Frontal fibrosing alopecia preceding the development of vitiligo: A case report

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Key words: frontal fibrosing alopecia; lichen planopilaris; pigment disorders; scarring alopecia; vitiligo.

INTRODUCTION

Frontal fibrosing alopecia (FFA) is a chronic, lymphocytic, scarring alopecia involving the hair follicles of the frontal scalp, eyebrows, eyelashes, and limbs. It is closely related to lichen planopilaris (LPP), differing in its distribution on the scalp. First introduced to the literature in 1994, the incidence of both conditions is increasing, with genetic, immune, and environmental pathogenic factors proposed.[1] The pathogenesis remains unclear, yet a key component is the destruction of hair follicular stem cells through an inflammatory response at the hair bulge. Several cases were reported of FFA or LPP co-existing with vitiligo and hypothesized that the coexistence of both entities may be attributed to a shared mechanism of CD8+ cytotoxic lymphocyte infiltration.[2]

Ten reported cases of FFA or LPP co-existing with vitiligo exist in the literature.[3-8] However, in all cases, vitiligo preceded the development of scarring alopecia. Here we present an unusual case of pre-existing FFA with rapid progression after the development of vitiligo.

Abbreviations used:

FFA: frontal fibrosing alopecia
LPP: lichen planopilaris
PPD: paraphenylenediamine

CASE REPORT

A 43-year-old Egyptian woman presented to the clinic with a 3-year history of asymptomatic, progressive hair loss of the eyebrows and frontal scalp in a band-like distribution. She noted worsening of her hair loss 9 months prior, shortly after the appearance of a depigmented lesion on her right frontal hairline. She reported that the depigmented lesion gradually expanded down onto the right side of her forehead and was associated with rapid hair loss of her adjacent frontal hairline. She had no history of white skin lesions or hair loss, and she denied any other areas of hair loss or scalp symptoms.

Her medical history was significant for hypothyroidism, diabetes, anemia, and Raynaud phenomenon, all of which were well controlled. Her medications included levothyroxine, amlodipine, and ethinyl estradiol and norethindrone oral contraceptive. She denied any family history of scarring alopecia, vitiligo, type I diabetes, or autoimmune disease. She reported use of a new, self-applied hair color product that contained paraphenylenediamine (PPD) just preceding the onset of her depigmented patch.

On examination, there was recession of the frontal hairline, notable bilateral thinning of the eyebrows, and facial papules. Perifollicular erythema and scale surrounded a well-circumscribed, depigmented patch of skin with evidence of complete scarring hair loss on the right side of the forehead (Fig 1). The patient underwent initial scalp biopsy to evaluate for cicatricial alopecia. The histologic findings were consistent with that of FFA, demonstrating perifollicular fibrosis with associated perivascular and perifollicular distribution of lymphocytes and interface change. Although pathology findings were inconclusive for vitiligo, as this biopsy was performed just outside the depigmented patch to

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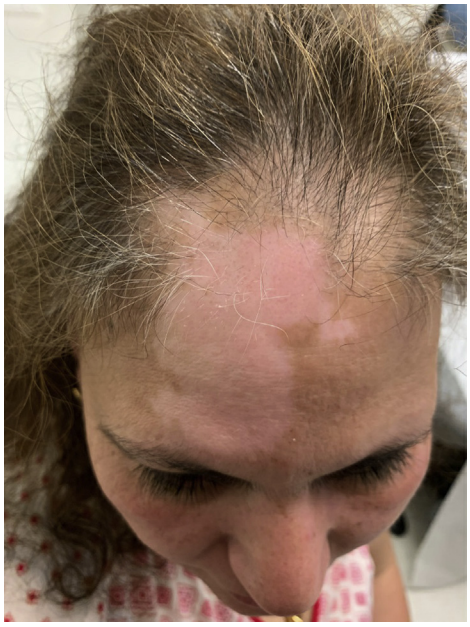

Fig 1. Vitiligo associated with complete hair loss and accelerated FFA progression.

contain hair follicles for evaluation, the clinical findings, fluorescence with Woods lamp examination, and timeline of PPD use were consistent with chemical-induced vitiligo. The patient started on oral hydroxychloroquine and topical pimecrolimus 1% cream twice daily. Additionally, she was advised to avoid PPD, an agent that uses oxidative reactions to fix color and is a known skin sensitizer.

DISCUSSION

Vitiligo and scarring alopecia are theorized to develop simultaneously, with pathogenesis beginning with the destruction of keratinocytes.[5,6] Cytotoxic lymphocytes attach to antigens in the keratinocyte basal layer and outer hair root sheath, stimulating apoptosis. It is well understood that melanocytes and keratinocytes form functional units, suggesting a mechanism for subsequent apoptosis of surrounding melanocytes and decreased melanocyte survival.[5,6,9]

Interestingly, in this case, melanocyte destruction was delayed by 2 years after FFA onset. We postulate that during the FFA-only period, an indolent inflammatory infiltrate was present. With either disease evolution or an external trigger, the inflammatory process intensified, leading to accelerated follicular stem cell destruction and localized decrease in melanocyte survival. Patient-specific risk factors, such as thyroid disease, rosacea, and possible PPD sensitization may have contributed to the abrupt inflammatory change. PPD, commonly found in hair dye and black henna tattoos, has been implicated to cause chemical-induced vitiligo.[10] In addition, recent findings suggest an increased likelihood of hypopigmentation among FFA patients.[11] Although LPP patients in the study had similar melanocyte counts within affected areas of the scalp compared with controls, FFA patients showed markedly decreased melanocyte levels contributing to hypopigmentation.[11]

The true nature of FFA remains highly debated and poorly understood. Yet it is likely multifactorial, with immune pathways, hormonal influences, and environmental factors playing roles. This case highlights an interesting deviation from previous reports of LPP/FFA-vitiligo coexistence, challenging previously proposed etiologies. It provides evidence of a mechanism for immune conditions to derive from existing scarring alopecia, rather than the reverse. Among ever-evolving theories, one thing is for certain, the relationship is complex. Further studies are warranted to prevent and slow disease progression in this patient population.

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