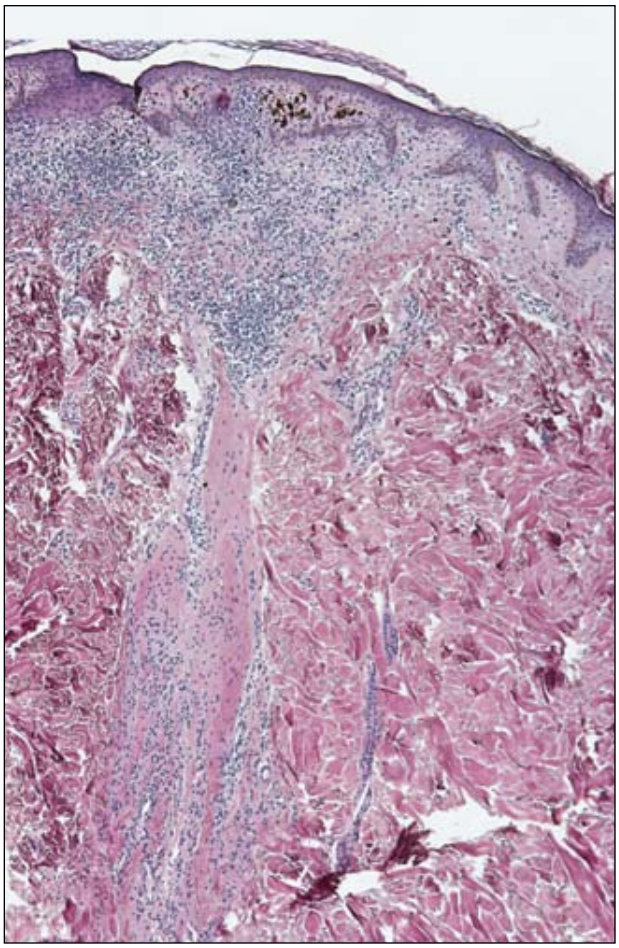

**Figure 2.** Epidermal vacuolization of the basal layer, with a dense surrounding lymphocytic infiltrate, and a fibrotic tract inferior to the infundibular portion of the hair follicle (H&E, original magnification ×4).

acuminate lesions are the most consistent clinical features of lichen planopilaris and represent dilated hair follicles filled with keratin plugs (Figure 1). The lesions can be widely distributed but have a predilection for the scalp, trunk, extremities, axilla, and groin. The lesions usually itch but may be asymptomatic. Hair follicle destruction and scarring of these lesions eventually result in a patchy smooth alopecia that can be clinically difficult to distinguish from other forms of alopecia. Lesions of typical lichen planus of nonfollicular skin, mucous membranes, and nails are present in only 50% of cases and usually follow the onset of cicatricial alopecia.[6]

Recently, the spectral nature of lichen planopilaris accounts for the varying clinical and pathologic presentations of this disease. Matta et al[7] detailed 3 variants of lichen planopilaris representing degrees of clinical and histologic scarring. These ranged from pure follicular involvement, without evidence of clinical scarring, to cicatricial alopecia. Kossard et al[8] described postmenopausal women with frontal fibrosing alopecia as a variant of lichen planopilaris, and Grunwald et al[9] characterized a scarring alopecia of the vulva as another variant. Recently, lichen planus follicularis tumidus[10] and linear lichen planopilaris of the face[11] also have been described as variants. Of historical significance, the Graham Little-Piccardi-Lassueur syndrome described by Graham Little in 1915 is the association of scarring alopecia of the scalp, hyperkeratotic follicular papules on glabrous skin, and noncicatricial alopecia of the axilla and pubic area.[1,8,12] There is still controversy as to whether this syndrome represents a form of lichen planopilaris or a separate disease entity.

The cause of lichen planopilaris is unknown. Some authors believe that lichen planopilaris is a subtype of lichen planus.[13] Infections, drugs, genetic factors, and immunologic abnormalities have been proposed as

potential etiologies of lichen planopilaris.[14-16] Smith et al[17] suggested that microorganism overgrowth causing an immune response limited to the follicle could be involved in disease pathogenesis. Recently though, there has been diverging immunofluorescence evidence that lichen planopilaris is not a subtype of lichen planus but a different disease altogether.[18]

Histologically, lichen planopilaris evolves through a progressive series of changes that often overlap, supporting the spectral nature of the disorder. Early lesions show dilated follicles containing plugged keratinous material and a perifollicular lymphocytic infiltrate.[5] As the lesions evolve, a lichenoid pattern of lymphocytic inflammation involving the infundibulum and isthmus of the hair bulge, along with perifollicular fibrosis, can be visualized.[4,10,19] The external root sheath shows vacuolar degeneration of the basal layer,[10] as well as cytoid bodies scattered along the follicular epithelial basement membrane.[4] The inferior segment of the follicle, reticular dermis, and interfollicular epidermis typically are spared. In late-stage lesions, inflammation can be minimal or slight with loss of distinctive lichenoid changes, but perifollicular fibrosis is striking.[20] Waldorf[5] documented flattened rete ridges and the complete absence of remnant pilosebaceous structures in late-stage smooth plaques; however, many late-stage and end-stage biopsy specimens are nondiagnostic and must be categorized as end-stage primary scarring alopecia.

Immunofluorescent microscopy findings of lichen planus have been extensively characterized; however, limited, conflicting data are available for lichen planopilaris.[6,13,21] In his series, Mehregan et al[6] showed the immunofluorescent findings in lichen planopilaris were similar to those of typical lichen planus in most patients. He reported interface immune deposits involving the hair follicle along the infundibulum and isthmus, with prominent IgM and IgA labeling of Civatte bodies. There was also fibrin deposition at these sites. In contrast, Ioannides and Bystryn[18] showed linear deposits of IgG and IgA at the basement membrane zone of the follicular epithelium, absence of Civatte body staining, and rare fibrin deposition.

Lichen planopilaris can overlap with other scarring alopecias, and a diagnosis often cannot be made by clinical features alone.[22] The presence of acuminate follicular papules on the body, in addition to scarring alopecia, is a helpful clinical clue to the diagnosis. Biopsies of the scalp are helpful but often do not show characteristic changes of lichen planopilaris. Immunofluorescent microscopy findings also are often variable. The diagnosis of lichen planopilaris is best made by a combination of clinical and suggestive histologic and immunofluorescent findings.[6] Other disorders that present with scalp alopecia can mimic lichen planopilaris; for example, the early stages of chronic cutaneous lupus erythematosus can present solely as a scarring alopecia of the scalp.[1,20] It can be differentiated histologically[22] and through direct immunofluorescent microscopy studies, which demonstrate immunoglobulin deposits at the dermal-epidermal junction. Pseudopelade of Brocq is an ill-defined syndrome of slowly progressive cicatricial alopecia, without clinically evident folliculitis.[23] Some clinicians believe it represents an end stage of various diseases, such as discoid lupus erythematosus, scleroderma, or lichen planopilaris, whereas others believe it is a separate entity. Histologically, it has never been clearly defined and as such is a diagnosis of exclusion. Lichen planopilaris also should be differentiated from inflammatory folliculitis, follicular degeneration syndrome in African Americans, alopecia areata, cicatricial pemphigoid of Brunsting-Perry, and keratosis follicularis spinulosa decalvans (a genetic disorder with progressive scarring alopecia).[1,20]

Lichen planopilaris is a chronic and often progressive disease, with little potential for hair regrowth following follicular inflammation and destruction.[13] Most therapeutic reports are anecdotal, and controlled trials have not been performed. Generally, treatment is not satisfactory and mimics therapy for typical lichen planus.[8,13,14,24] A common, though not uniformly successful treatment regimen, includes systemic corticosteroids, alone or in combination with high-potency topical glucocorticoids.[6] Other agents such as retinoids, griseofulvin, intralesional steroids, and hydroxychloroquine also have been used, but with little success.[2,8,11,21] Two recent case reports have shown some efficacy with thalidomide or cyclosporine, but no controlled trials have been completed.[25,26] In most cases, once treatment is discontinued, there is often a relapsing course of the condition.[6]

## Conclusion

Lichen planopilaris is a condition with a spectral nature and may present with widespread truncal alopecia. It has yet to be determined whether lichen planopilaris is a variant of lichen planus or truly a distinct entity. Newer immunosuppressive agents show promise for the treatment of this skin disorder.

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