Reply to: “Updated diagnostic criteria for frontal fibrosing alopecia”

To the Editor: We thank Vaño-Galvan et al for their letter regarding criteria for frontal fibrosing alopecia (FFA).1 The differential diagnosis of FFA is broad, and diagnostic criteria can aid in identifying this entity. We agree with many of the suggestions to our diagnostic criteria proposed in our article on the Mayo Clinic cohort of FFA patients.2

We will include “in the absence of follicular keratotic papules on the body” because such lesions are in the triad of Graham Little-Piccardi-Lassueur syndrome. The lonely hair sign is moved to the minor criteria, as this is a helpful clinical tool for diagnosing FFA.3

Trichoscopy has been increasingly used for distinguishing features of FFA from other forms of alopecia, such as androgenetic alopecia.1 We added trichoscopic features to the list of minor criteria; however, upon close clinical examination, perifollicular erythema and hyperkeratosis might be seen without the use of trichoscopy (Fig 1). We agree that histopathology is not necessary for the diagnosis of FFA and have changed this to a minor criterion. However, a biopsy obtained from the frontotemporal or eyebrow regions showing histopathologic features of LPP would help the diagnosis of FFA ruling out mimickers, especially in patients with maintained brows, no symptoms or facial papules, and no clinical signs of erythema or scales.

Eyebrow involvement has been reported as a common finding in FFA (Fig 2). Early eyebrow loss before hair loss at other sites has also been reported.5,6 As the authors note, bilateral eyebrow involvement with facial papules and histopathology suggestive of FFA or lichen planopilaris would be a case in which criteria should allow for diagnosis of early FFA. With diffuse bilateral eyebrow involvement as a major criterion, such cases would meet criteria for diagnosis.

Because histopathologic examinations of patients with FFA have rarely shown features of lichen planopilaris, we agree that this feature can be removed from the criteria. We have added body hair involvement to the minor criteria.

Pruritus and pain have been identified as common presenting symptoms in patients with FFA.5,7 Therefore, we kept symptoms as a minor criterion because they might help differentiate FFA from other types of alopecia.

We appreciate the input of Vaño-Galvan et al. Compiling consensus diagnostic criteria can be challenging and have updated our diagnostic criteria for FFA (Table I).

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Table I. Revised proposed criteria for diagnosis of frontal fibrosing alopecia

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<th>Criteria</th>
<th>Major</th>
<th>Minor</th>
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<td>1. Cicatricial alopecia of the frontal, temporal, or frontotemporal scalp on examination, in the absence of follicular keratotic papules on the body</td>
<td>1. Perifollicular erythema, perifollicular hyperkeratosis, or solitary hairs on physical or trichoscopic examination in a field of frontal/frontotemporal cicatricial alopecia</td>
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<td>2. Diffuse bilateral eyebrow cicatricial alopecia</td>
<td>2. Histopathologic features of cicatricial alopecia in the pattern of FFA or LPP on biopsy*</td>
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<td>3. Involvement (hair loss, perifollicular erythema, or perifollicular hyperkeratosis) of additional FFA sites: occipital area, facial hair, sideburns, or body hair†</td>
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<td>4. Noninflammatory facial papules</td>
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<td>5. Preceding or concurrent symptoms, such as pruritus or pain, at areas of involvement</td>
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Diagnosis requires 2 major criteria or 1 major and 2 minor criteria.

FFA, Frontal fibrosing alopecia; LPP, lichen planopilaris.

*Peri-infundibular and peri-isthmal lymphoctic inflammation, interface changes at the infundibular–isthmal epithelium, peri-infundibular and peri-isthmal fibrosis, increased hair in the catagen and telogen phases, and polytrichia.
†Involvement of locations associated with LPP, such as the vertex or occipital scalp, does not preclude diagnosis of FFA.

REFERENCES


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