Rapidly progressive diffuse fibrosing alopecia

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Key words: alopecia; baldness; cicatricial pattern hair loss; fibrosing alopecia; fibrosing alopecia with a pattern distribution; frontal fibrosing alopecia; graft versus host disease; hair loss; lichen planopilaris.

INTRODUCTION

Generally, cicatricial alopecias are relentlessly and slowly progressive. We describe clinical and histologic features of rapidly progressive cicatricial alopecia in a young man, with overlapping features of frontal fibrosing alopecia (FFA), lichen planopilaris (LPP), fibrosing alopecia with a pattern distribution (FAPD), and cicatricial pattern hair loss (CPHL), suggesting that these conditions may exist along a spectrum.

Abbreviations used:

CPHL:	cicatricial pattern hair loss
FAPD:	fibrosing alopecia with a pattern distribution
FFA:	frontal fibrosing alopecia
GvHD:	graft-versus-host disease
LPP:	lichen planopilaris

CASE REPORT

A 25-year-old man presented with progressive hair thinning for 3 years in July 2017. Physical and global examination showed that although scalp hair thinning was not very apparent at that time (Fig 1, *A*), small patches of hair loss similar to the pencil-eraser–sized areas of focal atrichia described by Olsen[1] were observed (Fig 1, *B*). The eyebrows, beard, armpit hair and pubic hair were sparse, and vellus hair of the limbs was absent. There were no lesions typical of lichen planus found elsewhere. Dermoscopic examination showed confluent white dots and loss of follicular ostia but no prominent hair miniaturization or high ratio of vellus hair (Fig 1, *C*). Results of complete blood count, liver function tests, free testosterone, syphilis IgG, thyroid function tests, antinuclear factor, and rheumatoid factor were either normal or negative. A scalp biopsy from the temporal aspect of the right scalp showed lichenoid folliculitis and concentric fibrosis around both the isthmus and infundibulum, with some follicles destroyed and replaced by connective tissue (Fig 1, *D* and *E*). Given the clinical manifestations and dermoscopic and histopathologic examination findings, a diagnosis of CPHL was rendered. The patient was treated with oral glycyrrhizin capsules, topical tacrolimus, and halometasone.

One year later, the patient presented again with significant hair thinning and numerous small patches of hair loss. Alopecia now involved the entire scalp, including bilateral temporal areas, occipital scalp, and hairline (Fig 2, *A* and *B*). Prominent perifollicular keratosis was now present, most prominently in the apical scalp and forehead regions (Fig 2, *C*). Perifollicular erythema was absent, and the patient denied abnormal sensation or other inflammatory symptoms. Dermoscopic examination showed evident loss of follicular ostia and plentiful confluent irregular white dots (Fig 2, *D*).

TrichoScan (Tricholog, Freiburg, Germany), an objective, noninvasive, and non–observer-dependent technology,[2] was used to quantify the percentages of vellus and terminal hairs. The absence of significant hair miniaturization was confirmed, and the percentage of vellus hairs was only 5.1% (not shown). Biopsy specimens from the frontal,

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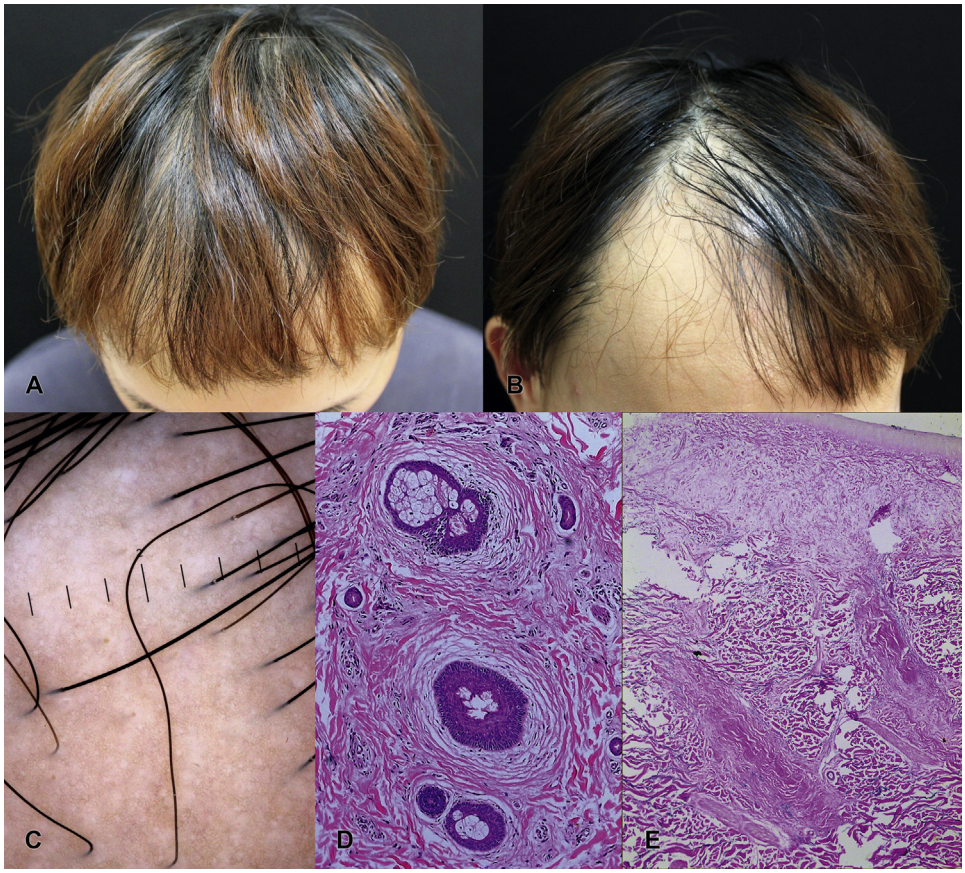

Fig 1. **A**, Hair thinning was not very apparent. **B**, Small patches of hair loss can be observed. **C**, Confluent white dots and loss of follicular ostia are noted, but no prominent hair miniaturization or high ratio of vellus hair is observed. **D** and **E**, Lichenoid folliculitis and concentric fibrosis around both the isthmus and infundibulum, with some follicles destroyed and replaced by connective tissue. Original magnifications, × 100. **D**, H&E stain. **E**, Victorian blue van Gieson stain.

temporal, and occipital scalp showed similar histopathologic changes, with more prominent lymphocyte infiltration around the isthmus and infundibulum compared with the biopsy a year earlier, and the number of follicles had significantly decreased, with some follicles destroyed and some telogen follicles remaining (Fig 3, *A-C*).

The clinical manifestations and dermoscopic and histopathologic examination show overlapping features with FFA, LPP, FAPD, and CPHL. Overlapping feature and differences are noted in Table I. Given the diffuse small patches of hair loss and histopathologic changes of lichenoid folliculitis and concentric fibrosis, we prefer the designation *diffuse fibrosing alopecia*. Intralesional Diprospan injection (Schering-Plough, Heist-op-den-Berg, Belgium), oral hydroxychloroquine, and tranilast were prescribed, together with topical tacrolimus and halometasone. The appearance of the scalp remained stable over the next 6 months, and regrowth of eyebrows was observed.

DISCUSSION

FFA and FAPD are both classified as subtypes of LPP by many authorities,[3,4] although this classification is still debatable.[5] CPHL, as described by Olsen,[1] is also a relatively new entity similar to FAPD but lacking the perifollicular erythema and perifollicular keratosis seen in FAPD. A relatively specific sign of CPHL is the presence of pencil-eraser–sized areas of patchy scarring, generally affecting women older than 40 years of age.

Recently, it has been proposed that LPP may result from a collapse of the hair follicle's immune privilege.[6] Triggering agents may include drug reactions, viral hepatitis, and cutaneous graft-versus-host disease (GvHD). Moreover, at least some examples of

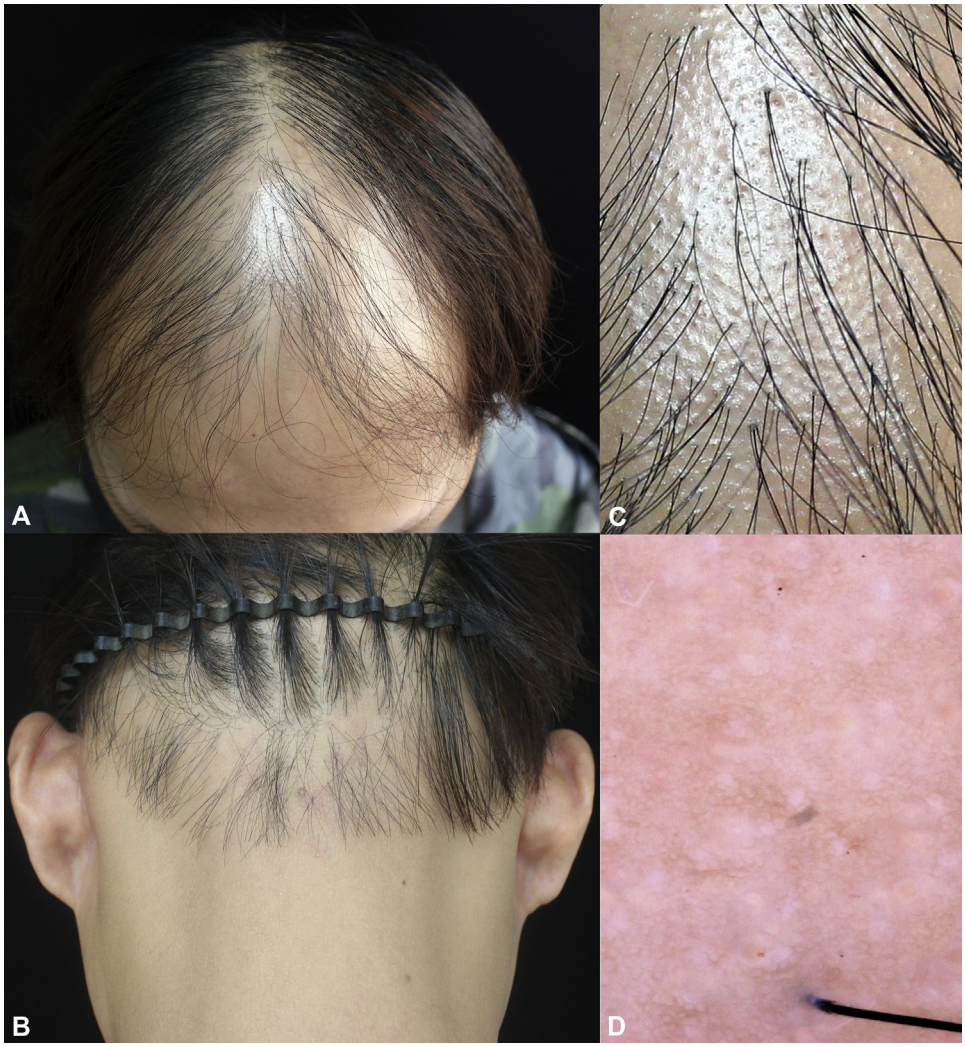

Fig 2. **A** and **B**, Significant hair thinning involved the entire scalp; vellus hair was absent, and there was no prominent hair miniaturization. **C**, Perifollicular keratosis was prominent in the apical scalp and forehead regions. **D**, Evident loss of follicular ostia and confluent irregular white dots were seen.

FAPD may represent a unique presentation of GvHD.[7,8]

As more attention is paid to alopecia subtypes, FAPD and CPHL are likely to be reported more often. FFA, once considered an uncommon condition, has now become common.[9] There remains much overlap in the spectrum of these entities,[10] and because the pathogenesis has not been clarified, distinctions between them remain speculative.[1]

There is wide overlap between FFA, LPP, FAPD, and CPHL, both clinically and histopathologically, and we prefer to use the general term *fibrosing alopecia* to refer to these cicatricial entities that share lichenoid folliculitis and fibrosis that mainly involves the upper portion of the follicle. We suggest that the disorders lie on a spectrum, where LPP represents patchy disease, FFA involves a bandlike area of the hairline, CPHL and FAPD involve androgen-dependent areas, and diffuse fibrosing alopecia represents the widespread counterpart. Varied presentations may relate to hormonal effects and other underlying triggers such as GvHD, and further studies are required to reveal the exact pathogenesis.

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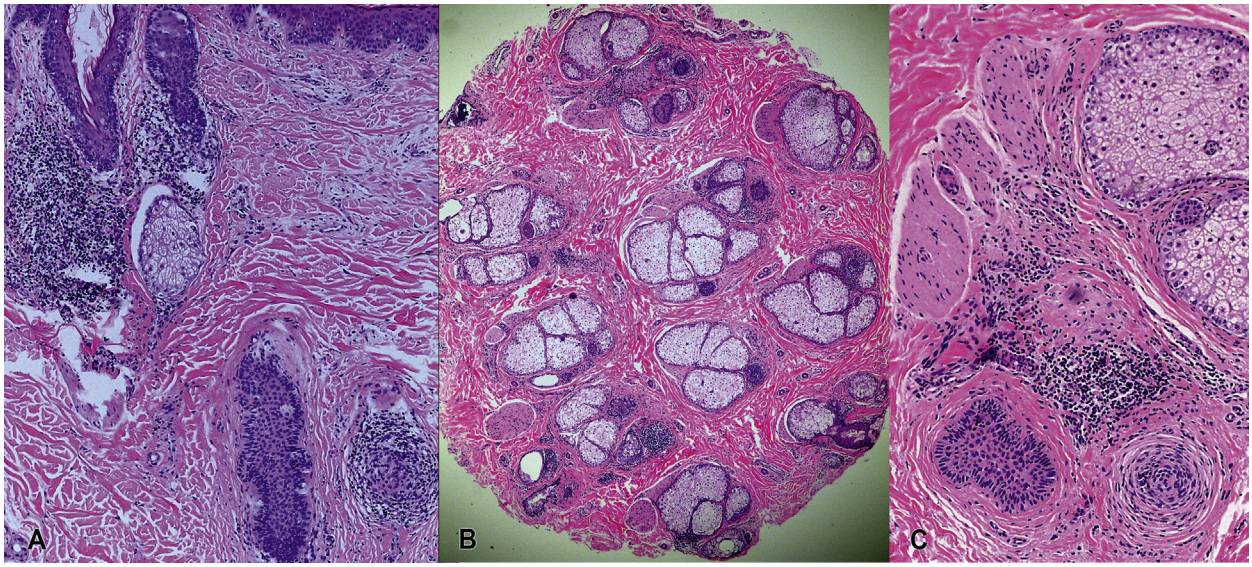

Fig 3. A-C, Dense lymphocyte infiltration around the isthmus and infundibulum and decreased number of follicles, with some telogen follicles remaining and some follicles destroyed. H&E stain. Original magnifications: **A**, × 100; **B**, × 40; **C**, × 200.

Table I. Comparison between FFA, LPP, CPHL, FAPD and the present case

Characteristic	FFA	LPP	CPHL	FAPD	Present case
Susceptible group	Mostly postmenopausal women	Mostly middle-aged women	Mostly women older than 40 years	Mostly older women	Young man
Location	Anterior hairline, temples	Mostly scattered	Androgen-dependent areas	Androgen-dependent areas	Diffuse
Pattern	Bandlike	Patchy	Pencil-eraser sized	Pattern distribution	Pencil-eraser sized
Other nonscalp involvement	Yes	Yes	No	No	Yes
Perifollicular keratosis	Yes	Yes	No	Yes	Yes
Perifollicular erythema	Yes	Yes	No	Yes	No
Hair miniaturization	No	No	May be present	Mostly yes	No
Slowly progressive	Mostly yes	Mostly yes	Mostly yes	Mostly yes	No
Histologically involved portion	Isthmus, infundibulum	Isthmus, infundibulum	Isthmus, infundibulum	Isthmus, infundibulum	Isthmus, infundibulum
Lichenoid folliculitis	Yes	Yes	Yes	Yes	Yes
Concentric fibrosis	Yes	Yes	Yes	Yes	Yes
Cicatricial	Yes	Yes	Yes	Yes	Yes

CPHL, Cicatricial pattern hair loss; *FAPD,* fibrosing alopecia with a pattern distribution; *FFA,* frontal fibrosing alopecia; *LPP,* lichen planopilaris.

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