

Chapter 4

A 26-Year-Old Woman with Multiple Erythematous Areas with Coexisted Hair Loss



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A 26-year-old woman presented with a six-month history of multiple erythematous lesions with coexisted hair loss and itching. No history of dermatologic or non-dermatological diseases was reported.

A physical examination revealed an area of non-scarring hair loss (9 cm × 6 cm) with the central erythema on the vertex and occipital areas (Fig. 4.1). Moreover erythematous lesions with follicular keratosis and coexisted hair loss on the trunk, upper and lower extremities were presented. A histopathological examination showed a mucin deposition within the hair follicles and sebaceous glands.

Based on the case description and the photographs, what is your diagnosis?

Differential Diagnoses

1. Mycosis fungoides.
2. Alopecia areata.
3. Primary follicular mucinosis.
4. Graham-Little syndrome.

Diagnosis

Primary follicular mucinosis.

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Fig. 4.1 A 26-year old woman with an area of non-scarring hair loss with the central erythema on the vertex and occipital areas



Discussion

Follicular mucinosis, also known as alopecia mucinosa, is a form of cutaneous mucinosis characterized by accumulation of dermal type of mucin in the pilosebaceous follicle and sebaceous glands [1]. Pathogenesis of the disease is not fully understood. There are two main clinicopathological forms of follicular mucinosis: a primary, benign variant and a secondary [1]. The primary form affects mainly children and young adults with the age range from 11 to 35 years [2]. Clinically, the disease is characterized by the presence of well-delimited, erythematous or brownish-erythematous papules, patches or plaques. Follicular keratosis or areas of alopecia may be presented. Hair loss in the course of follicular mucinosis is typically non-scarring and reversible; in very rare cases, scarring may occur [3]. The disease can be observed in any body area, however the scalp, neck and upper extremities are most commonly affected [1]. Other clinical variants of primary follicular mucinosis include the urticaria-like and acneiform. The diagnosis of follicular mucinosis is based on the clinicopathologic correlation. In histopathologic examination mucin deposits on the outer root sheath of the hair follicle, in addition to inflammatory infiltrates composed of lymphocytes, macrophages and eosinophils with folliculotropic lymphocytes are presented [4]. Mild-to-moderate potency topical corticosteroids are usually the first-line therapy for primary follicular mucinosis. Other therapeutic modalities include topical and oral antibiotics, retinoids, dapsone, imiquimod, topical pimecrolimus, and psoralen plus ultraviolet A (PUVA).

Differential diagnoses for the presented patient included mycosis fungoides, alopecia areata and Graham-Little syndrome.

Mycosis fungoides is the most common type of primary cutaneous T-cell lymphoma. It occurs more frequently in older adults and is more commonly observed in men compared to women. There are three main stages of cutaneous involvement of mucosis fungoides: patchy-, plaque, and tumor-stage. Lesions affects mainly photo-protected areas, in particular the buttocks, groin, breasts, upper thighs, and axilla and, less commonly, the distal extremities, head, and neck [4].

Alopecia areata, a form of non-scarring autoimmune hair loss, is characterized by the presence of hair loss areas within the skin which remains normal. Although the scalp is most commonly affected, hair loss can also be observed in other hair-bearing areas (such as eyebrows, eyelashes, pubic and axillary areas) [5].

Graham-Little syndrome, a variant of lichen planopilaris, is characterized by a triad consisting of patchy cicatricial alopecia of the scalp, non-cicatricial axillary and pubic hair loss and lichenoid follicular eruption. Middle-aged women are most commonly affected [3].

Based on the clinical manifestation and histopathological findings the diagnosis of primary follicular mucinosis was established. PUVA therapy and topical mometasone furoate were initiated.

Key Points

- Primary follicular mucinosis is a form of follicular mucinosis that affects mainly children and young adults
- It is characterized by the presence of well-delimited, erythematous or brownish-erythematous papules or plaques with or without alopecia

References

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