Australasian Journal of Dermatology

Australasian Journal of Dermatology (2017) ••, ••–•• doi: 10.1111/ajd.12557

SMALL CASE SERIES

Diffuse scarring alopecia in a female pattern hair loss distribution

Bonnie Fergie,[1] Gurpreet Khaira,[2] Vicki Howard[3] and Sally de Zwaan[1]

[1]*Dermatology Department, Royal Newcastle Centre,* [2]*Hunter Histo, and* [3]*Douglass Hanly Moir Pathology, Sydney, Newcastle, New South Wales, Australia*

ABSTRACT

We describe three cases of hair loss in a female pattern hair loss (FPHL) distribution with histologic features of lichen planopilaris (LPP). All patients had a history of diffuse, gradual hair loss in a Christmas tree pattern that clinically presented as FPHL on gross and dermoscopic examination. Notably, there were no characteristic clinical signs of LPP and no histologic features of FPHL. These cases are most consistent with cicatricial pattern hair loss (CPHL). This relatively new entity is similar to fibrosing alopecia in a pattern distribution (FAPD) in that they are both scarring alopecias confined to a FPHL distribution, but CPHL lacks the clinical signs of perifollicular erythema and perifollicular keratosis seen in FAPD. These three cases may present an early, subtle form of CPHL and will be of interest to clinicians and histopathologists alike.

Key words: cicatricial pattern hair loss, female pattern hair loss, fibrosing alopecia in a pattern distribution, lichen planopilaris.

INTRODUCTION

Diffuse scarring alopecia in a female pattern hair loss (FPHL) distribution with histological features of lichen planopilaris (LPP) was first described in 19 patients in 2000 and termed fibrosing alopecia in a pattern distribution (FAPD).[1] It is characterised by clinical features of LPP coupled with histological features of both LPP and FPHL. FAPD is currently classified along side LPP. In 2005 scarring alopecia in a FPHL distribution with histological features similar to FAPD, but lacking the clinical signs of follicular erythema and hyperkeratosis seen in FAPD, was described as cicatricial pattern hair loss (CPHL).[2] In addition, focal atrichia was noted as relatively specific clinical sign of CPHL.

CASE ONE

A 65-year-old Caucasian woman presented with 5 years of progressive, diffuse hair loss, maximally at the frontal scalp. She had minimal intermittent scalp pruritus. Her comorbidities were hypertension and high cholesterol. Her medications included perindopril, atorvastatin, amlodipine and indapamide. She had no family history of FPHL. On gross and dermoscopic examination there was sparse follicle density at the frontal and parietal scalp. No perifollicular erythema or hair casts were seen. No other mucocutaneous sites were involved. A 4-mm punch biopsy from the scalp vertex showed superficial interface perifollicular inflammation of lichenoid type with perifollicular fibrosis. A 4-mm punch biopsy from the scalp occiput showed mild superficial interface perifollicular inflammation, of lichenoid type with infundibular hypergranulosis and mild concentric follicular fibroplasia (Fig. 1). The number of hair follicular units was normal as well as normal total hair numbers, preserved terminal to vellus ratio and a normal telogen count. These changes were suggestive of mild LPP. The patient was treated with topical minoxidil 2%, topical betamethasone dipropionate lotion 0.5 mg/mL once daily and cyproterone acetate 50 mg/day, with some reduction in hair loss.

CASE TWO

A 34-year-old Caucasian woman presented with 4 years of progressive diffuse hair loss maximally at the frontal and

Abbreviations:

CPHL	cicatricial pattern hair loss
FAPD	fibrosing alopecia in a pattern distribution
FPHL	female pattern hair loss
LPP	lichen planopilaris

Correspondence: Dr Bonnie Fergie, Dermatology Department, Royal Newcastle Centre, Lookout Road, New Lambton Heights, NSW 2305, Australia. Email: bonniefergie@gmail.com

Bonnie Fergie, MBBS. Gurpreet Khaira, FRCPA. Vicki Howard, FRCPA. Sally de Zwaan, FACD.

Conflict of interest: none.

Submitted 19 April 2016; accepted 23 July 2016.

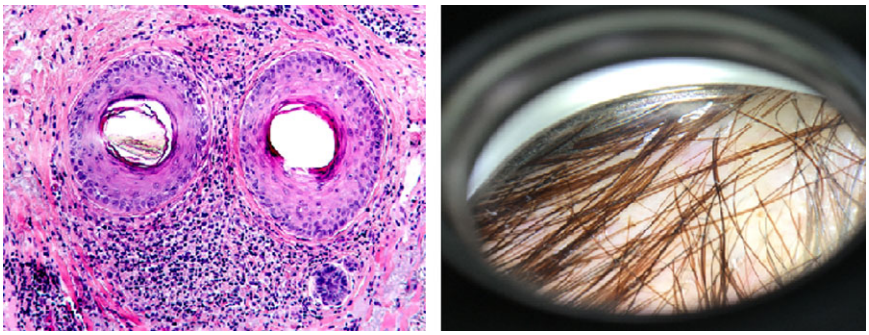

Figure 1 Case 1: histological and dermoscopic appearance. Haematoxylin and eosin.

parietal scalp. She experienced intermittent mild generalized scalp pruritus. She had no comorbidities and took no regular medication. She had a maternal aunt with FPHL. On gross and dermoscopic examination there was diffuse frontal and parietal scalp hair thinning. There was no perifollicular erythema, hair casts or follicular plugging and no loss of follicular ostia. There were no other mucocutaneous sites involved. A 4-mm punch biopsy from the scalp vertex showed focal young circumferential perifollicular fibrosis and mild perifollicular lymphocytic infiltrates and the presence of compound follicles (Fig. 2). The number of hair follicular units was normal, with normal total hair numbers, preserved terminal to vellus ratio and a normal telogen count. A 4-mm punch biopsy of the scalp occiput showed no specific inflammatory pattern, a normal number of follicles, a minor reduction in the terminal to vellus ratio and a normal telogen count. These histopathological features raised the possibility of subtle LPP. The patient was treated with topical minoxidil 2%, and betamethasone dipropionate lotion 0.5 mg/mL once daily. She declined to take spironolactone or other antiandrogens.

CASE THREE

A 36-year-old Caucasian woman presented with 12 years of progressive diffuse hair loss, ongoing pruritus and mild seborrhoeic dermatitis. Her comorbidities were depression, previous cervical intraepithelial neoplasia and asthma. Her medications were ketoconazole 1% shampoo prn and albuterol inhaler prn. She had no family history of FPHL. On gross and dermoscopic examination she had marked diffuse hair loss at the frontal scalp, vertex and temples. There was diffuse mild scalp erythema at the frontal region and a sparse follicle density but no perifollicular erythema or inflammation. No other mucocutaneous sites were involved. A 4-mm punch biopsy from the right frontal scalp showed mild superficial perifollicular interface inflammation of lichenoid type and V-shaped hypergranulosis in the infundibular region (Fig. 3). The number of hair follicular units was normal, as well as normal total hair numbers, a preserved terminal to vellus ratio and a normal telogen count. These histological features are suggestive of LPP. The patient was treated with topical minoxidil 2% and betamethasone dipropionate lotion 0.5 mg/mL twice daily. She declined to take spironolactone or other antiandrogens.

DISCUSSION

The relationship between cicatricial alopecia and FPHL is not fully understood. There appears to be an overlap in the clinical presentation and histology of FPHL, CPHL and FAPD. CPHL, as described by Olsen,[2] is characterised by the histological presence of cicatricial alopecia in a FPHL distribution lacking perifollicular erythema or follicular hyperkeratosis. It characteristically affects women over 40 years of age and has clinically obvious focal atrichia.

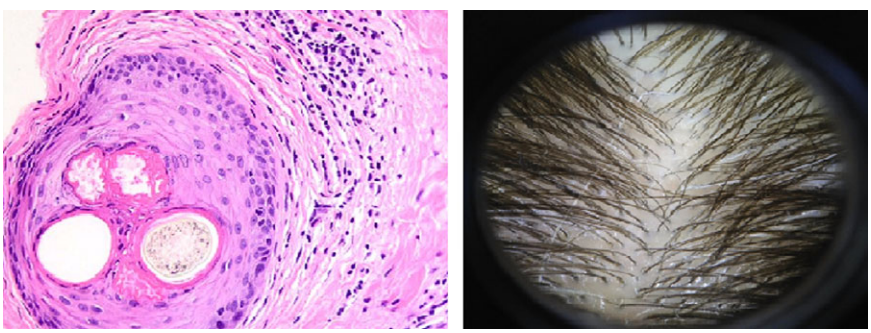

Figure 2 Case 2: histological and dermoscopic appearance. Haematoxylin and eosin.

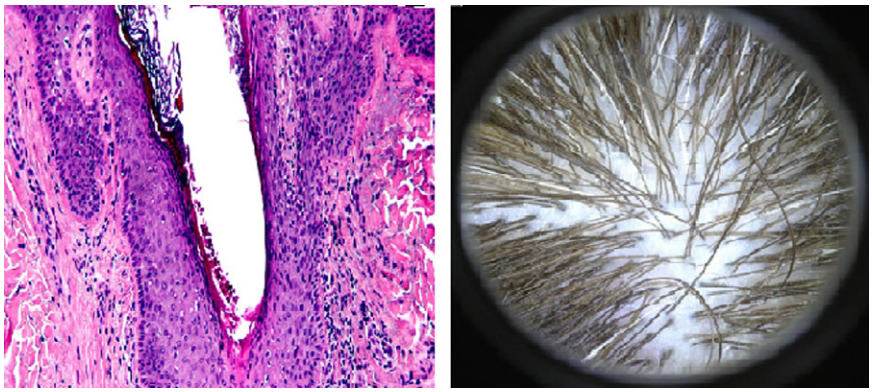

Figure 3 Case 3: histological and dermoscopic appearance. Haematoxylin and eosin.

This relatively specific sign is described as pencil–eraser-sized areas of patchy scarring. Histology shows a perifollicular lymphohistiocytic infiltrate surrounding the isthmus, sebaceous gland loss, and concentric lamellar fibrosis.

FAPD, as reported in 19 patients by Zinkernagel *et al.*[1] usually presents as central scalp scarring hair loss in women or men with underlying established FPHL or male pattern hair loss. Perifollicular erythema, follicular hyperkeratosis and loss of follicular orifices are evident on the central scalp. On histology there is a lymphohistiocytic infiltrate around the isthmus and infundibular region of the hair follicle, concentric perifollicular lamellar fibrosis, sebaceous gland loss and lymphocytic interface dermatitis. There is a reduction in the number of terminal hair follicles and miniaturised follicles may be present.

Frontal fibrosing alopecia, as described by Kossard,[3] is a scarring alopecia that occurs most often in postmenopausal women. The histology, presence of perifollicular erythema and hyperkeratosis are similar to FAPD but the distribution involves the fronto-temporo-parietal area. Mild lymphocytic inflammation has been also reported in approximately one-third of patients with androgenic alopecia alone and one-third of normal controls. A moderate perifollicular lymphohistiocytic infiltrate has been reported in 36% of cases of androgenetic alopecia.[4] Anti-androgens have been shown to ameliorate FAPD in some cases, which suggests a potential role of androgens in the development of inflammation and scarring.[5]

In the three reported cases, the clinical presentation of diffuse hair loss, lack of perifollicular erythema and perifollicular fibrosis is most consistent with CPHL. There was, however, an absence of obvious focal atrichia. This may have been because these areas were small and the women had pale skin. Two of the patients were 24 and 30 years old when their hair loss started, which is much younger than the average age for CPHL. The early onset may account for the subtlety of the focal atrichia. In cases such as these histology aids the diagnosis of a cicatricial alopecia. The three patients had been monitored over many years, which negates the possibility of previous active signs of inflammation being burnt out prior to examination.

All patients showed histological features suggestive of CPHL and FAPD. These included superficial perifollicular interface inflammation of lichenoid type and perifollicular fibrosis (case 1); compound follicles, mild perifollicular lymphocytic infiltrates and subtle perifollicular circumferential fibrosis (case 2); and perifollicular interface inflammation of lichenoid type and V-shaped hypergranulosis in the infundibular region (case 3). Histological features of FPHL were absent in our patients. If present, LPP may destroy miniaturised follicles. In these three patients the follicle count and ratios suggest that FPHL was not present in the areas biopsied. Although there were histological features of both CPHL and FAPD the clinical presentation is consistent with CPHL. A limitation of this study is that single biopsies were taken instead of multiple adjacent biopsies, as suggested by Sinclair *et al.*[6]

All cases were investigated for full blood count, urea, electrolytes, liver function tests, antinuclear antibody, thyroid studies, iron studies, zinc, glucose, calcium, phosphate, vitamin D and sex hormone levels. Results were normal except a low vitamin D level was found in case 3, 45 nmol/L (50–140) nmol/L. She was subsequently commenced on vitamin D replacement therapy.

CONCLUSION

These three cases of progressive, diffuse hair loss that presented clinically as FPHL are most consistent with CPHL. The patients' earlier age at onset and the lack of obvious focal atrichia vary from the characteristic presentation of CPHL. As focal atrichia may be subtle and difficult to discern in pale skin a biopsy may be required to confirm the diagnosis of a cicatricial alopecia. There remains an overlap in the spectrums of FPHL, CPHL and FAPD. The specific interplay between androgens, lymphocytic inflammation and the relation to scarring seen in CPHL and FAPD has not yet been characterised. Further research is needed into the pathogenesis of cicatricial alopecia and FPHL to direct future treatment.

REFERENCES

1. Zinkernagel MS, True RM. Fibrosing alopecia in a pattern distribution: patterned lichen planopilaris or androgenetic alopecia

with a lichenoid tissue reaction pattern? *Arch. Dermatol.* 2000; **136**: 205–11.
2. Olsen EA. Female pattern hair loss and its relationship to permanent/cicatricial alopecia: a new perspective. *J. Investig. Dermatol. Symp. Proc.* 2005; **10**: 217–21.
3. Kossard S. Postmenopausal frontal fibrosing alopecia. *Arch. Dermatol.* 1994; **130**: 770–4.
4. Whiting DA. Scalp biopsy as a diagnostic and prognostic tool in androgenetic alopecia. *Dermatol. Ther.* 1998; **3**: 24–33.
5. Olsen EA, Bergfeld WF, Cotsarelis G *et al.* Workshop on cicatricial alopecia. Summary of North American Hair Research Society (NAHRS)-sponsored workshop on cicatricial alopecia, Duke University Medical Centre, February 10 and 11, 2001. *J. Am. Acad. Dermatol.* 2003; **48**: 103–10.
6. Sinclair R, Jolley D, Mallari R *et al.* The reliability of horizontally sectioned scalp biopsies in the diagnosis of chronic diffuse telogen hair loss in women. *J. Am. Acad. Dermatol.* 2004; **51**: 189–99.