Dermpath Quiz: Which alopecia does the patient have?

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A 60-year-old woman presented with a longstanding history of bilateral hair loss of the anterior frontoparietal hairline [Figure 1]. Punch biopsies were performed for histological analysis [Figure 2a-d].

The sections most likely represent:

A. Trichotillomania

B. Frontal Fibrosing Alopecia

C. Traction Alopecia

D. Alopecia Areata

E. Central Centrifugal Cicatricial Alopecia

ANSWER

B. Frontal Fibrosing Alopecia

DISCUSSION

The histopathological sections demonstrated superficial dermal fibrosis and a focal peri-infundibular lymphoid infiltrate. Elastic tissue staining demonstrates a superficial wedge-shaped peri-infundibular scar. Given the clinical appearance, the changes are diagnostic of frontal fibrosing alopecia (FFA).

First described by Kossard in 1994, FFA represents a cicatricial alopecia with follicular destruction and is clinically distinguished by striking bilateral frontoparietal hairline recession.[1] It is considered to be a variant of lichen planopilaris (LPP). FFA is more frequently seen in postmenopausal women with the overall prevalence in premenopausal women being 6%.[2,3] Interestingly, the youngest reported patient with FFA is a 17-year-old female and this further emphasizes the uncertainty of the etiopathogenesis of FFA.[3] Clinically, the loss of hair is gradual, over several years, asymptomatic and typically involves the bilateral frontoparietal hairline. This occurs in a symmetrical pattern; the alopecic band of hair can reach up to 8 cm in diameter and give an appearance of a "clown" pattern of alopecia.[2] Furthermore, in up to 60% of published FFA cases in the literature, physical exam findings such as scarring of the alopecic skin, perifollicular erythematous papules, and loss of eyebrow hair have been noted.[2] A case series by Dlova has reported that more than 50% of ethnic patients with FFA had coexisting lichen planopilaris.[4] Thus rightly, Berliner *et al*. recommended an examination of all the patients with FFA for coexisting LPP and vice versa.[5] In cases of FFA, the differential diagnosis includes lichen planopilaris, traction alopecia, and ophiasis inversus. Given the scarring nature of FFA, other cicatricial alopecias may be entertained in the differential diagnosis, such as discoid lupus erythematosus, central centrifugal cicatricial alopecia, and pseudopelade of Brocq,[5] but usually the distinction with the latter two is made clinically.

Noted histological findings in FFA include reduction of the terminal hair follicles and their replacement by fibrous tracts. Usually, there is a lymphocytic infiltrate centered at the level of the isthmus and the infundibulum of the hair follicle. The hair bulb remains spared. Interestingly, the lymphoid infiltrate surrounds the perifollicular fibrosis with very few lymphocytes being interspersed within the fibrotic area.[1] The sweat glands and the overlying epidermis are unremarkable in FFA and there is commonly no lichenoid inflammation or interface change at the stage when the biopsy is done.[1] Early lesions of FFA histologically may only demonstrate perifollicular mucinous fibrosis and/or focal peri-infundibular lymphoid inflammation. Late lesions may lack

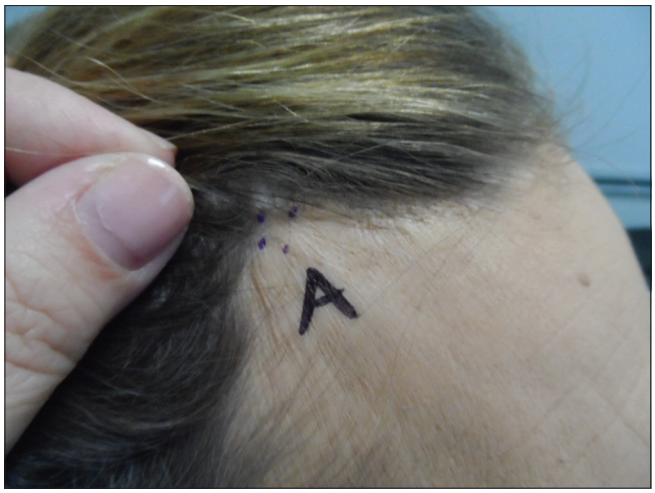

Figure 1: Bilateral hair loss of the anterior frontoparietal hairline

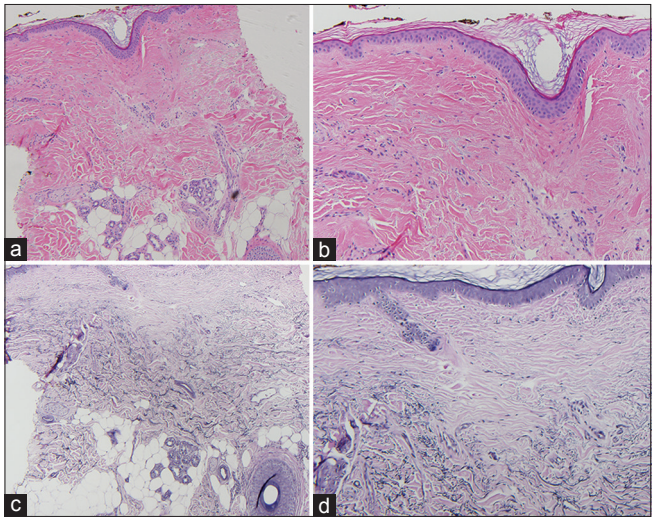

Figure 2: (a) H and E, ×40, (b) H and E, ×200, (c) EVG stain, ×40, (d) EVG stain, ×200

inflammation and demonstrate only wedge-shaped superficial scars at the infundibular level. Therefore, elastic tissue staining with elastic van Gieson (EVG) stain is particularly helpful in diagnosing biopsies of burnt-out patches of FFA by highlighting the superficial peri-infundibular scar in the dermis.[6] This pattern of scarring is characteristic of FFA and LPP, and helps to differentiate them from other scarring alopecias such as discoid lupus erythematosus and central centrifugal cicatricial alopecia.

As with other cicatrical alopecias, FFA remains a challenging clinical entity to treat. The temporal association with menopause of presence of kinky "genitalized" hairs at the margin suggests that hormonal treatments could be of benefit and roughly half of all patients respond to oral dutasteride.[7] Other treatment options include topical, oral, and intralesional steroids, pioglitazone, antimalarials, and retinoids.

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