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Lichen Planopilaris Presenting with Different Morphologies: 3 Case Reports

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Abstract

Lichen planopilaris is a primary cicatricial alopecia involving hair follicles with a lymphocytic inflammatory process destroying the follicles. Clinically it presents with grouped keratotic follicular papules surrounded by erythema. It is classified as classic LPP, frontal fibrosing alopecia (FFA), and Lassueur Graham-Little Piccardi syndrome. Pseudopelade of Brocq (PPB), can be considered as end-stage LPP which leaves clinical appearance of "footprints in the snow" appearance. Scarring alopecia represents a challenge to patients as well as treating dermatologist where early diagnosis is essential as treatment is difficult in late stage disease. Dermatoscopy accompanied by biopsy is indicated in such cases to start the treatment at the earliest. Three cases of lichen planopilaris with different morphologies is presented here.

Keywords: Lichen Planopilaris; Scarring Alopecia; Pseudopelade of Brocq.

Introduction

Lichen planopilaris (LPP) is a chronic cutaneous disorder selectively involving hair follicles with a lymphocytic inflammatory process that gradually destroys the follicle; thus, called as a primary cicatricial alopecia. It is a disease of unknown etiology, whose pathogenesis is poorly understood, despite a suspected autoimmune origin.[1]

LPP usually affects young adult females, although the age range is wide and can also affects males.[2] It commonly develops in association with lichen planus affecting the skin, mucosa and nails. Clinically it presents with grouped keratotic follicular papules surrounded by erythema in the initial stage which can be confirmed by histological examination, however a late disease has no specific signs, thus making it difficult to separate from other scarring scalp conditions even with the histopathological examination.[3] Early diagnosis is required for early initiation of therapy and halt the progress of the disease.

Case reports

Case 1

A female aged 36 years presented with hair loss and itching over scalp for last 1 year. Of alopecia with atrophy and shiny skin involving parietal scalp was present with typical foot print in snow appearance [Fig. 1a]. No other parts of body, nails or oral mucosa wear affected. Dermatoscopic findings showed lack of follicular orifices and perifollicular scales [Fig. 1b]. Histopathology showed hyperkeratosis, acanthosis and orthokeratosis in epidermis. Dermis showed perifollicular lymphocytic infilterate, periappendegeal and perivascular inflammatory infilterate [Fig. 1c]. Changes were suggestive of lichen planopilaris.

Case 2

A 38 year old male presented with single lesion over scalp for last 5 months. Lesion was itchy and progressive in nature. Patient was tchronic tobacco chewer having tobacco for last 15 years. On

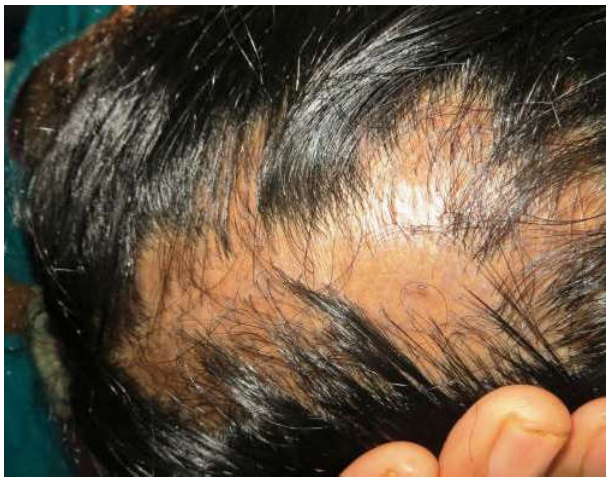

Fig. 1a: Atrophic patches of alopecia involving parietal scalp with typical foot print in snow appearance.

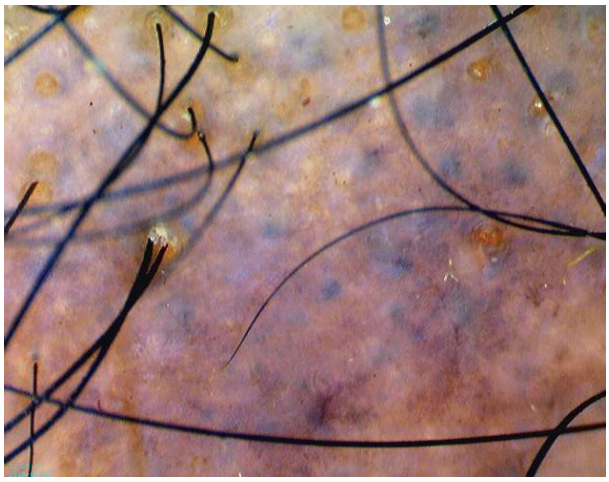

Fig. 1b: Dermatoscopic showing lack of follicular orifices and perifollicular scales.

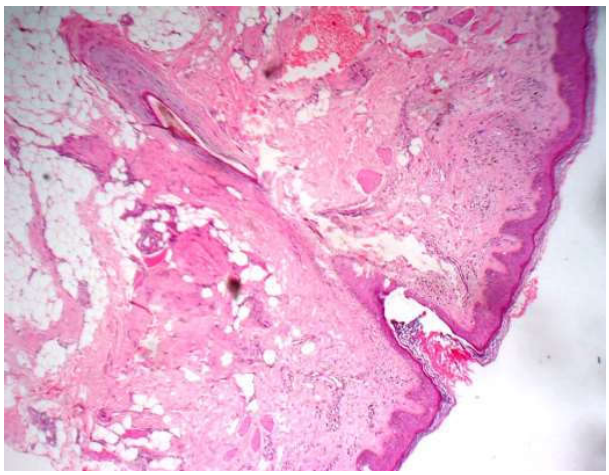

Fig. 1c: Hyperkeratosis, acanthosis, and orthokeratosis in epidermis with periappendegeal and perivascular inflammatory infilterate in dermis. [H&E stain 4X].

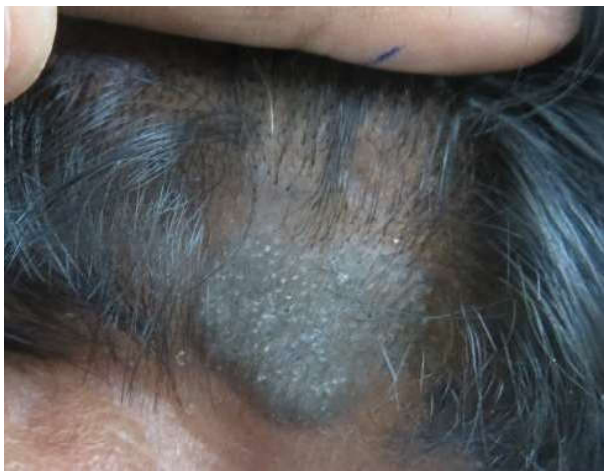

Fig. 2a: Single well defined hyperpigmented plaque with follicular plugging over left side of occiput.

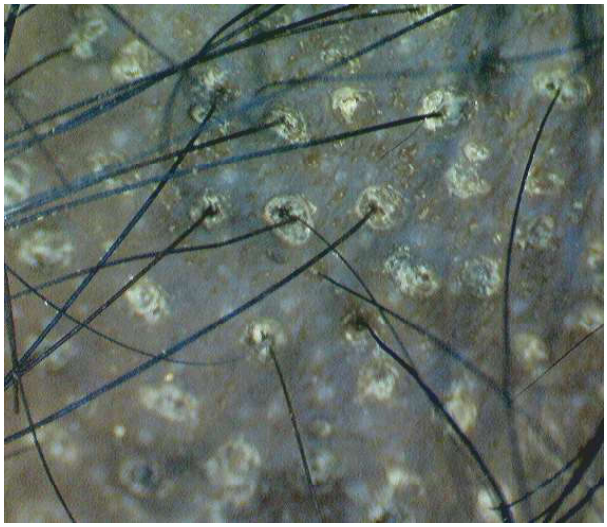

Fig. 2b: Dermatoscopy showing pigmentation, and perifollicular scales, follicular plugging and reduced follicular ostia.

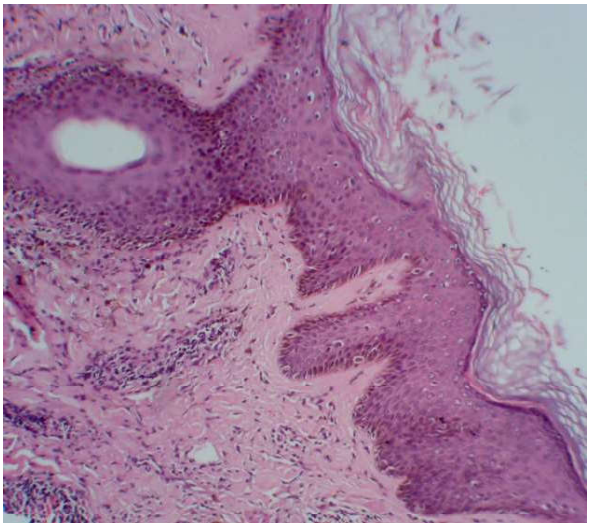

Fig. 2c: Follicular plugging, hyperkeratosis, focal wedge shaped hypergranulosis, elongated rete ridges and focally dense band like perifollicular lymphocytic infilteration at the base of rete ridges and infundibulum. [H&E stain 4X]

Fig. 3a: Single well defined hyperpigmented plaque with follicular plugging over vertex.

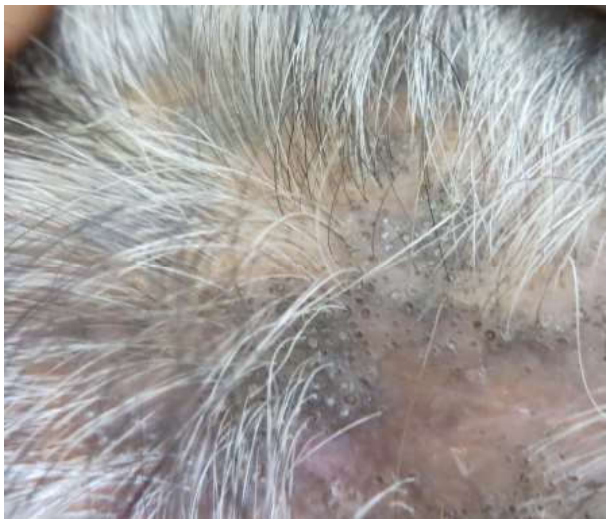

Fig. 3b: Multiple patches of scarring alopecia over parietal and occipital regions.

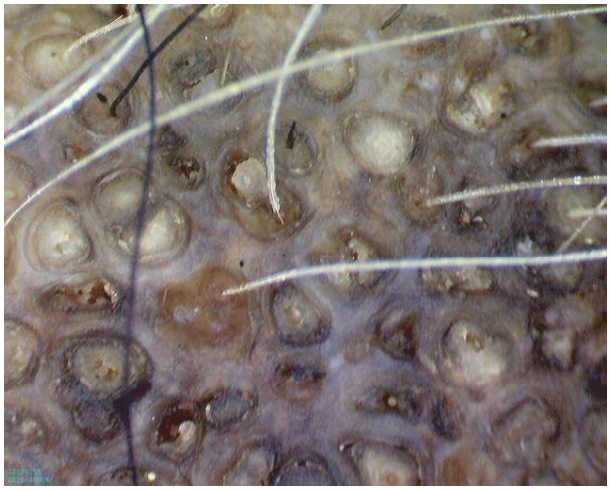

Fig. 3c: Dermatoscopy showed reduced follicular orifices, white dots as well as perifollicular scale.

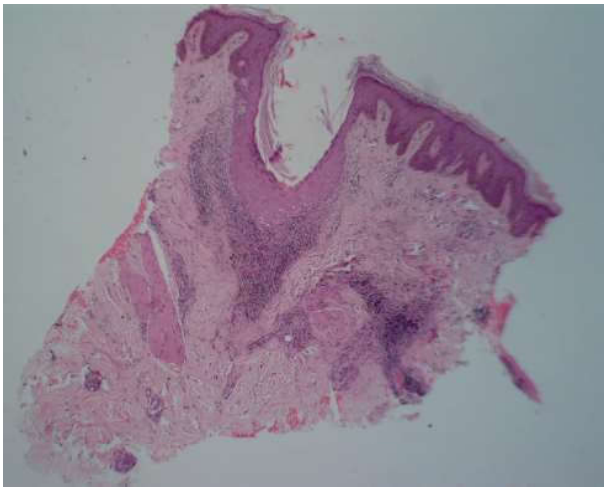

Fig. 3d: Follicular plugging and intense lymphocytic inflammatory infiltrate in perifollicular region. [H&E stain 4X].

examination, single well defined hyperpigmented plaque with follicular plugging was present over left left side of occiput [Fig. 2a]. Violaceous pigmentation was present over bilateral buccal mucosa. Nails showed clubbing and longitudinal melanonychia. No other areas of body were affected. Dermatoscopy showed pigmentation, perifollicular scale, follicular plugging and reduced follicular ostia [Fig. 2b]. Biopsy showed follicular plugging, hyperkeratosis, focal wedge shaped hypergranulosis, elongated rete ridges and focally dense band like perifollicular lymphocytic infilteration at the base of rete ridges and at the level of infundibulum [Fig. 2c]. Changes were suggestive of LPP.

Case 3

A 65 yr old male presented with multiple patches of hair loss over scalp for last 1 month. Lesions were progressive in nature but non itchy. On examination, single well defined hyperpigmented plaque with follicular plugging over vertex and multiple patches of scarring alopecia over parietal and occipital regions were present [Fig. 3a&b]. Dermatoscopy showed reduced follicular orifices, white dots and perifollicular scales [Fig. 3c]. Histopathological examination showed follicular plugging and intense lymphocytic inflammatory infiltrate in perifollicular region [Fig. 3d]. No evidence of any other body parts ie skin, nails or mucosal involvement was seen. Dermatoscopic and Histopathology finding showed changes of lichen planopilaris.

Patients were given topical steroids and tacrolimus 0.1% cream to applied.

Discussion

LPP is also known as lichen follicularis or follicular lichen planus, and was initially described by Pringle in 1895.[4] It is an uncommon inflammatory scalp disorder that is clinically characterized by perifollicular erythema, follicular hyperkeratosis, and permanent hair loss. It is considered a follicular variant of lichen planus. According to the NAHRS (North American Hair Research Society) classification, LPP has been subdivided into 3 variants: classic LPP, frontal fibrosing alopecia (FFA), and Lassueur Graham-Little Piccardi syndrome.[5]

LPP is more common in females (60 to 90%) than in males[6,7] and the age of onset of LPP is frequently between 40 and 60 years.[8] Two of the patients were males while one was female with 2 in their thirties and one with 65 years of age. Endogenous or exogenous agents such as drugs, viruses or contact sensitizers have been proposed as potential triggers for the autoimmune response observed in LPP[6].

Symptomatically, it presents with itching, pain or burning when inflammation is present, aggravated by heat and also associated with seborrheic dermatitis but not pathognomic for the disease. The early lesions are characterised by a follicular violaceous erythema and keratotic plugs, commonly located at the periphery of expanding areas of alopecia. Some hairs affected by the inflammation process can persist in the centre of the bald area with some tufted hairs. A positive pull test of anagen hairs is commonly present at the margin of alopecia, which indicates disease activity. Atrophic scarring without follicular units is seen in late stage of LPP without any typical papules of lichen planus on the scalp, as was seen in 1st case.

Parietal and fronto-vertical regions are the most frequently involved part of the scalp.[9]

Scarring alopecia of the scalp can be associated with other regional involvements in 17–28% patients in one study,[6] while 50%[9] in another study. Lesions of typical lichen planus over nonfollicular skin, mucous membranes, and nails are present in only 50% of cases and usually follow the onset of cicatricial alopecia.[7] Two of the patients didn't showed involvement of any other site while case 2 had oral involvement in the form of purplish pigmentation over bilateral buccal mucosa and nail changes showing clubbing and longitudinal melanonychia. This can be attributed to the habit of tobacco chewing in him since last 15 years.

Patients may present with prominent and characteristic multifocal irregular areas of patchy scarring hair loss resembling moth eaten alopecia which is similar to secondary syphilis and alopecia areata.[10] The presence of small asymptomatic truncal follicular papules is not noticed by the patients unless closely looked for by the dermatologist, so careful examination of all body regions are mandatory in evaluation of scarring scalp conditions.

The main diagnostic challenge for LPP is DLE, thus making it difficult to separate from other scarring scalp conditions even with the histopathological examination. However there are many clinical and histopathological points which can help the dermatologists to differentiate between these two primary lymphocytic cicatricial conditions.[11]

Pseudopelade of Brocq (PPB), can be considered as end-stage LPP, may leave the clinical appearance of "footprints in the snow" as presented by Case 1. If most of the lesions of PPB are old and the LPP is not expanding with perifollicular inflammation and hyperkeratosis, it is impossible to distinguish PPB from LPP. Dermoscopy shows lack of follicular orifices in the centre of bald areas and on the margin, the pink/red translucent inflammation is clearly perifollicular, with keratin surrounding and extending along the proximal part of the hair shafts.

Treatment is difficult and response with most of the drugs is poor. Topical corticosteroids and intralesional corticosteroid injection are commonly utilized as first-line therapy for LPP.[12]

Conclusion

Scarring alopecia represents a challenge to patients as well as treating dermatologist where early diagnosis is essential for initiation of an early and effective therapy to save the hair follicles from the irreversible damage.This can be done by dermatoscopy supplemented by histopathology if required.

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