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Review of clinical and demographic features of frontal fibrosing alopecia

Frontal fibrozing alopesi'nin klinik ve demografik özelliklerinin gözden geçirilmesi

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Abstract

Aim: Frontal fibrosing alopecia (FFA) is a progressive scarring alopecia that occurs on the frontotemporal area. The ethiopathogenesis of FFA is still unclear. Genetic predisposition, hormonal and environmental factors, and the use of personal care products have been put forward as etiological factors. Our aim was to evaluate the clinical and demographical features of FFA patients retrospectively during a two-year follow-up period.

Methods: Patients who admitted to our dermatology out-patient clinic between January 2016 and January 2018 were included in the study. Diagnosis was made histologically or based on the typical clinical presentation (ear to ear hair loss). Age at first presentation and onset of FFA, relevant medical comorbidities, family history, menopause status, clinical features, and medical therapies administered were recorded.

Results: All the patients were female and post-menopausal. Seven patients (87.75%) had hair loss on the eyebrows. Lonely hairs were observed in five patients (62.5%). Depression of frontal veins was observed in two patients (25%). Perifollicular erythema was observed in three patients (37.5%), and perifollicular scale was observed in four patients (50%). One of the patients (12.25%) exhibited a doll hair line. Two patients (25%) had early menopause history. The mean age of onset for graying hair was 30.87±11.39. Three patients (37.5%) had a history of premature hair graying. All the patients dyed their hair, and the mean age of hair dyeing was 31.5±10.50.

Conclusion: Perifollicular erythema and perifollicular scaling are important trichoscopic features of FFA. Environmental factors, autoimmune processes, and hormonal factors play a role in the development of FFA in genetically predisposed individuals. Premature hair graying and/or use of hair dyes (especially at early ages) can be a triggering or etiopathogenic factor for FFA.

Keywords: Cicatricial alopecia, Frontal fibrosing alopecia, Trichoscopy, Post menopause

Öz

Amaç: Frontal fibrozan alopesi (FFA) frontotemporal bölgede lokalize, ilerleyici bir sikatrisyel alopesidir. Etiyopatogenezi halen tam olarak bilinmemektedir. Genetik yatkınlık, hormonal ve çevresel faktörler, kişisel bakım ürünleri etyolojide suçlanmaktadır. Amacımız son 2 yıl içerisinde gözlenen FFA'lı olguların klinik ve demografik özelliklerinin incelenmesidir.

Yöntemler: Ocak 2016- Ocak 2018 tarihleri arasında dermatoloji polikliniğine FFA nedeniyle başvuran hastalar çalışmaya dahil edildi. Tanı histopatolojik olarak ya da tipik klinik bulgulara dayanılarak konuldu. Hastaların başvuru esnasındaki yaşı, hastalık başlangıç yaşı, ilişkili olabilecek tıbbi hastalıkları, aile öyküleri, menapoz durumları, klinik bulguları ve aldıkları tedaviler kaydedildi.

Bulgular: Hastaların hepsi kadın ve postmenapozal idi. Yedi hastanın (%87,75) kaşların da dökülme mevcuttu. Yalnız saçlar hastaların beşinde (%62,5) saptandı. Frontal venlerde çökme hastaların ikisinde (%25) gözlendi. Perifoliküler eritem üç hastada (%37,5), perifoliküler skuam dört hastada (%50) gözlenirken, bebek saçı şeklinde saç çizgisi bir hastada (%12,25) gözlendi. İki hastada (%25) erken menapoz öyküsü mevcuttu. Saçların beyazlama yaş ortalaması 30,87±11,39 idi. Üç hastada (%37,5) saçlarda erken beyazlama öyküsü mevcuttu. Tüm hastalar saçlarını boyatmıştı. İlk kez saç boyatlma yaş ortalaması 31,5±10,50 idi.

Sonuç: Perifoliküler eritem ve perifoliküler skuam FFA'nın önemli trikoskopik bulgularıdır. Çevresel faktörler, otoimmün proçesler, hormonlar genetik olarak yatkın bireylerde hastalığın ortaya çıkışını kolaylaştırmaktadır. Erken saç beyazlaması ve saç boyaları (özellikle erken yaşta maruz kalmak) FFA'da tetikleyici veya etyolojik faktör olabilir.

Anahtar kelimeler: Sikatrisyel alopesi, Frontal fibrozan alopesi, Trikoskopi, Post menapoz

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Introduction

Frontal fibrosing alopecia (FFA) is a progressive scarring alopecia that occurs on the frontotemporal area [1]. Band-like scarring alopecia localized on the frontal, temporal, and parietal hairline and a partial or complete loss of eyebrows are often observed in FFA [2,3]. The incidence of the disease has been increased since it was first described 20 years ago. There are no epidemiological data about incidence and prevalence of FFA. It is a rare form of scarring alopecia [4]. It is considered a variant of lichen planopilaris and mainly affects postmenopausal women. FFA is rarely seen in men with an incidence of 3-5% among all patients. [1,5]. The ethiopathogenesis of FFA is still unclear. Genetic predisposition, hormonal and environmental factors, and use of personal care products have been put forward as etiological factors. Early menopause and history of hysterectomy are common in FFA patients. Treatment is usually disappointing, and management of the condition can be difficult [1].

Our aim was to evaluate the clinical and demographical features of FFA patients retrospectively during a two-year follow-up period.

Materials and methods

Patients who admitted to our dermatology out-patient clinic with FFA between January 2016 and January 2018 were included in the study. Diagnosis was made histologically or based on the typical clinical presentation. Age at first presentation and onset of FFA, relevant medical comorbidities, family history, and menopause status were recorded as demographic features. Patients who started menopause before 40 years of age were classified as early menopause patients [6]. Clinical features (such as lonely hairs, facial papules, depression of frontal veins, and eyebrow and eyelash involvement) and symptoms (such as pruritus, trichodynia, and trichoscopy findings such as perifollicular erythema and scale), and medical therapies administered were recorded. Photographs of patients were taken, and each patient gave written consent. In this study, the Helsinki Declaration principles were applied.

Results

During two year periods, 16350 patients admitted to our dermatology out-patient clinic. Eight (0.048%) of these patients were FFA. All the patients with FFA were female (100%). The mean age of the patients with FFA was 63±7.11 years. All the patients with FFA were post-menopausal (100%). The mean age of onset of disease was 54.87±10.06 years. Family history was unremarkable for all patients with FFA (100%). No patients had any symptoms such as pruritus, pain, or trichodynia. Seven patients (87.75%) had hair loss on the eyebrows, whereas one patient (12.25%) had no involvement of the eyebrows. None of the patients had eyelash involvement, facial papules, or accompanying lesions compatible with lichen planus. Lonely hairs (Figure 1) were observed in five patients (62.5%). Depression of the frontal veins (Figure 2) was observed in two patients (25%).

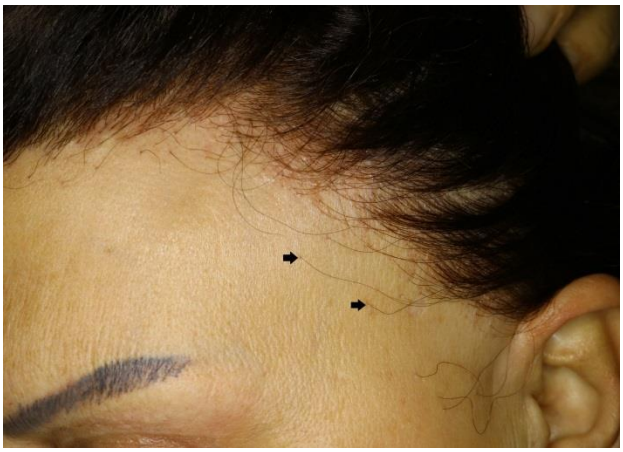

Figure 1: Lonely hairs on the atrophic background

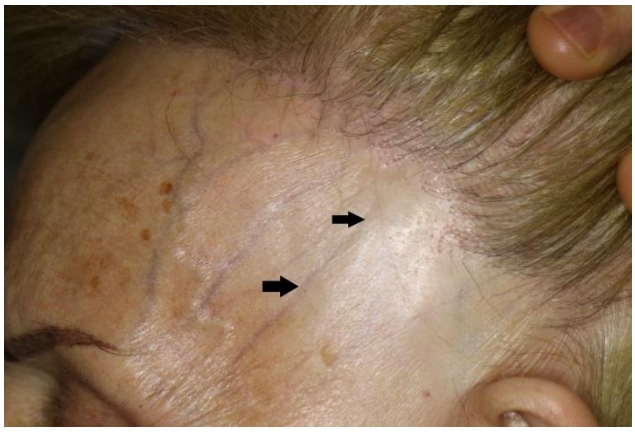

Figure 2: Depression of frontal veins on the atrophic background.

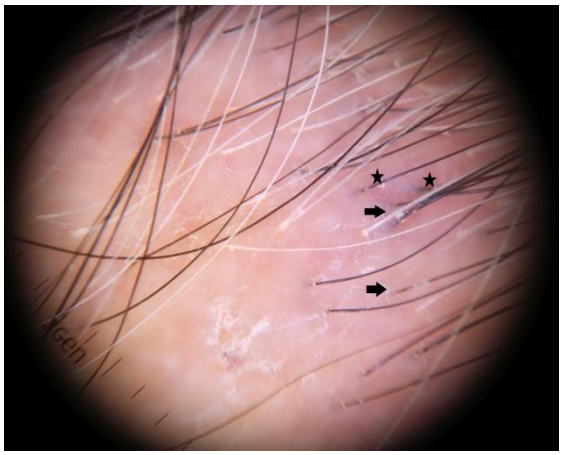

Figure 3: Perifollicular erythema (*) and perifollicular scale (arrow)

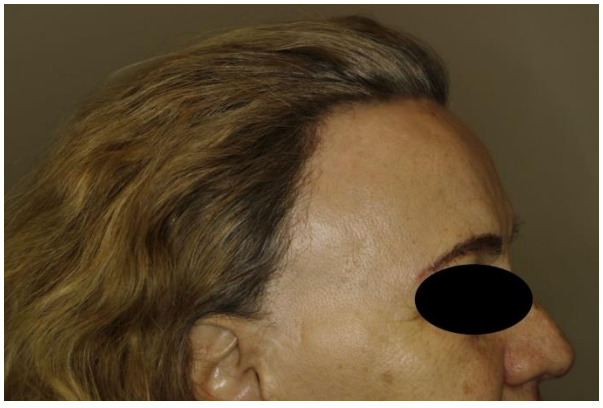

Figure 4: Doll hair line

Perifollicular erythema (Figure 3) was observed in three patients (37.5%), and perifollicular scale (Figure 3) was observed in four patients (50 %). Perifollicular hyperkeratosis was not observed in any patients upon trichoscopic investigation. One of the patients (12.25%) exhibited a doll hair line (Figure 4).

Six patients (75%) had loss of eyebrows as the initial symptom of the disease. Occipital region involvement was not observed in any patients. Two patients (25%) had thyroid disease, whereas six patients (75%) did not. The mean age at menopause was 42.25±6.60; in addition, none of the patients had a history of hysterectomy. Two patients (25%) had early menopause history. The mean age at onset of graying hair was 30.87±11.39. Three patients (37.5%) had a history of premature hair graying. All the patients dyed their hair, and the mean age of hair dyeing was 31.5±10.50. None of the patients had accompanying lichen planus lesions. The treatment methods used in our study are summarized in Table 1.

Table 1: Distribution of treatment methods in patient with FFA

Treatment methods	n (%)
Intralesional corticosteroids + topical corticosteroids + tacrolimus oinment	2 (25)
Intralesional corticosteroids + topical corticosteroids	1 (12.5)
Hyroxychloroquine + tacrolimus oinment	1 (12.5)
Topical corticosteroids	1 (12.5)
Intralesional corticosteroids + tacrolimus oinment +topical minoxidil	1 (12.5)
Topical minoxidil	1 (12.5)
No treatment	1 (12.5)

Discussion

Frontal fibrosing alopecia is a form of primary cicatricial alopecia that predominately affects women over the age of 50 and has a chronic course. In another study, the mean age of patients with FFA was 61.4, and the mean age of FFA onset was 59.8 [5]. In our study, the mean age of the patients with FFA was 63±7.11, and the mean age of FFA onset was 54.87 ±10.06 years. These results are consistent with those of other studies in the literature [7].

Trichoscopy is a non-invasive and effective technique used to distinguish FFA from other alopecias, such as alopecia areata, traction alopecia, and cicatricial margin alopecia. Absence of vellus hairs, absence of follicular openings, and presence of perifollicular scales and follicular hyperkeratosis are diagnostic features of FFA that are revealed in trichoscopic examination [8]. Especially during activation of the disease, local inflammation signs, such as perifollicular erythema and hyperkeratosis, can be observed via trichoscopic examination [7]. In our study, we detected perifollicular erythema in 37.5% of patients, and perifollicular scale was observed in 50% of patients, whereas perifollicular hyperkeratosis was not observed in any patients. This may be because the patients were under treatment. We think that trichoscopic examination is very useful for the diagnosis of FFA.

In a study, lonely hairs were found in 64% of patients, and depression of frontal veins was found in 8.7% of patients [7]. Eyebrow involvement is another common finding in FFA; the incidence of eyebrow involvement was 73–81% in the largest published series [9]. In our study, eyebrow involvement was found in 87.75% of FFA patients. In addition, lonely hairs were detected in 62.5% of patients, and depression of frontal veins was observed in 25% of patients. The incidences of lonely hairs and eyebrow involvement found in our study are similar to those in the literature, whereas the incidence of depression of frontal veins that we found was not consistent with the literature. This issue may be related to our small sample size.

Patient exhibition of a hairline reminiscent of that of a toy doll has been defined as a clinical sign of FFA. The doll hairline is highly suggestive of FFA. It is claimed that in the presence of this sign, further and more invasive investigations, such as videodermatoscopy and scalp biopsy, are unnecessary [2]. In our study, we found the doll hairline in one patient (12.5%) who had progressive FFA. We believe that the doll hair line can be a sign of progressive FFA and is not useful for early diagnosis.

A study suggested that incidences of early menopause and gynecologic surgery were higher in patients with FFA [8]. In Turkey, the average age of menopause onset is 47 years [10]. The incidence of early menopause is 1–4% in the normal population [11]. In our study, the incidence of early menopause was 25%, and the mean age at menopause onset was 42.25±6.60 in patients with FFA. FFA is not solely an immune-mediated disease. It is known that unknown hormonal mechanisms play a role in the etiopathogenesis of FFA. We found that early menopause is common in patients with FFA, which is consistent with the literature.

Environmental factors, such as the use of certain facial moisturizers, have also been put forward as triggering factors for FFA in the literature. The incidence of facial product usage, including usage of moisturizers and sunscreens, was higher in women with FFA [12].

Strazzulla et al. found that the incidence of facial moisturizer usage was 94% in male FFA patients. In the same study, the incidence of facial moisturizer usage was high in patients with FFA. They also found that the incidence of sunscreen usage was higher in the FFA group than in the control group (p=0.0012). They hypothesized that certain components of cosmetics may contribute to the development of FFA. In addition, chemical ultraviolet filters, including benzophenone-3, may have an endocrine disrupting effect, which has been shown in some animal studies [13].

In our study, we investigated the incidence of hair dyeing. The relationship between hair dyeing and FFA has never been investigated before. All our patients had a history of hair dyeing, and three patients (37.5%) had a history of hair dyeing at early ages because of premature hair graying. Hair dyes include many kinds of chemical agents. We suggest that exposure to certain chemical agents in hair dyes, like exposure to certain agents in facial moisturizers and sunscreens, could lead to the development of FFA.

Graying of the hair before the age of 20 in whites is referred to as “premature hair graying” (PGH). PGH could be associated with early exhaustion of melanocyte reservoirs due to genetic factors. This issue could be related to environmental factors, inflammation, or to psychological stress. The incidence of PGH is 26.4% in our country [14]. In our study, we found that the incidence of PHG was 37.5% in patients with FFA, which is higher than that of the normal population. In the literature, an association between FFA and PGH has not been identified. It is known that autoimmune disorders could be associated with PHG [15]. A T-cell mediated autoimmune reaction against hair follicles has been detected in FFA patients. In addition, FFA could be accompanied by autoimmune disorders like autoimmune thyroid disease [16]. We believe that PHG and FFA

could have the same pathogenetic mechanisms associated with autoimmunity.

This study has some limitations, including its retrospective design and lack of a control group. Another limitation is its small sample size.

In conclusion, FFA is a type of cicatricial alopecia that commonly affects post-menopausal women. Trichoscopic examination is a non-invasive and effective technique for the diagnosis of FFA. Perifollicular erythema and perifollicular scaling are important trichoscopic features of FFA. The etiopathogenesis of FFA is still unclear, although environmental factors, autoimmune processes, and hormonal factors appear to play a role in the development of FFA in genetically predisposed individuals. PHG and/or hair dyeing (especially at early ages) can be a triggering or etiopathogenic factor for FFA. Further studies are needed to evaluate the role of PHG and hair dyeing in the pathogenesis of FFA.

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