Linear Alopecia Mucinosa Along Blaschko Lines

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We present a case of linear reversible non-inflammatory alopecia mucinosa in a 25-year-old Nigerian patient. The patient exhibited an arciform bald patch on the scalp and follicular mucinosis in both lesions. The possible relationships between the unusual linear arrangement of alopecia mucinosa along Blaschko's lines and lichen striatus are discussed. Key words: Follicular Mucinosis; Lichen striatus.

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Several genetic and acquired dermatological disorders can show a linear arrangement. A common pattern of distribution of linear skin diseases follows the Blaschko lines, which in human beings probably reflect functional X-chromosome mosaicism (1–2). We report a case of reversible alopecia mucinosa which followed a linear arrangement along the Blaschko lines.

CASE REPORT

A 25-year-old Nigerian patient consulted us in June 1989 because of an odd, bizarre-shaped arciform bald patch on the right side of the scalp. Anamnesis revealed that the patient had noticed a progressive thinning of the hair on the right parietal side of his scalp 5 months earlier. He did not complain of itching, tingling or pain.

The clinical examination revealed the presence of an arciform bald patch (23 cm long and 4 cm wide) on the right parietal side of the scalp (Fig. 1). The skin of the involved area was apparently normal, with no signs of erythema, infiltration or follicular plugging. Several hair shafts of various length appeared broken. No exclamation point hairs were seen. This clinical picture suggested a traction alopecia.

A careful clinical examination showed the presence of follicular papules with linear arrangement on the right lateral aspect of the trunk (Fig. 2). The patient was not aware of the presence of these lesions and did not have other cutaneous or mucosal abnormalities. His clinical history was not remarkable except for malaria at the age of 16. Laboratory investigations including a complete blood count, serum electrolyte levels, serum thyroxin and triiodothyronin indexes were normal. Chest X-ray examination was negative.

A scalp biopsy revealed a normal density of hair follicles in different phases of the hair cycle. The dermis showed the presence of an inflammatory lymphocytic infiltration with exocytosis which surrounded catagen and telogen follicles as well as the upper portion of the anagen follicles. Reticular degeneration with formation of cystic spaces was evident in the outer root sheaths of anagen follicles (Fig. 3). The epithelial sac of telogen follicles was enlarged and showed large cystic spaces containing a granular basophilic material (Fig. 4). Giemsa and Alcian Blue stains confirmed the deposition of mucin within the hair follicles. A punch biopsy obtained from follicular papules on the trunk showed perifollicular lymphocytic infiltration associated with mucin deposition within the outer root sheaths.

The patient was not treated but was re-examined at monthly intervals. After 6 months his hairs had completely regrown and after 1 year the follicular papules on the trunk had regressed, leaving a residual hyperpigmentation.

DISCUSSION

Alopecia mucinosa is a clinical condition of unknown etiology in which hair loss is due to a distinctive pathological follicular alteration: follicular mucinosis (3,4). Alopecia mucinosa is usually characterized by slight infiltrated erythematous plaques that often show scaling and follicular plugging (5,6). Cicatricial alopecia can be seen in persistent lesions as the result of follicular destruction. Alopecia mucinosa can however occasionally cause a loss of scalp hair, with normal-appearing scalp skin as in our patient and in the patient reported by Snyder et al. in 1984 (7).

The absence of skin erythema and infiltration in these cases can perhaps be explained by the low density of the inflammatory infiltration. In our patient, in fact, the lymphocytic infiltration was inconspicuous and affected almost exclusively the perifollicular dermis and the hair follicles. The linear arrangement of both the bald patch and the follicular papules constitute to our knowledge a unique observation. As a matter of fact, the topographic distribution of these lesions followed the lines of Blaschko both on the scalp and on the trunk.

Although there are many acquired linear lesions of the skin which can follow the Blaschko lines, such as psoriasis, eczema, lichen planus and lichen striatus, no cases of linear alopecia mucinosa have hitherto been reported. Follicular mucinosis is a root sheath mucinosis, described first by Kreibich in 1925 (8), which is thought to represent a reactive pattern of follicular epithelium. This pathological pattern can be observed in association with numerous conditions other than alopecia mu-

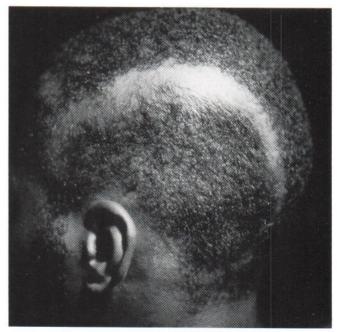


Fig. 1. Arciform bald patch on right side of the scalp.

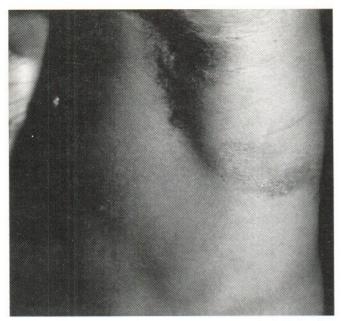


Fig. 2. Follicular papules with linear arrangement on the lateral aspect of the trunk.

cinosa, such as mycosis fungoides, angiolymphoid hyperplasia with eosinophilia, and eczema (9). Electron microscopy studies have suggested that mucin is produced by follicular keratinocytes (10). The observation that follicular mucinosis is invariably associated with a predominantly lymphocytic inflammatory infiltration which affects follicular structures supports the view that mucin is produced by hair follicle keratinocytes stimulated by T-lymphocytes (11). Distribution of inflammatory skin conditions in linear streaks has been extensively reported in the literature, both as part of a more widespread picture or as an entirely linear distribution.

Lichen striatus is an entirely linear inflammatory skin condition which often appears following the lines of Blaschko (12).

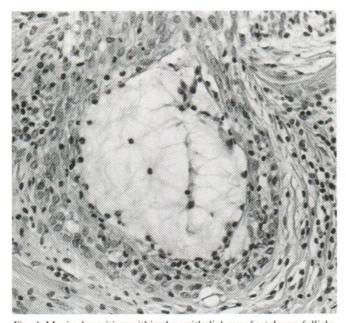


Fig. 4. Mucin deposition within the epithelial sac of a telogen follicle. H&E, ×250.

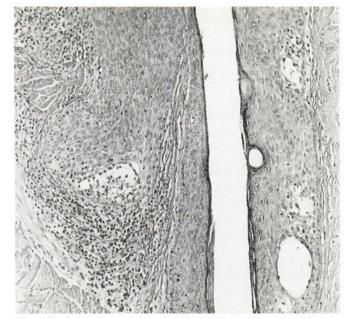


Fig. 3. Reticular degeneration associated with lymphocytic infiltration of the outer root sheath of an anagen follicle. H&E, ×125.

The pathological picture of lichen striatus is characterized by an inflammatory lymphocytic infiltration in the superficial dermis, with exocytosis in the epidermis. In rare cases, the infiltrate may affect eccrine glands and pilosebaceous units (13). In our patient the entirely linear distribution of alopecia mucinosa along the lines of Blaschko may suggest some relationship between lichen striatus and alopecia mucinosa. We may speculate that in this case the lymphocytic infiltration of lichen striatus selectively affected hair follicles inducing mucin production. The possible occurrence of follicular mucinosis in lichen striatus has been sporadically reported in the literature (9). According to this hypothesis, our case could be interpreted as one of alopecia mucinosa secondary to lichen striatus.

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