

World Journal of Advanced Research and Reviews

eISSN: 2581-9615 CODEN (USA): WJARAI Cross Ref DOI: 10.30574/wjarr Iournal homepage: https://wjarr.com/



(RESEARCH ARTICLE)



Frontal Fibrosing Alopecia: A prospective observational study

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World Journal of Advanced Research and Reviews, 2023, 18(01), 1017-1028

Publication history: Received on 19 February 2023; revised on 19 April 2023; accepted on 21 April 2023

Article DOI: https://doi.org/10.30574/wjarr.2023.18.1.0507

Abstract

Introduction: Frontal fibrosing alopecia (FFA) is a scarring alopecia of unknown etiology that affects mainly postmenopausal women. It is responsible for progressive recession of the frontotemporal hairline and sometimes occipital hairline with inconsistent eyebrow hair loss. We aimed to investigate the demographics, clinical and trichoscopic features of FFA in the Moroccan population.

Patients and methods: We conducted a prospective study over a period of 2 years and 1/2, from January 2020 to July 2022. We enrolled twenty four patients, with clinical and/or histopathological diagnosis of FFA, seen at the dermatology department of Ibn Rochd University Hospital in Casablanca, Morocco. Data regarding demographics, clinical and trichoscopic findings were collected.

Results: Twenty Four patients with FFA met the inclusion criteria. There were all women and half of them were postmenopausal (50%). The average age of disease onset was 46,95 years old. Pruritus was the most reported symptom (80%). An emotional factor was found in 40% of the cases and 45% of the patients reported the use of traditional products to their hairs. All patients presented with frontotemporal hairline recession with parietal or occipital involvement in 70% and 50% of the cases respectively, and 16 patients experienced eyebrow loss. Facial micropapules were found in 9 patients (45%), followed by patchy hyperpigmentation in 6 patients (30%) and follicular hyperpigmentation in 4 patients (20%).

The majority of patients presented mild FFA (grades I and II), with a recession of less than 3 cm of the frontotemporal hairline (85%). The most frequent trichoscopic findings were perifollicular erythema (80%) and follicular hyperkeratosis (70%), followed by decreased or absence of vellus hairs (60%), lonely hairs (45%), perifollicular bluegray pigmentation (35%), perifollicular brownish pigmentation (30%) and loss or absence of follicular openings (30%). Tufted hairs and white patches of scarring alopecia were found in 25% and 20% patients, respectively.

Conclusion: Our study supports the role of cosmetic products or emotional factors in the physiopathology of FFA. Trichoscopy seems a valuable tool in the diagnosis of AFF. Perifollicular erythema, follicular hyperkeratosis and decreased of absence of vellus hairs are easily identified and are very suggestive of the diagnosis.

Keywords: Frontal fibrosing alopecia; Trichoscopy; Lichen planopilaris; Emotional factors

1. Introduction

Frontal fibrosing alopecia (FFA) is a distinctive form of primary lymphocytic cicatricial alopecia that affects mainly postmenopausal women (1,2). It presents clinically with progressive recession of the frontotemporal hairline and sometimes occipital hairline with inconsistent eyebrow hairloss (3). Pruritus, pain and burning can be observed in variable degrees on the affected sites (1). It is considered as a variant of lichen planopilaris (LPP) (4). The objective of

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this study was to investigate epidemiological data, clinical presentation and trichoscopic features of FFA in the Moroccan population through a prospective series of twenty four cases.

2. Material and methods

2.1. Study

This is a single-center prospective descriptive study that included all the patients diagnosed with FFA, seen in the dermatology department of Ibn Rochd University Hospital of Casablanca, between January 2020 and July 2022. Diagnosis of FFA was made histologically or based on the typical clinical presentation (irregular recession of the frontotemporal and preauricular hairline with eyebrow loss) and characteristic dermoscopic findings. The trichoscopic examination was performed by a single examiner, the histological interpretation was done by the same anatomopathologist.

2.2. Data collection

For each patient, a detailed history was recorded including gender, age, age onset, disease duration, comorbidities, gynecological status, regular hair cosmetic product use, family history of hair loss, and symptoms (pruritus, trichodynia or burning).

Data regarding clinical examination were noticed (clinical severity, lonely hairs, facial papules, lichenoid skin, occipital involvement, eyebrow or eyelash involvement, affectation of body hair, mucosal or nail changes, association with another skin disorder.

The clinical severity of FFA was classified based on a clinical scale, measuring the area of cicatricial skin produced by the recession of the frontal and temporal hairline. This classification included 5 grades of severity: I (<1 cm), II (1-2.99 cm), III (3-4.99 cm), IV (5-6.99 cm), and V (>7 cm), which were grouped for statistical purposes as mild FFA (grades I and II) and severe FFA (grades III, IV, and V). In each patient, the largest measure (frontal or temporal) was used to define the grade of severity. We defined the normal frontal implantation hairline using the measurements recently published by Ceballos et al. (5).

Hairlines and eyebrows were examined by handheld dermatoscope (Dermlite DL4; 3Gen). Clinical photographs were taken by a smartphone and trichoscopic pictures were recorded by the digital camera of the smartphone attached to the dermatoscope.

3. Results

3.1. Patient demographics

Twenty Four patients with FFA met the inclusion criteria and were included in the study. They were all women. FFA diagnosis was confirmed by histology in 12 patients (40%), while 16 patients (60%) were clinically diagnosed. The mean age was 52,4 years old (range, 36-69 years), and the average age of disease onset was 46,95 years old (range, 35-65 years). The median duration of FFA was 5,45 years. Half of women were postmenopausal (50%), and 2 of them had received hormone replacement therapy (10%).

The most frequent comorbidities were: arterial hypertension in 3 patients (15%), thyroid dysfunction in 2 patients, obesity in 2 patients, dyslipidemia in 2 patients, and depression in 2 patients (10%). One patient suffered from anemia (5%), another from diabetes (5%) and another had a history of breast cancer (5%).

A family history of hair loss disorders was found in 20% of the cases, with 2 cases of FFA (10%) and 2 cases of androgenic alopecia (10%). Among the patients, 30% reported taking contraception by estrogen and progestin during their lifetime, while 45% reported the use of traditional products (henna, lemon, rassoul) and 15% used hair dyes. An emotional factor was found in 40% of the cases (bereavement, divorce, marital dispute).

Pruritus was the most reported symptom (80%), followed by burning (25%) and trichodynia (20%). Three patients were asymptomatic (15%).

Demographic characteristics of patients are summarized in Table 1.

 Table 1
 Demographic characteristics of patients

Patients	Age (years old)	Age at disease onset (years old)	Duration (years)	Hormonal status	Comorbidities	Family history of hair loss	Triggering factor	Symptoms
1	57	44	13	postmenopausal	anemia	-	-	pruritus
2	60	50	10	postmenopausal	arterial hypertension	AAG	Traditional products, hair dyes, bereavement	Pruritus, trichodynia
3	45	40	5	Premenopausal, contraception	-	FFA	Hair dyes, traditional products	Pruritus, trichodynia
4	65	50	15	Postmenopausal, contraception	dyslipidemia, obesity	-	Hair dyes, traditional products	burning
5	45	42	3	premenopausal	-	-	-	pruritus
6	48	44	4	Postmenopausal, HST	thyroid dysfunction	-	-	Pruritus, burning
7	57	55	2	postmenopausal	arterial hypertension	-	Traditional products, bereavement	-
8	45	43	2	premenopausal	obesity	-	-	Pruritus, burning
9	47	43	4	premenopausal	depression	FFA	divorce	pruritus
10	46	41	5	premenopausal	-	-	Traditional products	pruritus
11	49	46	3	premenopausal	depression	-	Traditional products	pruritus
12	36	35	1	premenopausal	thyroid dysfunction	-	-	-
13	59	53	6	Premenopausal, contraception	thyroid dysfunction, diabete	-	Traditional products, emotional factor	pruritus

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14	45	36	9	Premenopausal, contraception	-	-	-	pruritus
15	41	40	8	postmenopausal	postmenopausal		-	
16	57	55	2	postmenopausal	nopausal breast cancer - emotional factor		-	
17	51	42	5	postmenopausal		-		
18	62	52	10	postmenopausal	depression	-	Traditional products, emotional factor	pruritus
19	41	39	2	Premenopausal, contraception	-	AAG	Emotional factor	pruritus
20	69	65	4	Postmenopausal, contraception	-	-	-	Pruritus, burning, trichodynia
21	58	52	10	premenopausal	-	-	-	pruritis
22	66	63	3	Postmenopausal, HST	thyroid dysfunction, dyslipemia	-	Emotional factor	Pruritus, burning, trichodynia
23	49	43	6	postmenopausal	-	-	Traditional products	pruritus
24	47	39	3	Premenopausal	-	-	-	-

3.2. Clinical characteristics

Eight of our patients were phototype III (40%) and 16 patients were phototype IV (60%).

Recession of the frontotemporal hairline was found in all 24 patients (100%), this was followed by recession of the temporoparietal hairline in 70% of the cases and occipital hairline involvement in 50%. Eyebrow and eyelash were partially affected in 80% and 10% respectively. Body hair loss was found in 8 patients (20%), especially in axillary and pubic area. Lonely hairs were present in 13 patients (45%).

The facial lesions most commonly reported in our study were facial micropapules in 13 patients (45%), patchy hyperpigmentation in 10 patients (30%) and follicular hyperpigmentation in 8 patients (20%).

Seven patients (15%) had associated lichen planus with mucosal and nail involvement, and one patient (5%) had associated lichen striatus.

The majority of patients presented mild FFA (grades I and II), with a recession of less than 3 cm of the frontotemporal hairline (85%).

Clinical characteristics of the patients are shown in Table 2.

3.3. Trichoscopic features

The most frequent findings were perifollicular erythema (80%) and follicular hyperkeratosis (70%), followed by decreased or absence of vellus hairs (60%), perifollicular blue-gray pigmentation (35%), perifollicular brownish pigmentation (30%), lonely hairs (30%) and loss or absence of follicular openings (30%). Tufted hairs and white patches of scarring alopecia were found in 25% and 20% patients, respectively.

Trichoscopic features of these 24 subjects are shown in Table 3 and Figure 1.

 Table 2 Clinical characteristics

	phototype	recession of the frontotemporal hairline	mesurement (mm)	recession of the temporoparietal hairline	occipital hairline involvement	eyebrow loss	eyelash loss	body hair loss	lonely hair	facial lesions		
1	IV	+	8	+	+	+	+	+	+	follicular hyperpigmentation, facial micropapules		
2	IV	+	30	-	-	+	-	-	-	patchy hyperpigmentation		
3	III	+	5	-	-	+	-	-	-	-		
4	III	+	15	+	+	+	-	-	-	-		
5	III	+	8	+	-	+	-	-	+	facial micropapules		
6	IV	+	5	-	-	+	-	-	+	follicular hyperpigmentation		
7	IV	+	14	-	-	+	-	-	-	-		
8	III	+	20	+	-	+	-	-	-	patchy hyperpigmentation		
9	IV	+	30	+	+	+	+	+	-	facial micropapules		
10	IV	+	60	-	-	-	-	-	-	facial micropapules, patchy hyperpigmentation		
11	IV	+	20	+	-	+	-	+				
12	IV	+	5	-	-	-	-	-	-	follicular hyperpigmentation		
13	IV	+	8	-	+	-	+	+				
14	III	+	3	+	+	+	-	-	+	-		
15	IV	+	8	+	+	+	-	-	-	patchy hyperpigmentation, facial micropapules		
16	III	+	10	+	+	+	-	+	+	-		
17	IV	+	13	+	-	-	-	-	-	facial micropapules		
18	IV	+	12	+	+	+	-	+	+	follicular hyperpigmentation, facial micropapules		

19	IV	+	20	-	-	+	-	+		
20	III	+	10	+	+	+	-	-	+	-
21	IV	+	25	+	+	+	-	-	-	patchy hyperpigmentation, facial micropapules
22	IV	+	8	+	+	-	+	+		
23	III	+	12	+	+	+	-	-	+	-
24	IV	+	8	+	-	-	-	-	+	patchy hyperpigmentation, facial micropapules

Table 3 Trichoscopic features

	tufted hairs	perifollicular erythema	perrifollicular blue-gray pigmentation	perifollicular brownish pigmentation	decrease or absence of vellus hair	loss of absence of follicular openings		scarring white patches	presence of lonely hairs
1	+	+	+	-	+	-	+	-	-
2	-	+	-	-	-	-	-	+	-
3	-	+	+	+	-	+	-	-	-
4	-	+	-	-	+	-	-	+	-
5	-	+	-	-	+	-	+	-	+
6	-	-	+	-	+	+	-	-	-
7	-	+	-	-	+	-	+	-	+
8	+	+	-	+	+	+	+	+	-
9	-	+	+	+	-	-	+	-	-
10	-	+	-	-	-	-	-	-	+
11	+	-	-	+	-	+	+	+	-
12	+	+	-	-	+	-	-	+	-

13	-	+	+	-	+	-	-	-	+
14	+	+	-	-	+	-	-	-	-
15	-	+	-	+	+	+	+	-	-
16	-	+	-	-	-	-	+	+	-
17	-	-	+	-	+	-	-	-	+
18	-	+	-	-	-	-	-	-	+
19		+	+	-	-	-	-	-	-
20	+	-	-	+	-	+	+	-	+
21	+	+	-	-	+	-	-	+	-
22	-	+	+	-	-	+	-	+	+
23	-	+	-	+	+	+	+	+	-
24	-	+	+	-	+	-	+	+	+
Total	25%	70%	35%	30%	60%	30%	70%	20%	30%



Figure 1 (A): Vellus hairs; (B): Tufted hairs; (C): Perifollicular erythema; (D): Follicular hyperkeratosis

4. Discussion

Information regarding FFA in Moroccan population is still limited because of less availability of literature. Our study provided demographic data, clinical characteristics, and trichoscopic findings in Moroccan patients. The largest series was recently reported in Europe by Kanti et al., who collected 490 cases of AFF in a cross-sectional and descriptive study (2).

FFA has been known to predominantly occur in postmenopausal women; however, recent data from the literature revealed a much larger proportion of premenopausal as evidenced by the average age of our patients (1,6). Studies conducted in African patients reported higher percentages of premenopausal women (50–73%) and a lower mean age at the time of disease onset compared studies conducted in Caucasian patients (7,8). The hypothesis that a change in sex hormone balance following menopause may trigger the early onset of disease can be controversial. Additionally, female patients with FFA have not been identified to have abnormal hormone levels compared to unaffected postmenopausal women, and the disease does not exclusively affect hormone-dependent hair, as indicated by eyebrow and eyelash involvement (4,9). Moreover, rare cases have also been reported in men (10). Therefore, the association between hormonal imbalance and FFA is still inconclusive and requires further study to elucidate their relationship.

The etiopathogenesis of FFA remains unclear, it is believed that an autoimmune reaction against the pilosebaceous unit, as well as hormonal factors, may be involved in the etiopathogenesis of the disease (11). Also, previous reports have suggested genetic inheritance among several familial cases of FFA (7,12,13). However, many studies, including our study, have revealed only a small percentage of genetic inheritance of FFA (1,14). Other factors that could potentially impact FFA are the environment or emotional factors and the personal care products as shown by the use of traditional products in our study (15). Recently, Aldoori et al. suggested through an epidemiological case-control study, the responsibility of chemicals contained in sunscreens (16).

Despite the fact that FFA is now widely accepted as a variant of LPP, our study aligns with previous studies that reported low occurrences of LPP and lichen planus (1,14,17).

Regarding comorbidities, Asian and African patients showed a lower coexistence of thyroid disease with FFA; in contrast, Caucasian patients exhibited a high level of coexistence between these conditions (1,2,14,17). In our context, arterial hypertension was the most frequent found comorbidity, followed by thyroid disease.

FFA classically involves hair loss at the frontotemporal hairline. All patients in our study demonstrated hair recession in this typical area. Loss of hair at the temporoparietal hairline was also observed in our study. The involvement of the temporoparietal area was previously reported in Caucasian patients at variable frequencies but has been less observed in African patients (8,14,18). The prevalence of occipital hairline involvement in our study was found in 50% of the cases, while in the literature, it is reported in 6,7-32% of patients (1,2,14,19). Involvement of the posterior hairline has got little attention in most FFA published studies. Possible reasons are the lower frequency of disease symptoms in the occipital area compared to the frontal hairline, the lower aesthetical impact of posterior scalp alopecia and difficulties in clinical detection of hair loss in the occipital area (20). The partial or complete loss of eyebrows is another common manifestation, reported in 40– 95% of the patients, that may present before or after involvement of the scalp; sometimes, it is the only indicator of FFA (8,21,22). Our study showed high percentages of eyebrow alopecia (80%), which aligned with the results of many reports. In fact, FFA is a generalized hair disorder (23), as evidenced by the fact that 20% of our patients had depilation in other body areas.

FFA may cause pain, itching or burning sensations in the band across the frontal hairline (3). Additionally, our study reported various FFA-associated facial lesions. Interestingly, facial hyperpigmentation has predilection for patients with darker skin types as it has been reported predominantly in African, Hispanic, and South Asian patients (24–26). Histopathological evaluations have proposed that facial hyperpigmentation was associated with the prevalence of LPPig, another uncommon variation of lichen planus (26–28). However, it is difficult to conclude if LPPig is one of the clinical spectrums of FFA or if it is a distinct condition occurring coincidently with FFA. Moreover, during AFF, facial down may be affected, resulting in non-inflammatory follicular micropapules, especially on the temples (29). Such a picture was found in 45% of the cases in our series; its presence is very suggestive of the diagnosis, especially in the early forms.

The diagnosis of FFA is usually based on clinical presentation; the use of trichoscopy increases the accuracy of the diagnosis, by showing specific and sensitive signs, and helps differentiate FFA to other conditions, especially when diagnosis of the disease is doubtful (19). A study conducted in African patients revealed the absence of follicular openings in 75% of the cases (7), while preexisting data from the literature revealed that this sign was the most common

(14,17). Thus, our results suggest that the presence of prominent follicular ostia and frontal hair loss cannot rule out the existence of FFA. The "lonely hair" sign, a helpful diagnostic clue for FFA, was seen in the half of our cases (45%), which aligns with previous studies (7,17,21). Decreased or absence of vellus hairs in the hairline was seen in 60% of our patients. This absence was earlier identified by Lacarrubba et al. as a typical videodermatoscopic feature and was found in 94% of patients in their study. This finding corroborates the peculiar histopathological finding of early involvement and destruction of these follicles by a lymphocytic infiltrate (30). Our study emphasized the commonality of this process, and we believe that the absence of vellus hairs would help differentiate FFA to other forms of alopecia such as traction alopecia, androgenetic alopecia, and alopecia areata. Within the recessed hairline, perifollicular erythema and perifollicular scales were found in most of the cases; this result aligned with results of previous studies (14,17,21,31). We also observed follicular hyperpigmentation in 30% of our patients. This clinical characteristic was previously described over the affected areas in African and South Asian patients and seems to be a distinctive characteristic in patients with darkly pigmented skin (8,24). It was hypothesized that pigmentation in darker-skinned individuals may result from residual melanocytes at the scarring areas (32).

The activity and the evolution stage are also estimated by trichoscopy. Indeed, perifollicular erythema, blue-gray pigmentation and follicular hyperkeratosis, which respectively reflect inflammation, pigment incontinence and hyperkeratosis, are considered favorable prognostic factors for hair regrowth, whereas the disappearance of follicular openings and the white patches correspond to fibrosis, and thus to advanced stages of the lesions that are not easily reversible (33,34).

5. Conclusion

In conclusion, our study like that of Elloudi et al. (19), provides a better understanding of FFA in Moroccan population. We presented the demographic data, clinical manifestations and trichoscopic features of FFA patients to help diagnostic suspicion in initial cases, thus, minimizing scars and psychosocial impact.

Compliance with ethical standards

Acknowledgments

The researchers acknowledge and appreciate all patients who participated in this study

Disclosure of conflict of interest

No conflict of interest.

Statement of informed consent

Informed consent was obtained from all patients included in the study

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