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Case and Review

Diffuse Lichen Planopilaris Masquerading as Diffuse Alopecia Areata

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Alopecia areata · Lichen planopilaris · Androgenetic alopecia

Abstract
Introduction: Lichen planopilaris (LPP) is a primary lymphocytic cicatricial alopecia that represents a form of follicular lichen planus. ***Case Presentation:*** We describe a case of coexisting diffuse LPP and female pattern hair loss masquerading as diffuse alopecia areata in a 32-year-old female. ***Discussion:*** In complex cases such as this, dermoscopy-guided vertical and horizontal biopsies from androgen sensitive and insensitive areas are helpful in increasing diagnostic yield. Prompt initiation of treatment is key to halting disease progression. Long-term follow-up is important as resolution of clinical signs does not always correlate with the absence of disease progression.

Introduction

Lichen planopilaris (LPP) is a primary lymphocytic cicatricial alopecia that represents a form of follicular lichen planus [1]. LPP preferentially affects women, typically between ages 30 and 60 [2, 3]. Six variants have been described and are distinguished primarily based on the clinical distribution of alopecia. These variants are classic LPP, frontal fibrosing alopecia (FFA), Graham-Little-Piccardi-Lassueuer syndrome, fibrosing alopecia in a pattern distribution (FAPD), cicatricial pattern hair loss (CPHL), and LPP diffuse pattern (LPPDP) [1–6].

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Case Report

We present an atypical case of diffuse LPP masquerading as diffuse alopecia areata (AA). A 32-year-old woman presented for a second opinion regarding the treatment of AA. She reported a 10-year history of gradual hair thinning with a 3-year history of worsening diffuse scalp hair loss (shown in Fig. 1a, b) associated with complete madarosis and thinning of axillary and pubic hair. There was no personal or familial history of skin or hair diseases. Examination revealed diffuse hair thinning. Trichoscopy confirmed sporadic white dots, loss of follicular ostia, hair miniaturisation, and minimal perifollicular hyperkeratosis (shown in Fig. 1c, d). Perifollicular erythema, exclamation hair marks, and black dots were absent. Examination of the nails and oral mucosa was unremarkable. Given the diagnostic dilemma, four trichoscopy-guided horizontal and vertical biopsies were taken from androgen sensitive and insensitive areas. These showed fibrous tracts and perifollicular lichenoid lymphocytic infiltrate, suggestive of LPP (shown in Fig. 2). The background of a 10-year history of gradual ponytail thinning and hair miniaturisation also supports a coexisting diagnosis of female pattern hair loss. She was treated with oral corticosteroids, minoxidil, and spironolactone.

Discussion

Classic LPP presents with focal or multifocal alopecia. Frontotemporal hairline recession is characteristic of FFA, typically with eyebrows and less so with eyelash involvement. Occipital scalp and peripheral body hair can also be affected, while facial papules are also seen in a minority of patients. Eyelash loss, facial papules, and body hair involvement have been shown to be associated with severe FFA, while eyebrow loss is associated with mild forms [7]. Graham-Little-Piccardi-Lassueuer syndrome is a triad of scarring alopecia of the scalp, non-scarring alopecia of the axillary and pubic areas, and lichenoid papules on the trunk and extremities [1]. FAPD presents as central scarring alopecia with perifollicular erythema and follicular hyperkeratosis with histologic features of LPP and androgenetic alopecia [1]. FAPD was unlikely in this case as she had diffuse hair loss which also affected androgen-independent areas, and there was an absence of perifollicular erythema [8]. CPHL presents with features of female pattern hair loss and focal atrichia, histologic features similar to FAPD but lacking follicular erythema and hyperkeratosis [1]. This patient presented with diffuse hair thinning without a specific female pattern distribution associated with complete madarosis and loss of axillary and pubic hair, in keeping with LPPDP.

In a retrospective study of 20 patients with LPPDP, mild-moderate itch and pain of the scalp were found in all patients, but our patient had minimal to no symptoms [1]. Trichoscopy findings including perifollicular erythema and hyperkeratosis were also found to be pathognomic in this case series [1]. The lack of symptoms and trichoscopy findings in our patient suggest the chronicity of her disease. The complexity and atypical nature of our patient's presentation prompted dermoscopy-guided biopsies in locations with peripilar casts/peripilar white halo which can yield pathological diagnosis in 95% of cases [8, 9].

AA can be diagnosed by vertical or horizontal biopsy; however, when chronic, a peribulbar infiltrate may be missing. Hence, horizontal biopsy will show follicular counts revealing reduced terminal anagen, increased terminal catagen and telogen hairs, and miniaturised vellus hairs and follicular stelae. Additionally, reduced follicular count suggests poor prognosis [10]. In cicatricial alopecia, vertical section is advantageous, as it gives full view of the skin and hair, showing peribulbar inflammation and fibrosis [11]. In LPP, the focus of the lymphocytic infiltrate is the terminal follicle, with the miniaturised follicles spared. In FAPD and CPHL; however, predominantly miniaturised follicles are affected [1]. As such, in a case

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Fig. 1. a Hair thinning and loss affecting the vertex (androgen-dependent area). **b** Hair thinning and loss affecting the occipital scalp (androgen-independent area). **c** Trichosopy of an androgen-dependent site: loss of follicular ostia, white dots, miniaturised hair. **d** Trichoscopy of androgen-independent site: one peripilar scale.

where there is potentially chronic AA, a horizontal biopsy is preferred, while in LPP, vertical is preferred. Hence, to maximise accuracy in complex cases and increase diagnostic yield since the number of targeted follicles and the intensity of infiltrate are lower in LPPDP compared to classic LPP, vertical and horizontal biopsies are taken.

The aim of treatment is to halt disease progression as uncontrolled LPP results in irreversible hair loss, and hair regrowth is not expected unless treatment is started early [5]. Evidence to guide therapy is extremely limited as LPPDP is a newly diagnosed condition. In the retrospective case series described above, that included 20 LPPDP patients, disease progression was reported to have been arrested in 95% of cases after a year through combinations of intramuscular triamcinolone (14; 6–8 month course), hydroxychloroquine (8; 400 mg/day), clobetasol propionate (15; 0.05% cream), and topical minoxidil (20; 2/5%) [1].

It is, however, worth noting that the cicatricial alopecias can progress slowly and that a 12-month period may be insufficient to monitor for progression. The diffuse nature of LPPDP makes it even more challenging to monitor for disease progression. The LPP activity index (LPPAI) tool may be utilised, but subtle signs of progression may not be fully captured [12]. It should also be noted from the LPP literature that hydroxychloroquine may simply reduce the

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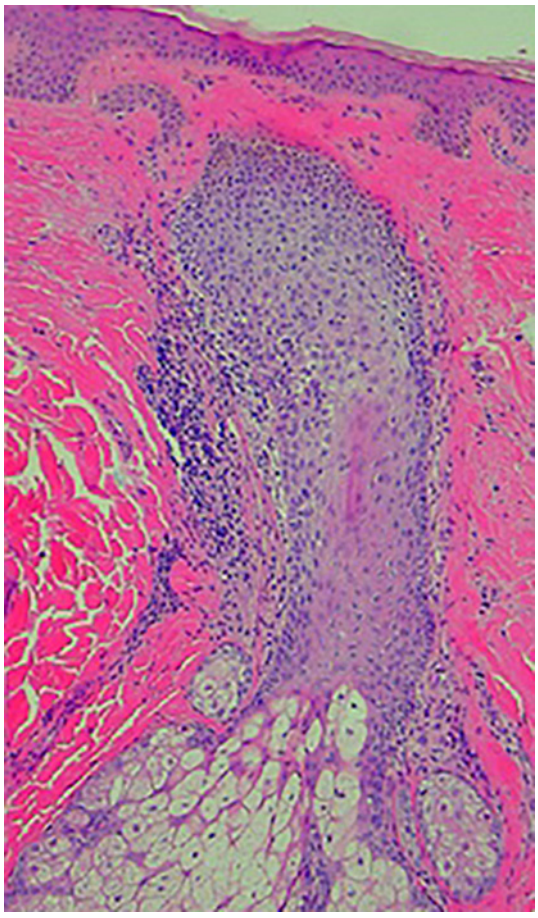

Fig. 2. Mid-power view (×10) of a punch biopsy (vertical section) showing a dense lichenoid lymphocytic infiltrate of the follicular epithelium.

symptoms and signs associated with activity without truly halting hair loss [13]. Hence, long-term follow-up is important as resolution of clinical signs does not always correlate with the absence of disease progression.

We highlight the importance of considering LPPDP when there is diffuse hair loss affecting the scalp and body, not only to halt the progression of a scarring alopecia but also to council patients regarding potential disease outcomes and avoid misdirected therapies. Dermoscopy-guided vertical and horizontal biopsies from androgen sensitive and insensitive areas are helpful in diagnosing these complex cases. The CARE Checklist has been completed by the authors for this case report, attached as online supplementary material (for all online suppl. material, see https://doi.org/10.1159/000538064).

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Statement of Ethics

Ethical approval is not required for this study in accordance with local or national guidelines.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

L.J.H.Y.: conceptualization, data curation, formal analysis, writing – original draft. N.M., D.W., and I.M.: formal analysis, supervision, validation, writing – review and editing.

Data Availability Statement

All data generated or analysed during this study are included in this article and its online supplementary material. Further enquiries can be directed to the corresponding author.

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