Ten clinical clues for the diagnosis of frontal fibrosing alopecia

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Frontal fibrosing alopecia was described by Kossard as a progressive scarring alopecia along the frontotemporal hairline.[1] It was initially thought to occur with postmenopausal women,[1-3] however, the description of its occurrence in premenopausal women has been on the rise.[4] Its incidence in male patients has rarely been described.[5-8] Although it is considered a variant of lichen planopilaris due to the similarity of the prominent histopathological findings, the pathogenesis of frontal fibrosing alopecia is not completely elucidated yet.[1,5-7,9-11] 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.[12] However, reports of familial cases suggest a genetic susceptibility with probable environmental triggers.[5,6,13,14]

The association of frontal fibrosing alopecia with androgenetic alopecia and autoimmune diseases such as hypothyroidism, vitiligo, Sjögren's syndrome and chronic cutaneous lupus erythematosus has been reported in global literature.[3,5,15,16] A relationship between frontal fibrosing alopecia and the use of facial cosmetics was recently postulated.[17] Facelifts and hair transplantion also, reportedly, trigger the disease.[18,19]

Clinically, frontal fibrosing alopecia affects the frontotemporal hairline, making its differentiation from male androgenetic alopecia and marginal alopecia areata in that location, difficult. Frontal fibrosing alopecia can also generate areas of alopecia in the auricular margins and, less frequently, may affect the occipital region.[2,3,5,7,10]

Patients may present loss of eyebrows in 50–80% of cases, involvement of eyelashes in 15% and can present with progressive and generalized disappearance of body hair.[2,3,5,11,20,21] Typically, loss of eyebrows and body hairs does not exhibit the classical inflammatory signs of erythema and perifollicular scaling.

Most patients are asymptomatic, although pruritus, pain and burning can be observed at affected sites.[5] As a primary cicatricial alopecia with a potential for irreversible damage to affected scalp areas, biopsy and histopathological confirmation is essential. Trichoscopy (dermoscopy of the scalp) and its findings increase diagnostic accuracy, and are useful in determining the best site for scalp biopsy, proving to be a valuable tool to assess disease activity, on follow-up consultations. The characteristic findings in trichoscopy of frontal fibrosing alopecia are erythema with perifollicular scaling in the peripheral part of the alopecic area, absence of vellus hairs and reduction of the number of follicular ostia. In addition, the presence of branched capillaries, honeycomb pattern of pigmentation and white dots have been described.[6,7,22,23]

Due to its insidious course, in some cases the late recognition of the disease and consequent delay in the institution of treatment could affect the evolution and prognosis of cases.[24]

The purpose of this study is to describe and illustrate 10 clinical clues that can facilitate the identification of frontal

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Figure 1: Patient with a high hair implantation line

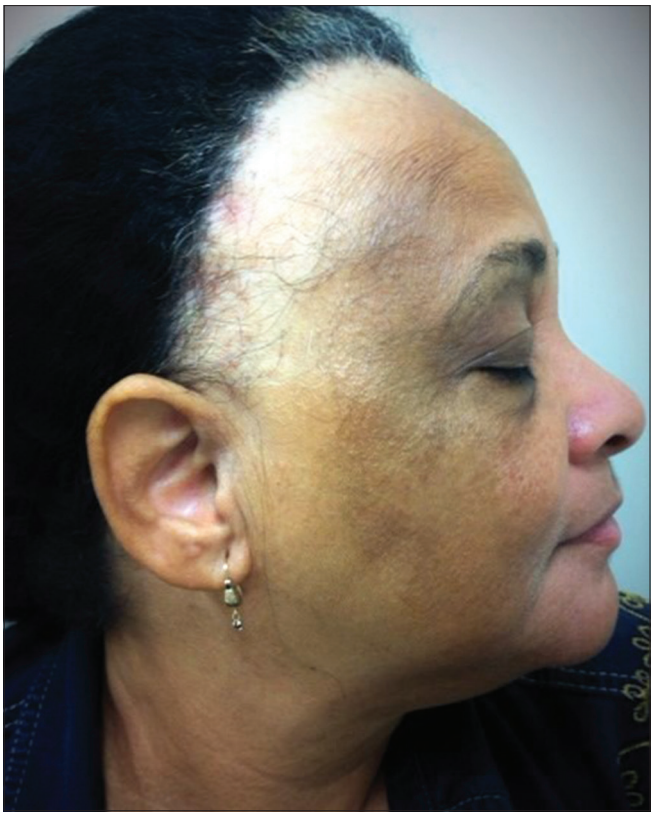

Figure 2: Patient with an evident difference in the appearance of marginal region skin

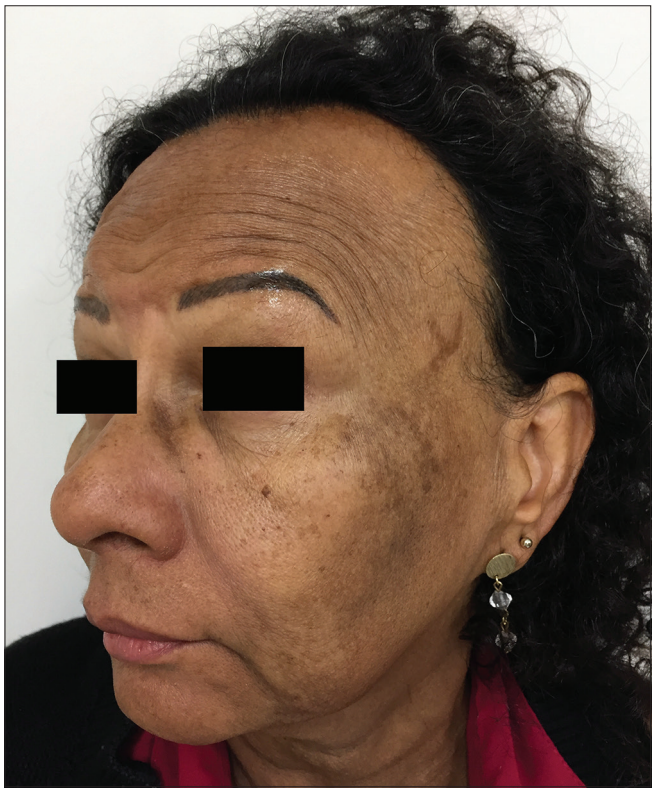

Figure 3: Patient contracting the frontal muscle, allowing the differentiation between frontal wrinkles and the marginal region of the scalp

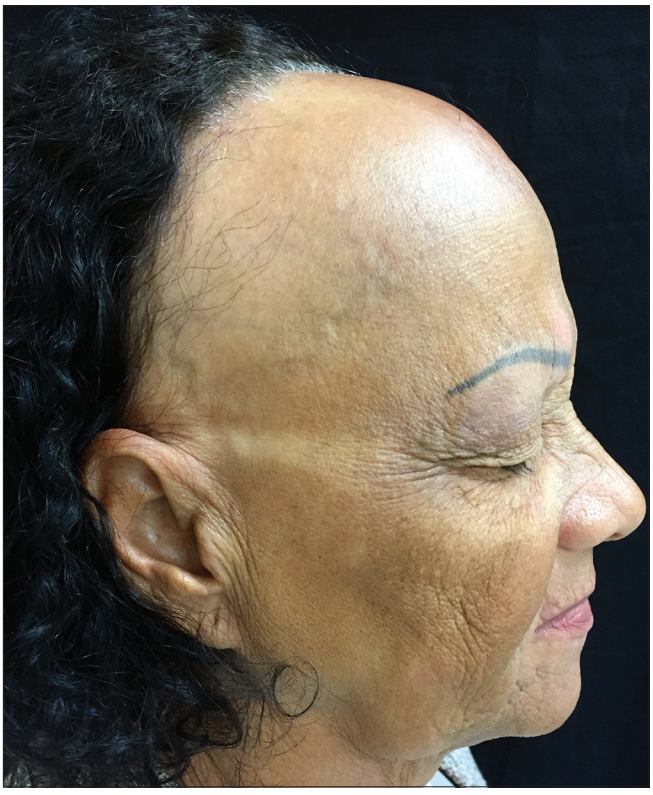

Figure 4: Patient with both eyebrow and eyelash loss. She resorted to micropigmentation of the eyebrows in order to cover up the hair loss in the region

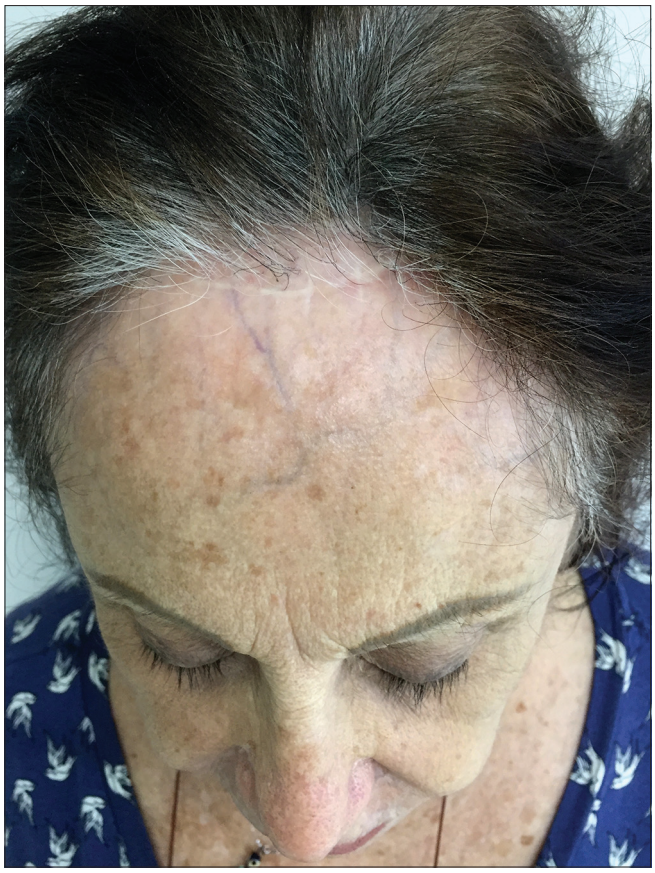

Figure 5: Patient with evident frontal veins. It is important to emphasize that this patient started the frontal fibrosing alopecia signs after having undergone a facelift procedure

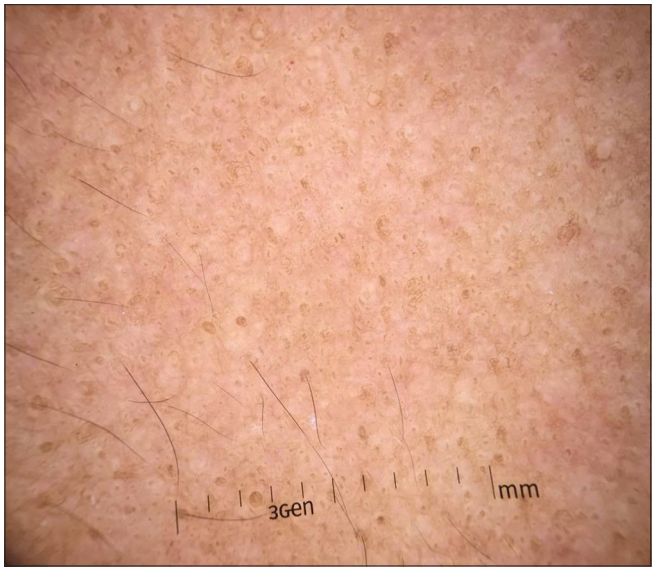

Figure 7: Trichoscopy evidencing the absence of vellus hairs in the capillary implantation line

fibrosing alopecia, aiming that residents having their first contacts with hair diseases would be able to formulate the diagnostic hypothesis of frontal fibrosing alopecia only from their clinical examination, even without complementary

Figure 6: Patient without vellus hair at the capillary implantation border

tools such as trichoscopy. Frontal fibrosing alopecia has been described only two decades ago, having increasing incidence in recent years. Proper clinical evaluation, leading to early and correct diagnosis, is fundamental to establish adequate treatment that can minimize symptoms, disease progression, scars and the impact on quality of life.[25]

1. High hairline is a critical finding for frontal fibrosing alopecia diagnosis. Fronto-parieto-temporal hairline recession[1,5-7,10,22,24] usually develops bilaterally and symmetrically,[5,10] and it can extend through to the retroauricular region and occipital margin [Figure 1]. Additionally, frontal fibrosing alopecia may cause pain, itching or burning sensations in the band across the frontal hairline.[5,7,10]
2. Skin color difference: The area affected by frontal fibrosing alopecia presents as pale skin with no follicular ostia, in contrast to normal facial skin which shows more signs of photodamage.[1] Hypopigmentation can be better visualized with the aid of a Wood's lamp [Figure 2].[26]
3. Contraction of the frontal muscle represents a simple and useful semiological maneuver that can be helpful in determining the extension of hairline recession, by precising the limits between facial and scalp areas. Contraction can be promoted by

Figure 8: Patient with isolated terminal hairs (lonely hairs) over the original hairline

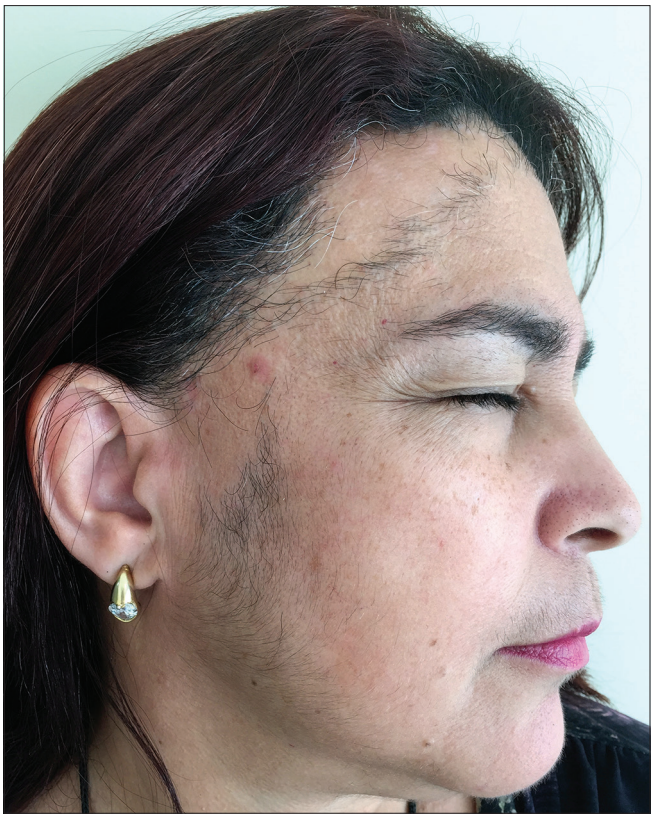

Figure 9: Patient with terminal hairs grouped over the original hairline (pseudo fringe sign)

Figure 10: Patient with facial normochromic and monomorphic papules

Figure 11: Patient with blue-grayish macules on the face, whose histopathological examination was compatible with lichen planus pigmentosus

requesting the patient to raise the eyebrows. This measure allows the differentiation between frontal wrinkles and the marginal region of the scalp, thus enabling quantification of the hairline retraction area [Figure 3].[27]

4. Bilateral eyebrow loss is another clinical warning sign for frontal fibrosing alopecia, and may represent one of the earliest manifestations which could precede alopecia by several months to years.[5,28] The distal third is commonly affected[6,7] and, unlike the scalp, it is unusual to find evident erythema or scaling on the eyebrow area.[11] Eyelash loss may also occur, albeit less frequently,[5,7,10] and appears to be related to more severe forms of frontal fibrosing alopecia.[5] This finding could, therefore, be effective as a prognostic marker and it may be considered, by some, as indicative for initiation of systemic treatment [Figure 4].[6]
5. More evident frontal veins can be detected by palpation as a localized depression, near the original hairline. Cutaneous atrophy is thought to be responsible, in addition, by the use of topical steroids [Figure 5].[26,29]
6. The disappearance of the vellus (fine, thin and light-colored hairs) at the scalp hairline is a relevant clinical and trichoscopic finding which corroborates the peculiar histopathological finding of early involvement and destruction of these follicles by a lymphocytic infiltrate [Figures 6 and 7].[11,23]
7. Lonely hairs: After progressive disappearance of vellus hairs on the frontal region, persistence of some isolated terminal hairs in the original hairline become evident, measuring 3–7 cm and may present perifollicular erythema and scaling [Figure 8].[21,25]
8. Pseudofringe sign is a retention of hairs along the frontotemporal hairline with a clinical aspect similar to the fringe sign, present in traction alopecia.[30] This sign, although infrequent, is apparently related to a good prognosis with less severe scalp involvement, greater sparing of eyebrows and less frequent facial papules [Figure 9].[31]
9. Facial papules in frontal fibrosing alopecia were described as follicular, normochromic and monomorphic papules,[32] representing the involvement of facial vellus hair, and are present in 6–37% of the cases.[5,33,34] They are randomly distributed on facial skin and are not easily noticed, being better visualized over the temples.[35] Involvement of the inframandibular and retroauricular areas can also occur, helping in differentiation from photodamaged skin.[33] These papules are associated with more severe forms of frontal fibrosing alopecia [Figure 10].[5,6]
10. Association with lichen planus pigmentosus: It is characterized by hyperpigmented macules with colors ranging from brown to blue-grayish and a reticulated or diffuse appearance, predominating over photoexposed areas. It occurs frequently in patients with higher skin phototypes and may precede the manifestations of the disease in the scalp. In the presence of facial hyperpigmentation in these patients, the possibility of lichen planus pigmentosus must be considered [Figure 11].[35-37]

Conclusion

Frontal fibrosing alopecia is a recently described disease and subsequent studies are still needed to better understand its causes and triggering factors. Its predominance in women is a well-defined aspect, and its cicatricial and irreversible character enhances the importance of early recognition.

Because there are no established diagnostic criteria for frontal fibrosing alopecia, the purpose of this study is to summarize the most relevant clinical findings of this disease by highlighting 10 clinical clues, contributing to a better recognition of the disease, and leading to diagnostic suspicion in initial cases, thus, minimizing scars and the psychosocial impact. However, it should be emphasized that studies are still needed to determine the sensitivity and specificity of these clinical findings for the diagnosis of the disease.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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