Dermatology Practical & Conceptual

Folliculitis Decalvans with Frontal Fibrosing Alopecia in a Dark Phototype: Presentation of Folliculitis Decalvans and Lichen Planopilaris Phenotypic Spectrum

Basma Karrakchou[1], Amani Fliti[1], Amal El Fiboumi[2], Fouad Kettani[3], Karima Senouci[1], Mariame Meziane[1]

1 Dermatology and Venereology Department, Ibn Sina Hospital, Mohammed V University of Rabat, Morocco
2 Department of medicine, Moulay Youssef Hospital, Mohammed V University of Rabat, Morocco
3 Histopathology Center of the United Nations, Rabat, Morocco

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Corresponding Author: Basma Karrakchou: M.D. Dermatology and Venereology Department, Ibn Sina Hospital, Mohammed V University, Rabat, Morocco. orcid number: https://orcid.org/0000-0001-5509-5412 E-mail adress: karrakchou.basma@gmail.com

Introduction

Lichen planopilaris (LPP) and folliculitis decalvans (FD) are two primary scarring alopecias recently associated in a phenotypic spectrum in which they occur simultaneously or in a bi-phasic presentation, either in the same scalp location or in different scalp areas [1]. We report a sequential onset of vertex FD and frontal fibrosing alopecia (FFA), a variant of LPP, as an exceptional presentation of this spectrum in phototype V.

Case Presentation

A 42-year-old premenopausal woman presented with a 10-year history of vertex pustules and crusts leading to scarring patches of alopecia. Nine years later, she presented pruritus in the frontal hairline. The physical examination found a phototype V patient with two vertex keloid patches of alopecia measuring 6 cm and 4 cm in diameter respectively, containing tufts, follicular pustules, hemorrhagic crusts, milky-red areas, and dilated vessels on trichoscopy (Figure 1, A and C).

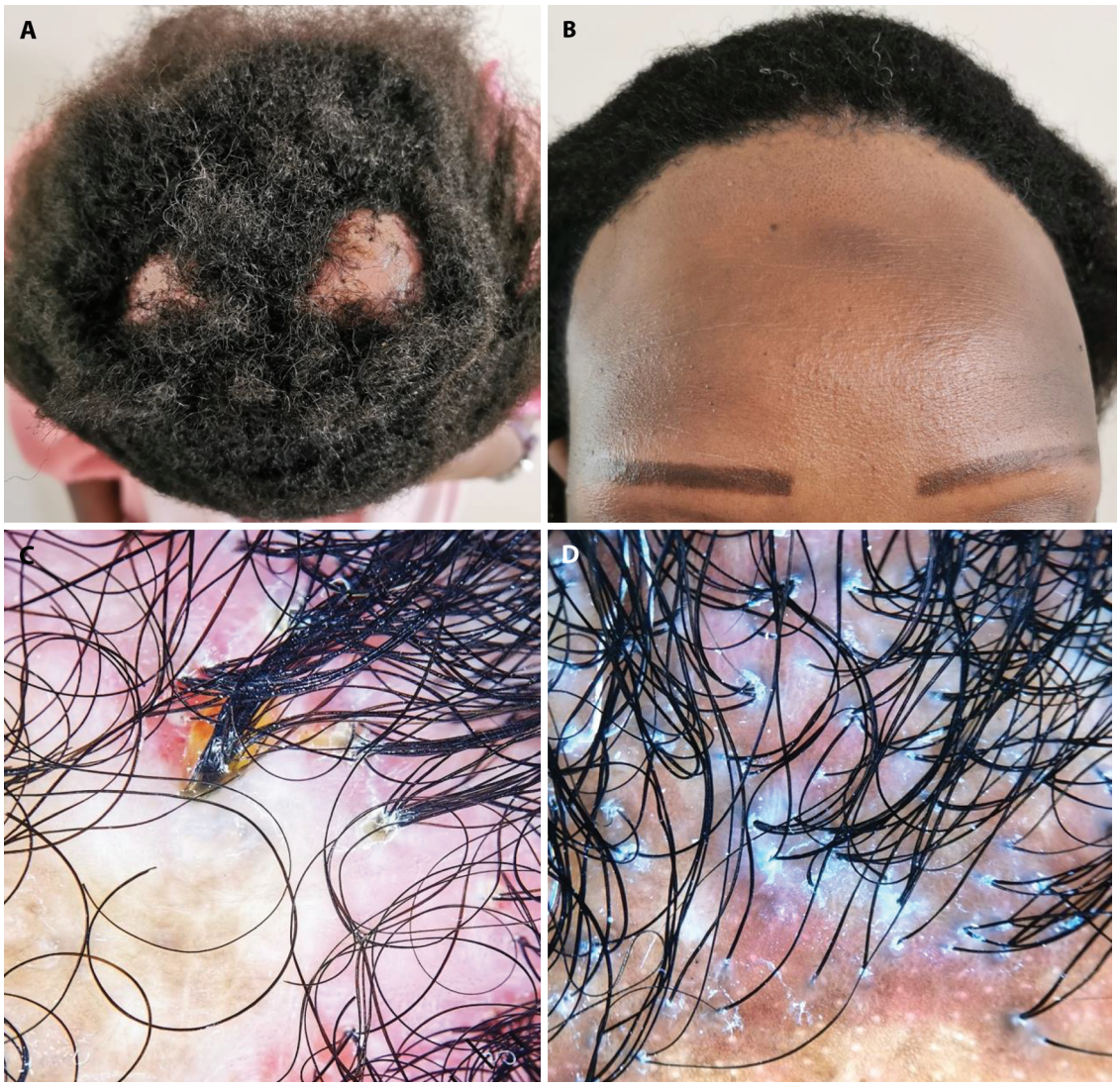

Figure 1. (A) Clinical aspect of folliculitis decalvans: 2 vertex patches of alopecia with tufted hairs, pustules and crusts on an underlying keloid scar. (B) Front view showing an associated frontal fibrosing alopecia, and lichen planus pigmentosus. (C) Vertex trichoscopy: pustules and hemorrhagic crusts surrounding tufts of more than 5 hairs. There were cicatricial milky-red areas with dilated vessels and no follicular openings. (D) Frontal hairline trichoscopy: perifollicular erythema, peripilar hyperkeratosis, and tubular hair casts. Absence of vellus hairs was noticed. Yellow dots were seen on dark phototype and corresponded to sebaceous glands.

There was an associated 2.5 cm linear frontal hairline recession with trichoscopy showing peripilar hyperkeratosis and erythema, tubular hair casts, and yellow dots (Figure 1, B and D). No vellus hairs were seen. Additional eyebrow loss covered by micropigmentation, facial papules, and facial hyperpigmentation were noticed.

A diagnosis of FD associated with FFA and lichen planus pigmentosus was confirmed by histopathology (Figure 2, A-D). No bacterial sample was taken from vertex patches of alopecia due to ongoing oral doxycycline (50 mg twice a day) at that time, with no improvement after 6 months. The decision was made to cease treatment and to prescribe oral low-dose isotretinoin (0.2 mg/kg per day for at least 6 months), intralesional injections of corticosteroids every 6 weeks for FFA and FD, and topical fusidic acid twice a week for FD. Within one year of treatment, FFA stabilization was achieved with no more peripilar erythema and hyperkeratosis, no hair loss progression, and no hair regrowth. A subsidence of keloid FD scars was obtained with persistence of some pustules, which needed the adjunction of oral azithromycin (500 mg per day, 3 days per week for 3 weeks) for remission. Currently, the patient is still under treatment, with a taper-off of isotretinoin dose after one year (0.1 mg/kg per day), in the view of its discontinuation.

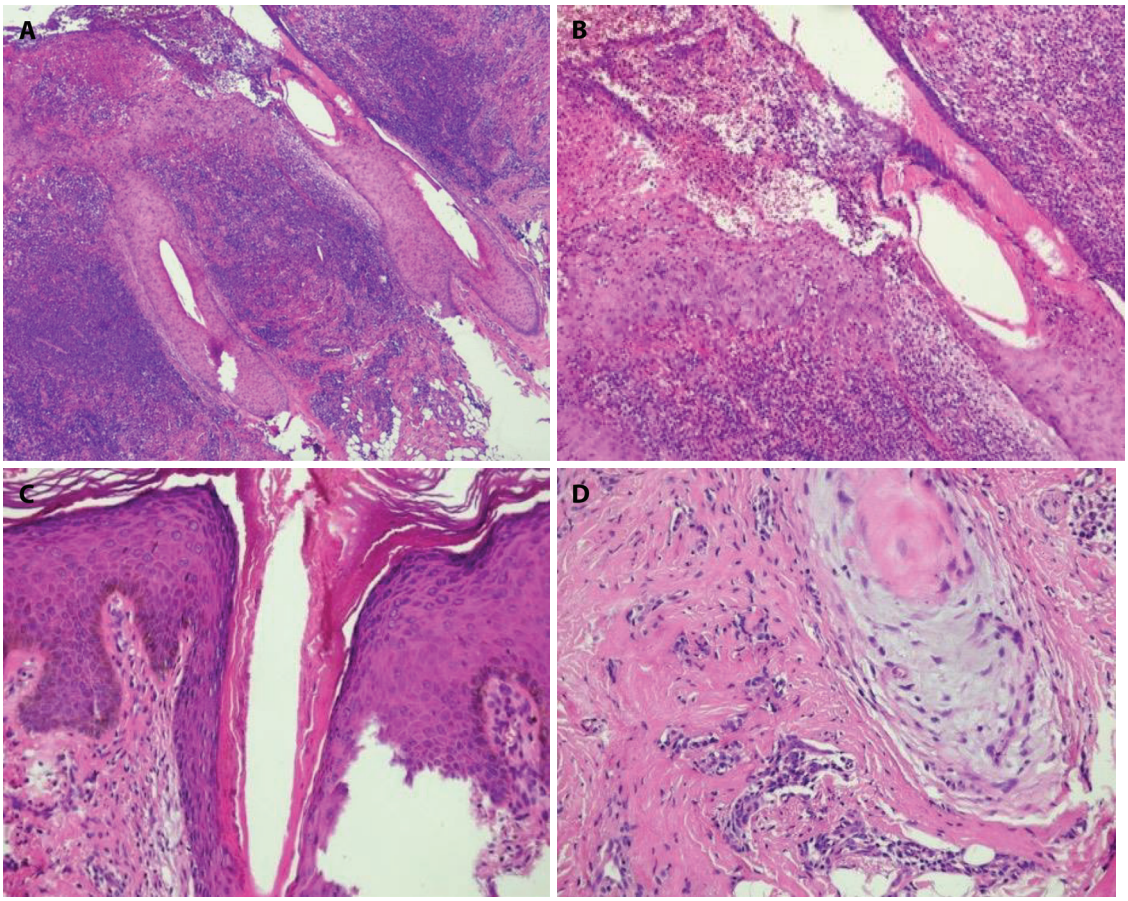

Figure 2. (A, B) Histopathology view (Hematoxylin-Eosin stain, (A) low magnification, (B) high magnification) of vertex scalp biopsy showing a dense peripilar neutrophilic infiltrate, infundibular pustule, and plasma cell exocytosis. They destruct focally hair shafts. Fibrosis is noted in deep dermis. (C, D) Histopathology aspect (Hematoxylin-Eosin stain, (C) low magnification, (D) high magnification) of frontal hairline scalp biopsy: isthmic and infundibular perifollicular hyperkeratosis associated with a moderate dermic lymphocytic infiltrate and dense fibrosis.

Conclusions

FD and LPP are primary scarring alopecias recently combined in the "FD and LPP phenotypic spectrum" (FDLPPPS) [1-5]. To date, mainly vertex scalp LPP cases have been reported in association with FD [1-5]. To the best of our knowledge, only 2 observations described FFA within the phenotypic spectrum, and the two patients were phototype II (Table 1) [1,5].

FFA is prominent in postmenauposal light skinned women. When there is additional lichen planus pigmentosus, as in our case, FFA mostly affects dark phototype females [6]. FD generally occurs in African American young men. And scalp keloid scars are described in FD only when associated with acne keloidalis nuchae, which occurs in 21% [7].

Trichoscopy has a prominent place by showing either separate features of FD and FFA, or a progressive switch from one to another using dynamic trichoscopy [1]. The aspect is then more or less a mixture of FFA activity signs, such as peripilar erythema and scaling, with FD activity patterns, as crusts and pustules. In late stages of scarring alopecia, there are milky-red areas and tufts in FD, and no follicular openings in FFA.

Histopathology is the hallmark of FDLPPPS' final diagnosis, especially when FD and LPP occur in the same area. The condition is characterized by more follicular packs and plasma cells exocytosis over neutrophils [2], and less bacterial infiltrate [8].

Both conditions progress into scarring alopecia, and early diagnosis is then compulsory to an adequate management based on anti-inflammatory agents (corticosteroids and isotretinoin) associated with oral antibiotics [1-5]. However, there is no consensus in FDLPPPS treatment, and the therapy duration remains to be established. A long-term follow up is also needed to manage the frequent disease flares occurring after treatment completion.

Table 1. Literature reported cases of Frontal Fibrosing Alopecia associated with Folliculitis Decalvans

Patient	Age (y), Gender	Fitzpatrick phototype	Onset	Clinical aspect	Trichoscopic aspect	Histology	Bacterial culture (scalp swab)	Treatment	Outcome	Reference #
1	42, F	II	Concomitant	Frontal hairline recession and vertex area of scarring alopecia.	Frontal trichoscopy: peripilar hyperkeratosis and erythema consistent with FFA. Vertex trichoscopy: tufts, crusts, follicular plugging, and pustules compatible with FD.	FFA: peri-isthmic fibrosis and lymphocytic infiltrate FD: Polytrichia, perifollicular neutrophilic infiltrate and microabces.	MSSA	Lymecycline 408mg bd followed by oral and topical fusidic acid for 3 weeks, and oral zinc sulfate 125mg bd	Intermittent recurrences controled with intermittent topical and oral fusidic acid and topical corticosteroids.	[1] Case 9
2	65, F	II	Sequential: FFA then FD	Pustular frontal hairline alopecia.	Frontal trichoscopy: peripilar hyperkeratosis erythema and pustules.	Frontal hairline scalp: Perifollicular lymphocytic dermic infiltrate and presence of plasma cells. Intraepidermal pustule with neutrophils were present.	Sterile	Topical corticosteroids for 1 month.	No disease flare occured within 3 months of follow up.	[5]
Our patient	42,F	V	Sequential FD then FFA	Linear frontal hairline recession and two vertex keloid patches of alopecia.	Frontal trichoscopy: peripilar hyperkeratosis, sliding sheaths, perifollicular erythema, and absence of vellus hairs. Vertex trichoscopy: tufts, pustules, and crusts, associated with cicatricial milky-red areas and dilated vessels.	FFA: infundibular hyperkeratosis and dermic lymphocytic infiltrate with fibrosis.FD: multicoupound hair structures and plasma cells exocytose with neutrophilic infiltrate and dermic fibrosis.	Not performed	Doxycyclines 50mg bd followed by long course oral isotretinoin 20mg/d and intralesional corticosteroids, and topical fusidic acid. Additional oral azithromycin 500mg/d for 3days per week during 3 weeks were prescribed.	Stabilization of FFA, subsidence of keloid patches of alopecia and drying of FD lesions within 1 year.	

FD = folliculitis decalvans; FFA = frontal fibrosing alopecia.

In conclusion, this case highlights the possible emergence of new variants within the FDLPPPS, as the occurrence of FD-FFA in a dark phototype. The keloid evolution of FD lesions is possible without an associated acne keloidalis nuchae in highly pigmented skins. FD and FFA have both a scarring evolution, and trichoscopy allows an early diagnosis orientation for an urgent management. Further case reports are however needed to fully characterize the FDLPPPS spectrum.

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