Case study

Folliculitis Decalvans and Lichen Planopilaris Phenotypic Spectrum Three cases report

ABSTRACT:

Introduction: folliculitis decalvans (FD) and lichen planopilaris (LPP) are two primary cicatricial alopecias (neutrophilic and "lymphocytic" respectively) recently associated in a phenotypic spectrum. The "folliculitis decalvans and lichen planopilaris" phenotypic spectrum FDLPPPS is a combination of the clinical and histological features of FD and LPP. The objective of our study is to analyze three cases of a specific disease in order to better understand its clinical characteristics, evolution, and treatment.
Materials & Methods: We reported 3 cases of FDLPPPS with clinical manifestations, trichoscopy, histopathological features, treatments, and follow up findings as a new presentation of this spectrum.
Results: We reported 3cases, two females, and one male, with an average age of 33 years (ranging from 20 to 42 years). The average duration of disease evolution before diagnosis was 6.3 years (ranging from 1 to 10 years). Clinically, they presented with one or multiple alopecic patches on the vertex with pustules and crusts, and a receding frontal hairline in one patient. Dermoscopy revealed signs consistent with lichen planopilaris and folliculitis decalvans. A diagnosis of phenotypic spectrum was established through histology. Patients received treatments including corticosteroids, antibiotics, hydroxychloroquine, or isotretinoin, with good improvement noted in one patient, moderate improvement in another, and stabilization in the third.
Conclusion: This phenotypic spectrum is a newly recognized entity, combining clinical and histological signs of FD and LPP. Early treatment is crucial to halt disease progression. The emergence of new variants within FDLPPPS, such as FD-frotnale fibrosing alopecia, underlines the need for further case series to better characterize this spectrum.

KEYWORDS: Phenotypic spectrum, lichen planopilaris, folliculitis decalvans, trichoscopy, fortnal fibrosing alopecia, alopecia

INTRODUCTION:

folliculitis decalvans (FD) and lichen planopilaris (LPP) are two primary cicatricial alopecias (neutrophilic and "lymphocytic" respectively) (1) recently associated in a phenotypic spectrum in which they occur simultaneously or in a bi-phasic presentation, in the same or in different areas of the scalp (2). The "folliculitis decalvans and lichen planopilaris" phenotypic spectrum FDLPPPS is a combination of the clinical and histological features of FD and LPP.
We report 3 cases of FDLPPPS as a new presentation of this spectrum.

MATERIALS & METHODS:

We reported 3 cases of FDLPPPS with clinical manifestations, trichoscopy, histopathological features, treatments, and follow up findings

RESULTS:

Case 1 [8]: A 42-year-old woman presented with a 10-year history of vertex scarring patches of alopecia. Since one year , 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, with negative pull test. trichoscopy found tufts, follicular pustules, hemorrhagic crusts, milky-red areas, and dilated vessels on trichoscopy (Figure 1, A and C). 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). Additional eyebrow loss covered by micropigmentation, facial papules, and facial hyperpigmentation were noticed.

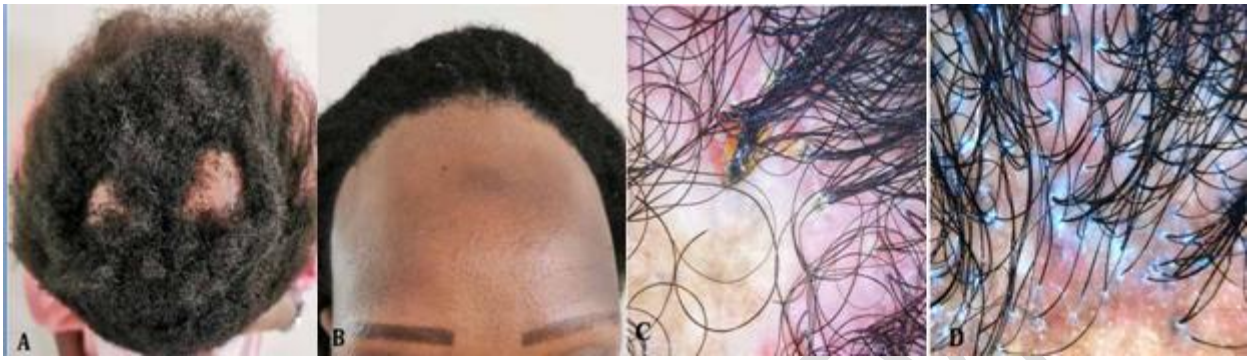

Fig 1: 2 vertex cheloid patches of alopecia(A) with trichoscopy(C) linear frontal hairline recession(B) with trichoscopy(D)

Histopathology view of vertex scalp biopsy showing a dense peripilar neutrophilic infiltrate, infundibular pustule, and plasma cell exocytosis and dermal fibrosis. (Fig 2 A, B)
Histopathology aspect of frontal hairline scalp biopsy showing perifollicular hyperkeratosis associated with a moderate dermic lymphocytic infiltrate and fibrosis.(Fig 2 C,D)
A diagnosis of FD associated with frontal fibrosing alopecia (FFA) and lichen planus pigmentosus was confirmed by histopathology.

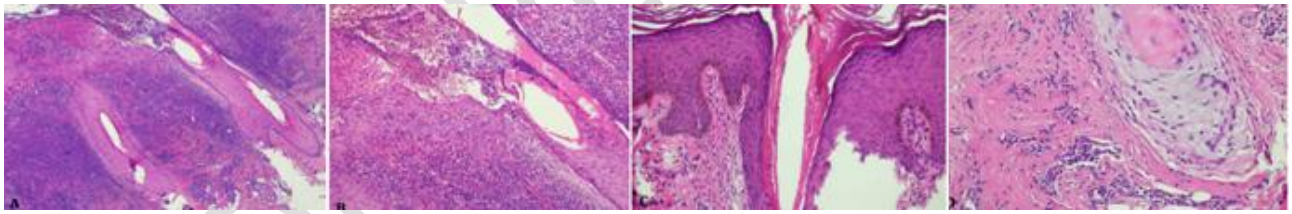

Figure 2. Histopathology view (A,C) low magnification, (B,D) high magnification) of vertex scalp biopsy (A,B) showing features consistent with folliculitis decalvans . and of frontal hairline scalp biopsy (C,D): showing features consistent with frontal fibrosing alopecia

A metabolic analysis as well as investigation of associated pathologies with lichen were mad and were without abonormality
No bacterial sample was taken from vertex patches of alopecia
the patient was treated by oral low-dose isotretinoin (0.2 mg/kg per day), intralesional injections of corticosteroids every 6 weeks for FFA and FD, topical fusidic acid twice a week for FD, and tacrolimus 0.1% for eyebrow.
Within one year of treatment, FFA stabilization was achieved , then some pustules have appeared, which needed the adjunction of oral azithromycin (500 mg per day, 3 days per week for 3 weeks) for remission [8].

Case2: A 37-year-old female patient presented with intermittent pustules and scales on her scalp for 3 years, preceding an extensive hair loss. Following systemic antibiotic therapy, she made a ;lgradual improvement with new hair grown.

However, 1 year ago Lesions of cicatricial alopecia appeared on the scalp with follicular papules, pustules, hemorrhagic crusts, perifollicular erythema, and scale build-up.

She was diagnosed with folliculitis decalvans and received different courses of topical/oral antibiotics along with corticosteroids
Nevertheless, her symptoms continued to progress slowly
Dermatological examination showed a wide patch of cicatricial alopecia on the scalp, measuring 15 ×20 cm , pull test hair was negative Trichoscopy revealed loss of partial follicular opening, follicles surrounded by milky red areas, dilated blood vessels, pustules, and follicular tufts. Fig 3

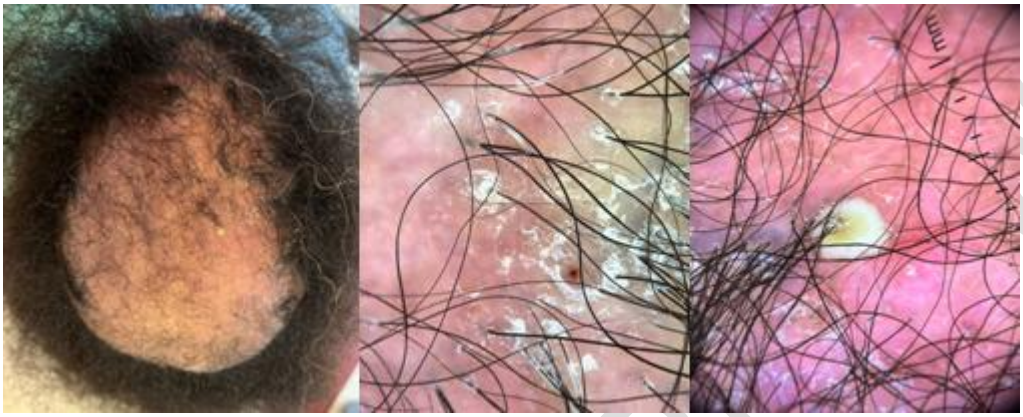

Fig 3: patch of cicatricial alopecia on the scalp with trichoscopy images

Otherwise, grouped, skin-colored, keratotic follicular papules were found on her neck and chest without any other lesions on the face, extremities, mouth, or nails,

Scalp biopsies from the center and the edge of the alopecia patch, with horizontal and vertical sections showed differnet findings :
Histopathological examination of the edge showed hyperkeratosis, mild epidermal hyperplasia, and a neutrophilic infiltration along the follicle fig 4(A,B)
Histopathological examination of the centre showed epithelial basal layer destruction in the hair follicle, with dense lymphocytic infiltration and partial destruction of hair follicles surrounding the hair follicle fig 4(C,D).

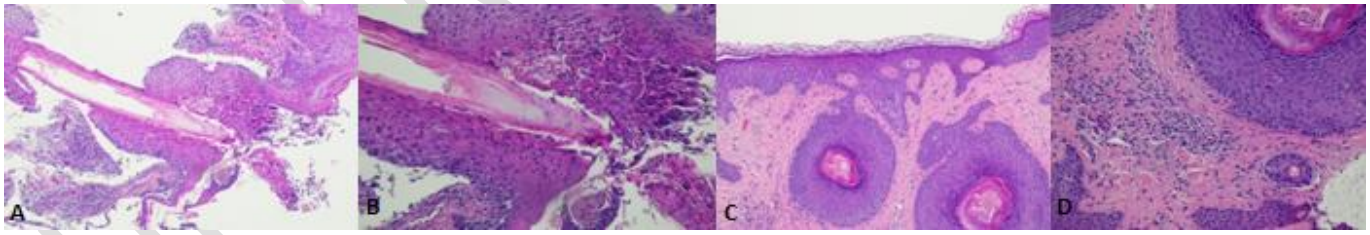

Figure 4. Histopathology view (A,C) low magnification, (B,D) high magnification) of scalp biopsy at the edge (A,B) showing features consistent with folliculitis decalvans . and of scalp biopsy at the center (C,D): showing features consistent with lichen planopilaris

These clinical, trichoscopic and histopathological findings were consistent with the diagnosis of FDLPPPS.

A metabolic analysis as well as investigation of associated pathologies with lichen were mad and were without abonormality
the patient was treated by oral doxyxycline 100 mg / day, hydroxychloroquine 400 mg / day topical fucidic acid and Propionate of clobetasol 0.05% : twice / week

Case3: A 20-year-old male treated since 8 years for pilar lichen with corticosteroids, he presented with scarring patches of alopecia, pustules and crusts on her scalp, and hair pull test was negative. Trichoscopy showed hyperkeratosis and erythema, tubular hair casts,milky red areas and tufts fig5.

Fig 5: scarring patches of alopecia on the scalp with trichoscopy images

A diagnosis of FD LPPPS was confirmed by histopathology

A metabolic analysis as well as investigation of associated pathologies with lichen were mad and were without abonormality the patient was treated by Diprostene IM injection 1injection /month Doxycyline 100mg/Day and topical Fusidic acid, with hair growth after 8 months

Table 1 : List of different cases

	Case 1	Case 2	Case 3
Gender/Age	42-year- old woman	37 year-old woman	20 year-old man
Physical examination	two vertex keloid patches of alopecia linear frontal hairline recession and eyebrow depilation	Alopecic plaque of vertex with pustules, hemorrhagic crusts and scales.	Several alopecic plaques on the vertex with crusts and pustules
Trichoscopy	Vertex : tufts, follicular pustules, hemorrhagic crusts, milky-red areas, and dilated vessels - Linear frontal hairline : peripilar hyperkeratosis and erythema, tubular hair casts, and yellow dots	peripilar hyperkeratosis and erythema, tubular hair casts tufts, follicular pustules, hemorrhagic crusts, milky-red areas, and dilated vessels	peripilar hyperkeratosis tubular hair casts milky-red areas, Des croutes , and tufts
Histology	FD + Frontal Fibrosing Alopecia (FFA)	FD+LPP	FD+LPP
Metabolic analysis	No anomalies	No anomalies	No anomalies

Treatment	Isotretinoine Intralesional corticosteroid injection Tacrolimus 0.1% Fusidic acid the day before showering	Hydroxychloroquine Doxycycline Propionate of clobetasol 0.05% Fusidic acid the day before showering	Diprostene IM injection 1injection /month Doxycyline 100mg/DAY Fusidic acid the day before showering
Evolution	Stable for 1 year	Evolutionary	Growth in 8 months

DISCUSSION

FD and LPP were classified into two distinct categories: "neutrophilic" primary scarring alopecia and "lymphocytic" respectively (1),

Morais et al (3) reported on 13 cases of lichen planopilaris with pustules, crusts, follicular tufts and histopathological findings of lymphocytic infiltration , They suggested the difficulty in differentiating lichen planopilaris and folliculitis decalvans, and the need for further study to elucidate whether lichen planopilaris with pustules constitute a new subtype of LPP or even a different disease.

Then Yip et al (2) reported other 13 cases, where the lesions developed biphasic clinical and histologic features of folliculitis decalvans and lichen planopilaris , Folliculitis decalvans presentations generally preceded those of lichen planopilaris . They were the first to introduce the term folliculitis decalvans lichen planopilaris phenotypic spectrum.

then 7 additional cases were reported by Egger et al.(4)

Ramos et al has previously reported on two cases of FDLPPPS diagnosed in pediatric patients.(5)

Between 2018 and 2023, only 39 cases of this spectrum have been reported in the literature, including only 3 cases of FFA+FD (2.7.8).

Yip et al hypothesize about the likely pathogenic mechanisms that underlie this spectrum of phenotypic appearances; in particular, abnormal hair follicle inflammatory responses and collapse of hair follicle immune privilege in response to microbiome dysbiosis (2).

FDLPPS is a combination of the clinical and histological features of FD and LPP (9-12).

Tufting, pustules and yellow crusts are clinically characteristic of FD, whereas perifollicular cast and erythema suggest LPP (3.4.5).

Regarding trichoscopy, both LPP and FD can show pronounced inflammation, hyperkeratosis, and lack of follicular ostia. Kang et al. [6] suggested that LPP presents peripilar white/silver scales, peripilar erythema, casts, concentric blood vessels, and violaceous coloration of interfollicular epidermis, while FD is characterized by peripilar white yellowish scales, peripilar hyperplasia, white and milky-red areas, and numerous hair tufts

Regarding histopathology, the identification of multicompound follicular structures, atrophy of the follicular epithelium, and prominent plasma cell component instead of predominance of neutrophils, should be considered for the diagnosis of FDLPPPS (3).

Early diagnosis is essential for appropriate management, based on anti-inflammatory agents combined with antibiotics. However, there is no consensus in FDLPPPS treatment, and the therapy duration remains to be established. Oral doxycycline or association of rifampicin / clinadamycin in combination with oral or intralesional steroids is commonly used as first line treatment.

Hydroxychloroquine, cyclosporine, retinoids and dapsone were all reported as possible treatments (2-5).

Long-term follow up is also needed to manage the frequent disease flares occurring after treatment completion.

CONCLUSION:

FDLPPS is a combination of the clinical and histological features of FD and LPP.

The emergence of new variants within FDLPPPS, such as FD-FFA, underlines the need for further case series to better characterize this spectrum.

REFERENCES:

1- Olsen EA, Bergfeld WF, Cotsarelis G, et al. Summary of North American Hair Research Society (NAHRS)-sponsored Workshop on Cicatricial Alopecia, Duke University Medical Center, February 10 and 11, 2001. J Am Acad Dermatol. 2003;48: 103-10.
2- Folliculitis decalvans and lichen planopilaris phenotypic spectrum: a case series of biphasic clinical presentation and theories on pathogenesis. Yip L, Barrett TH, Harries MJ. *Clin Exp Dermatol.* 2020;45(1):63-72. DOI: 10.1111/ced.13989. PMID: 31017678.
3- Morais KL, Martins CF, Anzai A, Valente NYS , Romiti R. Lichen Planopilaris with Pustules: A Diagnostic Challenge. Skin Appendage Disord 2018;4:61-6.
4- Egger A, Stojadinovic O , Miteva M. Folliculitis Decalvans and Lichen Planopilaris Phenotypic Spectrum-A Series of 7 New Cases With Focus on Histopathology. Am J Dermatopathol 2020;42:173-7.
5- Ramos PM , Miot HA. Exuberant tufted folliculitis. An Bras Dermatol 2019;94:115-6.
6- Kang H, Alzolibani AA, Otberg N, Shapiro J: Lichen planopilaris. Dermatol Ther 2008; 21: 249–256.
7- Lobato-Berezo A, González-Farré M, Pujol RM. Pustular frontal fibrosing alopecia: a new variant within the folliculitis decalvans and lichen planopilaris phenotypic spectrum? *Br J Dermatol.* 2022;186(5):905-907. DOI: 10.1111/bjd.20962. PMID: 34939665.
8- Karrakchou B, Fliti A, El Fiboumi A, Kettani F, Senouci K, Meziane M. Folliculitis Decalvans with Frontal Fibrosing Alopecia in a Dark Phototype: Presentation of Folliculitis Decalvans and Lichen Planopilaris Phenotypic Spectrum. *Dermatol Pract Concept.* 2023;13(4):e2023229. DOI: https://doi.org/10.5826/dpc.1304a229
9- Zhang X, Zhu M, Zhou J, Wu S, Liu J, Qin Q. Folliculitis Decalvans and Lichen Planopilaris Phenotypic Spectrum: A Case Report. Clin Cosmet Investig Dermatol. 2022 May 31;15:993-996. doi: 10.2147/CCID.S365566. PMID: 35677221; PMCID: PMC9167836.
10- Bolduc C, Sperling LC, Shapiro J. Primary cicatricial alopecia: Lymphocytic primary cicatricial alopecias, including chronic cutaneous lupus erythematosus, lichen planopilaris, frontal fibrosing alopecia, and Graham-Little syndrome. J Am Acad Dermatol. 2016;75:1081-99. doi: 10.1016/j.jaad.2014.09.058.
11- Bolduc C, Sperling LC, Shapiro J. Primary cicatricial alopecia: Other lymphocytic primary cicatricial alopecias and neutrophilic and mixed primary cicatricial alopecias. J Am Acad Dermatol. 2016;75:1101-7. doi: 10.1016/j.jaad.2015.01.056.
12- Bernárdez C, Molina-Ruiz AM, Requena L. Histologic features of alopecias: part II: scarring alopecias. Actas Dermosifiliogr. 2015;106:260-70. doi: 10.1016/j.ad.2014.06.016.