CASE 23

Patient: A 57-year-old Thai female from Bangkok

Chief Complaint: Asymptomatic hair loss on frontal hair line and generalized hair thinning for 1 year

Present History: The patient presented with a 1 year history of asymptomatic hair loss on frontal hair line and generalized hair thinning. There was no loss of hair on the other parts of the body. She was treated at clinic with oral and topical medications without improvement.

Past History: She was in good health and denied taking any medication.

Family History: No one in her family has similar skin lesion.

Physical Examination: unremarkable

Dermatological Examination (Figure 23.1-2): Ill defined band-like atrophic alopecia patch with perifollicular erythema on frontal hair line with recession of frontal hair line. Hair pulling test positive.

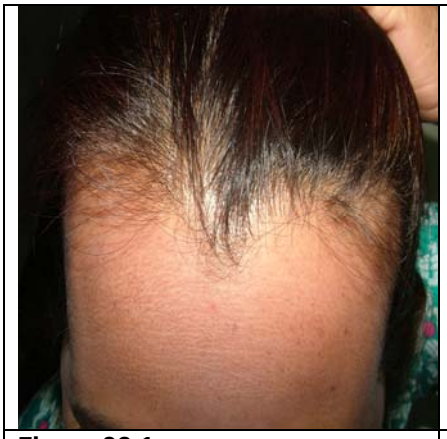

Figure 23.1

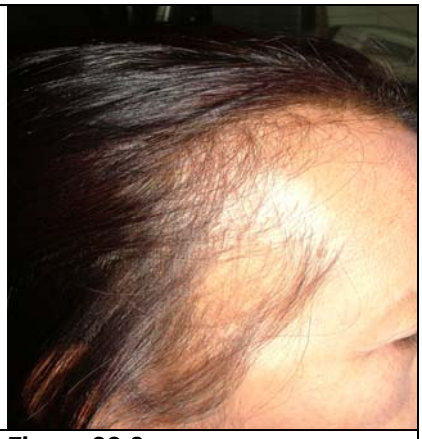

Figure 23.2

Histopathology (S09-18247) (Figure 23.3-4):

- Perifollicular lymphocytic infiltrate of fibrosis with follicular atrophy
- Some fibrotic tracts

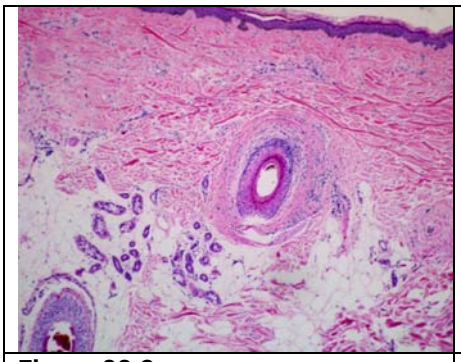

Figure 23.3

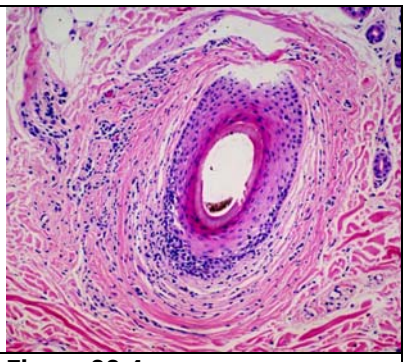

Figure 23.4

Diagnosis: Lichen planopilaris (Frontal fibrosing alopecia)

Treatment: 3% minoxidil apply bid
0.05% Clobetasol propionate scalp lotion bid
Intralesional triamcinolone acetonide (10mg/ml)

Presenter: Natta Tangkawattanakul

Consultant: Penpun Wattanakrai

Discussion:

Lichen planopilaris (LPP), also known as lichen follicularis or follicular lichen planus, was first described by Pringle in 1895.[1] It is an uncommon inflammatory hair loss disease characterized by autoreactive lymphocytic destruction of the hair follicle and progressive scarring alopecia of the scalp.[2] According to the cicatricial alopecia classification of the North American Hair Research Society, LPP is classified as a primary lymphocytic cicatricial alopecia.[3] LPP is the most frequent cause of adult primary scarring alopecia and is more

common in women (60 to 90% of the case) than men.[4-6] The age of onset is between 25-70 years. [4,7-8] This disease may develop alone or in association with other forms of lichen planus. The known incidence rate of LPP with lichen planus is variable according to reports, although 17-28% of LPP patients may have other forms of lichen planus.[7] LPP can be subdivided into 3 subgroups.

1. Classic lichen planopilaris
2. Frontal fibrosing alopecia
3. Graham-Little syndrome

The classical form of LPP commonly involves the vertex, but any region of the scalp can be affected. The early lesions are characterized by a follicular violaceous erythema and acuminate keratotic plugs commonly located at the periphery of the bald zone, corresponding to the margin of expanding areas of alopecia. Infrequently, pustules can be observed at the edges of the balding surface, which could mimic folliculitis decalvans. A positive pull test of anagen hairs is commonly present at the margin of alopecia, indicating the disease activity. After inflammation and hair shedding, atrophic scarring replaces the lesions. This corresponds to the end-stage of any cicatricial alopecia. Furthermore, these patches may coalesce to produce larger scarring patches, sometimes involving the entire scalp.[2] Most patients complain about symptoms of itching, pain, or burning when inflammation is present. This sensation is usually not intense and is aggravated by heat. The course of the disease is generally slowly progressive and well camouflaged by the remaining hair. [4-5] In other cases, a more rapid course may result in extensive hair loss within months.[9]

Histologically, a good biopsy needs to be done at the margin of an active, inflammatory lesion. A 4-mm punch is

the minimal necessary size.[10] In the active and early stage, histopathology shows a lichenoid interface inflammation with hypergranulosis, hyperkeratosis, hyperacanthosis, degeneration of basal keratinocytes and destruction of the basal layer in half cases.[10-11] A band-like subepidermal infiltrate of lymphocytes involving ("hugging") the follicles between the infundibulum and the isthmus, with sparing of the lower portion. Colloid bodies which are degenerated keratinocytes that stain pink with eosin are observed in the basal layer.[12] In late-stage lesions, fibrous tracts take the place of destroyed hair follicles and inflammation can be minimal or absent, with no distinctive lichenoid changes.

Frontal fibrosing alopecia, first described by Kossard in 1994[13], is a progressive band-like scarring alopecia mostly confined to the frontal hair line. Postmenopausal woman are predominantly affected though it can occur occasionally in premenopausal women and men.[14] The characteristic clinical findings in FFA include: follicular hyperkeratosis with perifollicular erythema, progressive recession of frontal and temporal hair line, and loss of follicular ostia. 52% of FFA patients also have eyebrow hair loss.[15] In contrast to classic LPP, FFA has no multifocal areas of scarring alopecia and is very rarely associated with lesions of lichen planus.

Graham-Little syndrome (also known as Graham-Little-Piccardi-Lassueur syndrome) is an uncommon condition of adults, characterized by scarring patchy alopecia of the scalp, noncicatricial axillary and pubic hair loss and lichenoid follicular eruption on the trunk and extremities.The scalp alopecia may develop at any time during the course of the disease and is marked by follicular hyperkeratosis or erythematous scaly patches. There are infrequent reports of face and eyebrow involvement. Most of the patients described are women ages 30 to 60 years. The syndrome can be associated with an eruption

of LP or LPP of the body hair, especially of the pubic and axillary regions.[2]

Treatment of LPP:

- The first-line therapy is ultra-potent topical corticosteroids or monthly intralesional injections of triamcinolone acetonide(3-10 mg/ml) in cases of local and severe inflammation.[2, 4]
- The second-line therapy with systemic oral corticosteroids; prednisolone 1 mg/kg/day for 15 days, tapered over 4 months can help stop rapid hair loss. A relapse after ceasing treatment is common.[2]
- The third-line therapy includes cyclosporine[2, 16], mycophenolate mofetil[17,18], Hydroxychloroquine[20] and topical 2% or 5% minoxidil lotion[2].

Our patient's her age and clinical features of band-like atrophic alopecia patch with perifollicular erythema and recession of frontal hair line were compatible with the frontal fibrosing alopecia subgroup of LPP. She was treated with intralesional triamcinolone acetonide, topical 0.05% Clobetasol propionate scalp lotion and topical 3%minoxidil.

References

1. Pringle JJ. Cited by Adamson HG. Lichen pilaris spinulosus. Br J Dermatol 1905; 17:78-9.
2. Assouly P, Reygagne P. Lichen planopilaris: update on diagnosis and treatment. Semin Cutan Med Surg 2009;28:3-10.
3. Olsen EA, Bergfeld WF, Cotsarelis G, et al. Summary of North American Hair Research Society (NAHRS)-sponsored Workshop on Cicatricial Alopecia, Duke University Medical Center, February 10 and 11, 2001. J Am Acad Dermatol 2003;48:103-10.
4. Chieregato C, Zini A, Barba A, Magnanini M, Rosina P. Lichen planopilaris: report of 30 cases and review of the literature. Int J Dermatol 2003;42:342-5.
5. Cevasco NC, Bergfeld WF, Remzi BK, de Knott HR. A case-series of 29 patients with lichen planopilaris: the Cleveland Clinic Foundation

experience on evaluation, diagnosis, and treatment. J Am Acad Dermatol 2007;57:47-53.
6. Kang H, Alzolibani AA, Otberg N, Shapiro J. Lichen planopilaris. Dermatol Ther 2008;21:249-56.
7. Tan E, Martinka M, Ball N, Shapiro J. Primary cicatricial alopecias: clinicopathology of 112 cases. J Am Acad Dermatol 2004;50:25-32.
8. Handa S, Sahoo B. Childhood lichen planus: a study of 87 cases. Int J Dermatol 2002;41:423-7.
9. Ross EK, Tan E, Shapiro J. Update on primary cicatricial alopecias. J Am Acad Dermatol 2005;53:1-37; quiz 8-40.
10. Sperling LC. Scarring alopecia and the dermatopathologist. J Cutan Pathol 2001;28:333-42.
11. Tandon YK, Somani N, Cevasco NC, Bergfeld WF. A histologic review of 27 patients with lichen planopilaris. J Am Acad Dermatol 2008;59:91-8.
12. Mehregan DA, Van Hale HM, Muller SA. Lichen planopilaris: clinical and pathologic study of forty-five patients. J Am Acad Dermatol 1992;27:935-42.
13. Kossard S. Postmenopausal frontal fibrosing alopecia. Scarring alopecia in a pattern distribution. Arch Dermatol 1994;130:770-4.
14. Faulkner CF, Wilson NJ, Jones SK. Frontal fibrosing alopecia associated with cutaneous lichen planus in a premenopausal woman. Australas J Dermatol 2002;43:65-7.
15. Naz E, Vidaurrazaga C, Hernandez-Cano N, et al. Postmenopausal frontal fibrosing alopecia. Clin Exp Dermatol 2003;28:25-7.
16. Mirmirani P, Willey A, Price VH. Short course of oral cyclosporine in lichen planopilaris. J Am Acad Dermatol 2003;49:667-71.
17. Tursen U, Api H, Kaya T, Ikizoglu G. Treatment of lichen planopilaris with mycophenolate mofetil. Dermatol Online J 2004;10:24.
18. Cho BK, Sah D, Chwalek J, et al. Efficacy and safety of mycophenolate mofetil for lichen planopilaris. J Am Acad Dermatol 2010;62:393-7.
19. Moreno-Ramirez D, Camacho Martinez F. Frontal fibrosing alopecia: a survey in 16 patients. J Eur Acad Dermatol Venereol 2005;19:700-5.
20. Chiang C, Sah D, Cho BK, Ochoa BE, Price VH. Hydroxychloroquine and lichen planopilaris: efficacy and introduction of Lichen Planopilaris Activity Index scoring system. J Am Acad Dermatol 2010;62:387-92.