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Pathomorphological approach to alopetic lesions on the scalp

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Hair loss is a common disorder but without enough knowledge in the evaluation of hair diseases. The differential diagnosis includes a lot of disorders causing cicatricial or noncicatricial alopecias.

We present two casuistic cases of patients with alopetic lesions on the scalp. The clinical examination signifies alopecia areata in both cases but with disappearance of visible follicular ostia. Pathomorphological findings clarifies the lesions as scarring or cicatricial alopecias, an uncommon and clinically diverse set of disorders that result in permanent and irreversible hair loss.

Key words: alopecia, cicatricial alopecia, sclerodermia "en coup de sabre", psoriasis vulgaris, lupus erythemathosus capilitii

Introduction

Alopecias of the scalp is divided into two groups, namely non-scarring or non-cicatricial and scarring or cicatricial forms. Pathomorphologically, a scar constitutes the end point of reparative fibrosis with permanent destruction of the preexisting tissue [4].

Non-scaring alopecia represents a complex group of hair disorders in which the hair shafts fall out, but the hair follicles are still alive and from this point of view the prognosis is better. The clinical entities include androgenic alopecia, telogen effluvium, alopecia areata, trichotillomania and traction alopecia.

Cicatricial alopecia encompasses a group of disorders characterized by permanent destruction of the hair follicle, which is replaced by fibrous tissue leading to progressive and permanent hair loss. According to the initially location of the inflammatory process cicatrical alopecia can be primary and secondary. In primary cicatricial alopecias the pilosebaceous follicle is the target of the destructive process. In secondary forms the follicular destruction is a consequence, and due to numerous aetiologies (Table 1) [1].

Table 1. **Secondary scarring alopecia**

- Aplasia cutis congenita
- Cicatricial bullous pemphigoid
- Infectious (kerion, staphylococcal)
- Neoplastic (primary, metastasis)
- Connective tissue disease (morphea)

- Trauma (burn)
- Metabolic (amyloid, mucin)
- Granulomatous (sarcoid)

Primary cicatricial alopecias are divided into several categories, including lymphocytic, neutrophilic and mixed (Table 2) according the current working classification proposed by the North American Hair Research Society [3]. The cause and pathogenesis are largely unknown, but the target is the hair follicle itself [6].

Table 2. **Working classification proposed by the North American Hair Research Society**

1. Lymphocytic
- Chronic cutaneous lupus erythematosus
- Lichen planopilaris (LPP)
  - Classic LPP
  - Frontal fibrosing alopecia
  - Graham–Little syndrome
- Classic pseudopelade (Brocq)
- Central centrifugal cicatricial alopecia
- Alopecia mucinosa
- Keratosis follicularis spinulosa decalvans

1. Neutrophilic
- Folliculitis decalvans
- Dissecting cellulitis / folliculitis (perifolliculitis capitis abscedens et suffodiens)

1. Mixed
- Folliculitis (acne) keloidalis
- Folliculitis (acne) necrotica
- Erosive pustular dermatosis

Materials and Methods

We present two different cases of alopetic lesions of the scalp.

First case is nine years old girl. She complains of hairless well-defined solitary lesion with linear form localized on the fronto-parietal part of the scalp. She and her parents treated it according medical prescription with local creams and solutions almost one year without visible effect.

Second case is 67 years old woman. She has red raised skin plaques covered with silvery white squamas on the skin of the knees, elbows and trunk. She is diagnosed as having psoriasis and the diagnosis is clinically and histological proven more than ten years ago. She has also psoriatic lesions on the scalp, presented as inflammatory plaques with white squamas which are not hairless. Last two months the patient has noticed another type of skin lesions on her capillicium, characterized with small oval alopetic maculae with very soften and thin skin without any visible hair.

Both patients followed laboratory and immunological investigations. The girl followed also examination of X-ray cranium and study for antibodies for Borrelia burgdoferi. All the results were in normal limits.

Skin biopsies of hairless lesions were obtained from the two patients. The biopsy tissue was fixed in 10% neutral buffered formaldehyde solution, processed routinely, and stained with hematoxyline-eosin.

Results and discussion

The histology of the first patient demonstrate thickening and moderate hyalinization of the collagen in middle and lower part of the dermis. Perivascular infiltrate are seen in middle and upper dermis (Fig. 1).

The clinical and histological characteristics of the skin lesions lead us to diagnosis of linear scleroderma, often called scleroderma en coup de sabre (SCS) for its resemblance to a sword-strike scar. The differential diagnosis include chronic cutaneous lupus erythematosus, frontal fibrosing cicatricial alopecia, pseudopelade of Brocq, or lichen sclerosus et atrophicus. The lack of surface changes, such as scaling, follicular plugging or perifollicular erythema, the linearity of the lesion and the extension of sclerosis onto the forehead help to distinguish it from the other alopetic lesions. According to Soma et al frontoparietal scleroderma, may occur along the lines of Blaschko [5]. Since both the unilateral distribution and the lesions along Blaschko's lines are the patterns created by genetic mosaicism, they suggest that a significant part of linear scleroderma and perhaps a smaller part of multiple morphea could be related to cutaneous mosaicism. We follow the patient till now because 25% of the cases with SCS could be worsened with Central Nervous System complications [2]. Alopetic lesion persists as it is classified as secondary cicatricial alopecia due to connective tissue disease- morphea but fortunately there is not involvement of any other organ or system.

Two biopsies were performed from the skin of the second patient. The first one was obtained from a lesion localized on the hand. The findings demonstrates hyper and parakeratosis, epidermal acathosis and elongation of rete ridges and moderate perivascular inflammatory infiltrate in upper derma (Fig. 2). All these characteristics are typical for psoriasis. The other biopsy was taken from alopetic lesion of the scalp. The findings were quite different and showed epidermal atrophy, moderate vacuole degeneration of the basal layer and lymphocytic infiltrate in upper derma (Fig.3). Direct immunofluo-

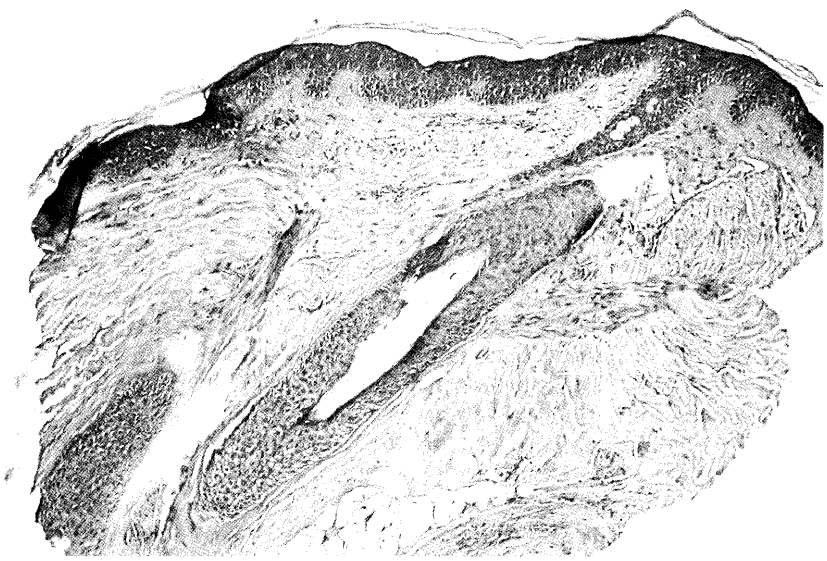

Fig. 1 Thickened connective tissue septae in hypoderma and hyalinization of the collagen (HE, x 20)

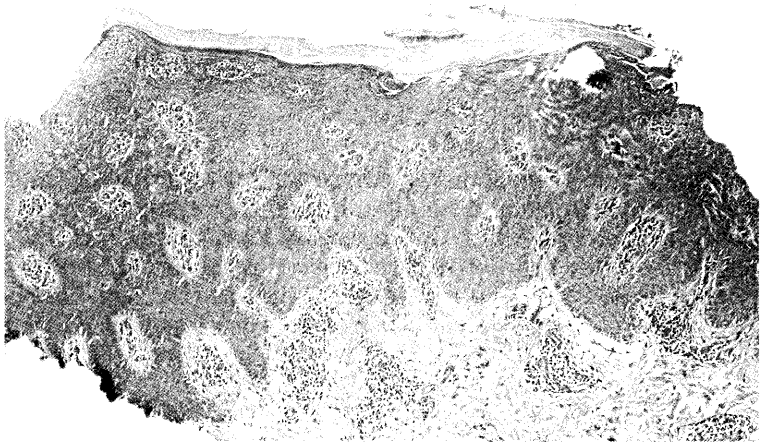

Fig. 2. Hyper and parakeratosis, epidermal acathosis and elongation of rete ridges and moderate perivascular inflammatory infiltrate in upper derma (HE, x 20)

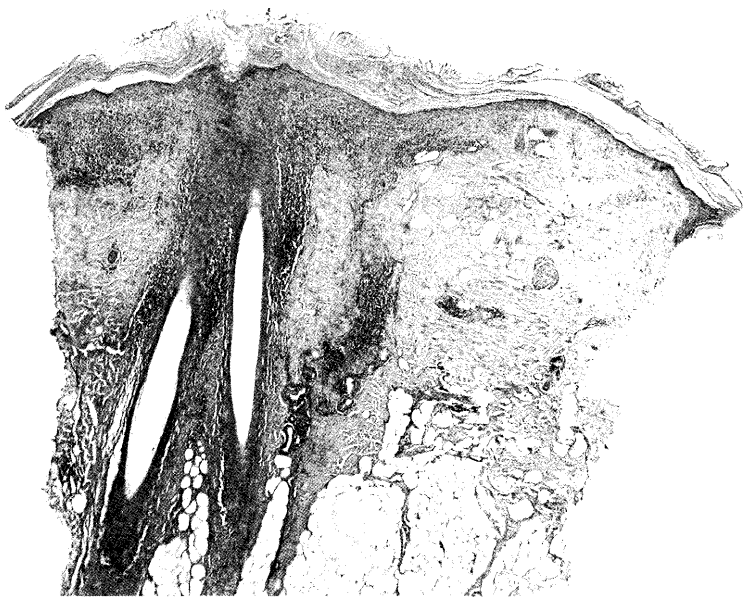

Fig.3. Epidermal atrophy, vacuole degeneration of the basal layer and lymphocytic infiltrate in upper derma (HE, x 20)

rescense revealed granular Ig M (+) deposit on the layer of dermo-epidermal junction. These findings correlate with the diagnosis of cutaneous form of lupus erythematosus chronicus discoides (LECD). We conclude that this patient has psoriasis vulgaris in association with LECD on the scalp. The two diseases are very different from clinical, morphological and therapeutic point of view. However, they both have multifactor immunological background and their pathogenetic pathway is disputable. Clearly is

that the alopetic lesions are due to LECD and are classified to lymphocytic subtype of primary cicatricial alopecia.

Conclusion

Alopetic lesions on the scalp are clinically similar but pathomorphologicaly quite different. Their correct interpretation from all points of view leads to correct diagnosis and consequently to correct treatment. Our cases have histologic and clinical overlap of cicatricial alopecia – due to a connective tissue disease, secondary subtype and lymphocytic primary subtype in association with systemic disease. The both have permanent destruction of the hair follicle that result in irreversible hair loss.

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