Lichen Planopilaris

Hong Jig Kim, M.D., Young Keun Kim, M.D., Tae Eun Kim, M.D.

Department of Dermatology, College of Medicine, Soonchunhyang University, Seoul, Korea

A 26-year-old Korean woman had a 5 month history of pruritic, keratotic papules on the scalp, trunk, and pubic area, which were accompanied by diffuse hair loss from the scalp, eyebrows and pubic area. She also had flat-topped violaceous papules on the scalp and pubic area, and white lace-like striae on the oral mucosa and hard palate.

Histopathologic study of the progressive stages of the disease revealed dilated follicles filled with horny material, hyperkeratosis, hypergranulosis and hydropic degeneration of the basal cell layer. Throughout the entire subepidermal region, there was an intense bandlike infiltration of mononuclear cells, which were most prominent at the lower pole of the hair follicles. The late stage of the disorder showed atrophy of the epidermis and atrophy of the hair follicles with fibrotic tracts, and mild mononuclear infiltrate at the sites of the former hair follicles.

This is the first recorded case, to the best of our knowledge, in Korea. (Ann Dermatol 1:51–54, 1989)

Key Words: Lichen planopilaris

Lichen planopilaris, which was first described by Pringle in 1895, is characterized by a triad consisting of lesions of lichen planus, spinous or acuminate lesions, and alopecia of the scalp or other hairy regions frequently showing atrophy.

The histopathologic features of lichen planopilaris are essentially those of lichen planus with particular localization of changes around the hair follicles.

We had the opportunity to observe a woman with extensive lichen planopilaris who presented diffuse hair loss of the scalp, eyebrows and pubic area, and follicular keratosis on the scalp, trunk, and pubic area. Typical lesions of lichen planus were seen on the oral mucous membranes, scalp, and the trunk, which were helpful in making the diagnosis of lichen planopilaris.

REPORT OF A CASE

A 26-year-old Korean woman developed a pruritic eruption of 5 month duration involving the scalp, trunk, and pubic area. Subsequently she extensive-ly lost hairs from the scalp, eyebrows, and pubic area. Her past medical and family histories were free of any pertinent medical problem.

On physical examination, her scalp and pubic area were almost completely barren of hair. Scattered over the trunk and the occipital region of the scalp were clusters of follicular spinous papules and flat-topped, violaceous papules (Fig. 1-3). Hairs in the affected areas were removed easily with gentle traction. Partial hair loss of the eyebrows was noted. However, the hairs of the axillae were not affected. There was keratosis pilaris on the upper arms and inner thighs which had occurred in childhood. There were several patches of white lace-like striae on the hard palate and buccal mucosa. The nails were normal.

The results of routine hematologic tests and serum chemical determinations were within normal limits. A chest x-ray showed active, chronic, and minimal tuberculosis.

The first biopsy specimens from the lichen planus-like papules of the scalp and pubic area showed dilated hair follicles containing keratotic plugs, hyperkeratosis, focal hypergranulosis, acanthosis, and hydropic degeneration of the basal cell layer or separation of the epidermis from the dermis. At the lower pole of the hair follicle was a dense mononuclear infiltrate (Fig. 4,5).
A second biopsy specimen from the scalp lesion taken six months later revealed atrophy of the epidermis, atrophy of the hair follicles with fibrotic tracts, and mild mononuclear infiltrate at the sites of the former hair follicles (Fig. 6). Direct immunofluorescent study revealed no specific finding.

Treatment with topical and systemic corticosteroids resulted in some flattening of the keratotic papules and decreasing pruritus, although alopecia persisted.

**DISCUSSION**

Since Pringle reported the association of lichen planus with follicular keratotic lesions, using the term 'lichen planopilaris' in 1895, there has been various speculation as to what clinically constitutes lichen planopilaris and whether or not it is an entity distinct from lichen planus.

The nomenclature previously suggested for this rare disorder—for example, "Graham Little syndrome", "Feldman's syndrome", and "lichen planopilaris", has been confusing.

Our patient, who has cicatricial scalp alopecia, follicular spinous lesions on the scalp, trunk, and
pubic area and noncicatricial alopecia of the pubic area, closely resembles the cases Graham Litte and Feldman described as “lichen spinulosus et folliculitis decalvans” and “lichen planus et acuminatus atrophicans”, respectively.

A review of the literature disclosed that the various names had essential features in common.

Clinically, the development of lichen planopilaris is usually an insidious process. Most of the patients give a history of gradual involvement over a period of several months (or even one or two years). In some the scalp may be affected first, as in our case, and in others the glabrous skin. Subjective symptoms are usually mild; however, our patient complained of severe pruritus. The clinical picture of this disease has been described as a triad consisting of lesions of lichen planus, spinous or follicular lesions, and alopecia of the scalp or other hairy regions frequently showing clinical atrophy.

The scarring alopecia has been confused with that of pseudopelade because specific papules of lichen planus are usually not apparent on examination. However, the cicatricial alopecia of our patient could be differentiated from that of pseudopelade, because she also had follicular keratotic lesions in the scalp, trunk, and pubic area. The flattened violaceous papules, which are typical lesions of lichen planus, on the occipital region of the scalp and the trunk, and white lace-like lesion of the oral cavity helped us confirm that lichen planopilaris is a follicular type of lichen planus; the presence of cutaneous follicular or spinous lesions is essential for the diagnosis, and frequently these coexist with typical cutaneous and mucosal lichen planus and with cicatrical alopecia of the scalp and noncicatricial alopecia of the axillary and/or pubic area, as in our patient.

The histopathologic picture of lichen planopilaris is essentially that of lichen planus, but the characteristic changes involve the follicles and perifollicular areas. At times, microscopic evidence of lichen planus in nonfollicular areas, in addition to lichen planopilaris, is present in the same sections. The histology shows a subepidermal bandlike infiltrate of mono-
nuclear cells, which were most prominent at the lower pole of the hair follicles and at the hair papillae. This damage to the hair papillae leads to destruction of the hair. At first the hair follicles are dilated and filled with plugs. Later on, the hair follicles and sebaceous glands disappear and fibrosis is seen in the dermis, which produces atrophic scarring. It is readily apparent that many difficulties exist in making a definitive diagnosis in the late stages of scarring alopecias. A histopathologic study of pseudopelade of Brocq has shown changes closely resembling the abnormalities of the end stage of various diseases including discoid lupus erythematosus, scleroderma, and lichen planopilaris.

Immunofluorescent findings of lichen planus have been extensively studied in the past decade. Characteristically, the subepidermal colloid bodies contain IgM, less frequently IgG, and rarely IgA, C3, and fibrinogen. Granular deposits of IgM and IgG in the basement membrane zone have also been reported. Like Fellner, we were unable to find any specific findings of immunofluorescence. However, Horn et al reported immunofluorescent findings of lichen planopilaris, which were similar to those of lichen planus, and supported the concept that lichen planopilaris is a variant of lichen planus.

In conclusion, the clinical picture of lichen planopilaris presents a combination of the following features: lesions of lichen planus, spinous or follicular lesions, and alopecia of the scalp, frequently showing clinical atrophy.

Among these features, the presence of cutaneous follicular lesions is essential for the diagnosis. The histopathologic picture of lichen planopilaris is that of lichen planus involving the follicle and perifollicular areas and is diagnostic for the disease. We wish to support the theory that lichen planopilaris, which is also called various other names but all have the essential features in common, is a follicular variant of lichen planus. Lichen planopilaris or follicular lichen planus is the name that most appropriately designates the clinical and histopathological features of the disease.

REFERENCES

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