doi: 10.1111/j.1365-4632.2004.02078.x

Cameo

Alopecia totalis incognito

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We report an 11-year-old boy with a strong family history of alopecia areata who initially developed alopecia areata with a single circular patch of hair loss on the scalp with exclamation mark hairs at the periphery, which evolved over time into alopecia totalis and then into a novel pattern of hair growth with fine depigmented hair, uniformly 5 mm in length. The hair has not grown any longer over a 48-month follow-up period. Scalp biopsies from the occipital scalp demonstrate dense peribulbar lymphocytic infiltrate and uniform miniaturized secondary vellus hairs. This previously undescribed pattern of alopecia areata is remarkable for total lack of alopecia.

Introduction

The Greek word alopecia refers to hair loss from any source, while the Latin word areata means occurring in patches or circumscribed areas. Alopecia areata is a familial, chronic-relapsing, organ-specific autoimmune disease affecting terminal anagen hair growth. Disruption of hair growth leads to baldness either in patches, diffusely or totally. Patchy hair loss may be patterned, and a number of terms such as ophiasis and sisaipho (the reverse of ophiasis) have been used to describe these patterns. The hair loss is most common on the scalp, but may occur anywhere on the body. Individual patches may coalesce to form total hair loss of the scalp or universal hair loss on the body. Also total hair loss may regrow incompletely to leave one or more circular patches of hair loss. The fluidity of the pattern of hair loss in individual patients has resulted in the term alopecia areata being applied to all patterns of hair loss seen with this disease. Other than the distinctive exclamation mark hairs that may be seen within patches, the scalp is totally bald in affected areas.

Depigmented hairs are commonly spared in the initial hair loss and regrowth, either spontaneous or medically assisted, and may be initially depigmented.

On histology affected terminal anagen hairs have a dense peribulbar lymphocytic (T cell) infiltrate, and there is an increased number of catagen and telogen hairs. Uniform miniaturization of terminal hairs into secondary vellus hairs is also a prominent feature. While miniaturization of hairs is also a feature of androgenetic alopecia, this patterned hair loss is confined to the crown.

We report an 11-year-old boy with a novel presentation of alopecia areata, sine alopecia. He has a fine down of depigmented hair, approximately 5 mm in length, all over the scalp.

Case Report

An 11-year-old boy first developed alopecia areata at the age of 18 months. This manifested initially with a solitary patch of hair loss on the parietal scalp, which was followed rapidly by several other patches of hair loss on the fronto-vertical scalp. Complete regrowth of all patches occurred spontaneously within 6 months. He was disease free until 4 years of age. Relapse was initially patchy, bur the hair loss became progressively confluent over 2 months and evolved into alopecia totalis (Fig. 1). His eyebrows and eyelashes were spared. His nails were normal.

There was no personal history of atopy or organ-specific autoimmune disease, however, there is a strong family history of both alopecia areata and thyroid disease. His mother, maternal grandmother, maternal uncle and both his siblings all have alopecia areata. His father has auto-immune thyroid disease. One sister has juvenile arthritis as well as alopecia areata.

Topical immunotherapy with: diphencypropenone (DPCP), 3,4-dihydroxy-3-cyclobutene-1,2 dione3 (squaric acid) and dinitrochlorobenzene (DNCB) all failed to regrow any hair and were each abandoned after 6 months. Oral prednisolone (initial dose 37.5 mg per day reducing to 0 mg over 8 weeks), topical PUVA (stopped after 23 treatments), topical anthralin daily for 6 months and topical minoxidil twice daily for

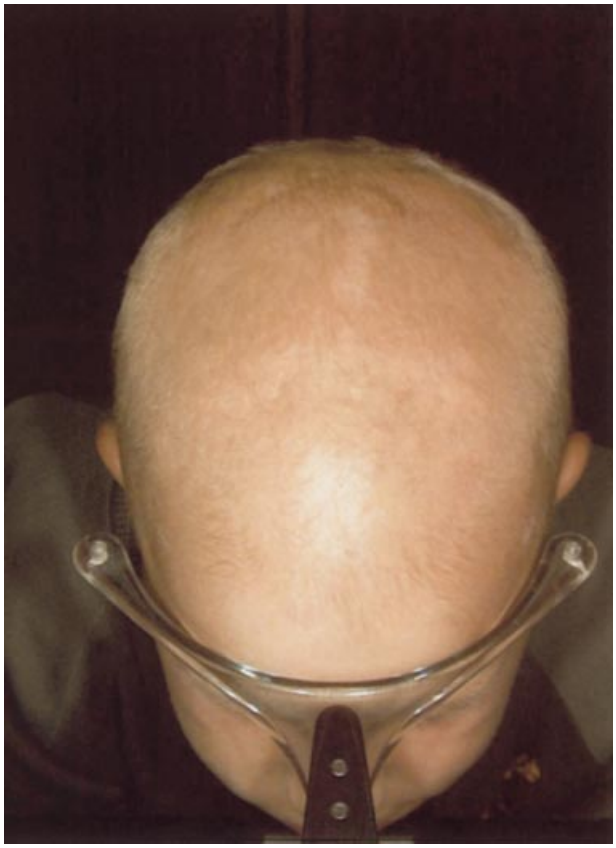

Figure 1 Vertex of the scalp showing fine downy hair

Figure 2 Fine downy telogen hairs on the inside of this subject's cap, indicating that the hair is cycling with a shortened anagen phase

6 months were all unsuccessful. No further treatment was initiated.

Six months after stopping all treatment his hair regrew spontaneously over his entire scalp. However the regrowth consists of fine, short and depigmented hair that fails to grow longer than 5 mm and never requires cutting (Fig. 2). The hairs have tapered tips.

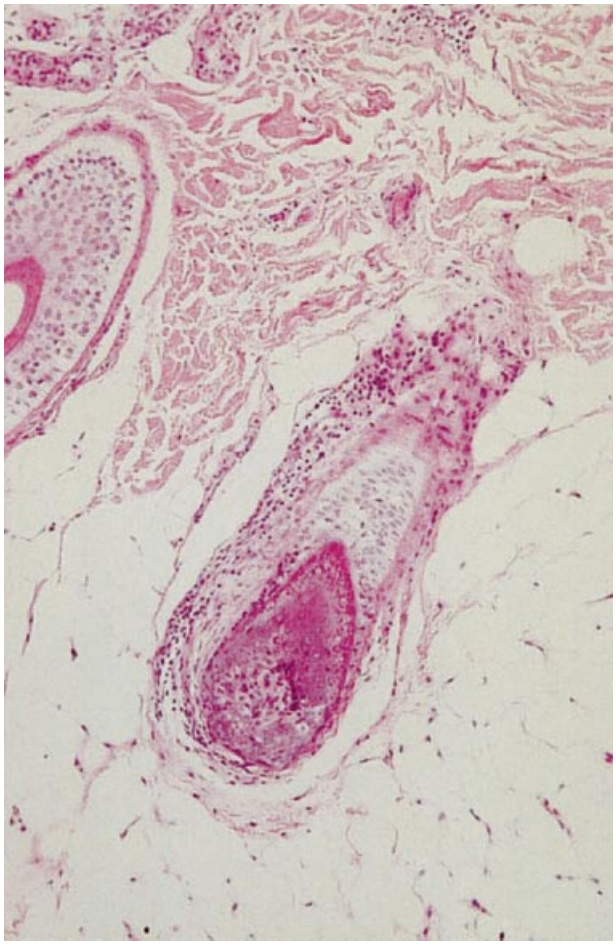

Figure 3 Histology of the occipital scalp in a vertical cross-section showing a peribulbar lymphocytic inflammatory infiltrate

A hair pull test was negative. On hair pluck the hairs were painful to remove. Examination of the bulbs demonstrated that the hairs were in anagen, and therefore not exclamation mark hairs.

Treatment with topical minoxidil was re-initiated in an attempt to prolong anagen duration and promote the growth of longer hairs, but was ceased after 6 months because of lack of effect.

Two 4-mm punch biopsies were taken from his occipital scalp for horizontal and vertical sectioning. On vertical section, the biopsy revealed a noncicatricial process with diffuse hair follicle miniaturization, an increase in catagen hairs and a peribulbar lymphocytic infiltrate (Fig. 3). Horizontal section showed global miniaturization (Fig. 4). Of 52 hairs counted, 24 were vellus-like hairs and 28 were terminal hairs. Among the terminal hairs there were several intermediate-sized hairs. Eleven hairs were in either catagen or telogen and many of those were also miniaturized.

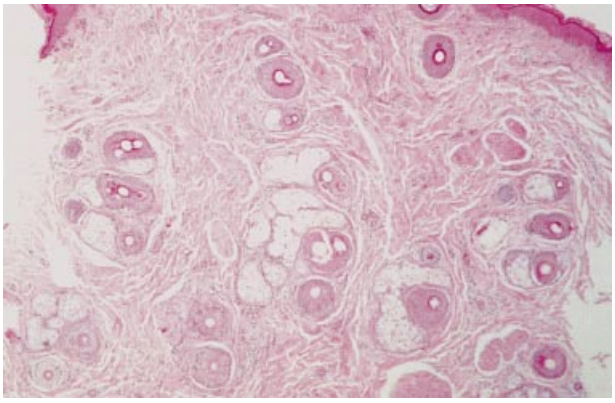

Figure 4 Histology of the occipital scalp in a horizontal cross-section showing normal follicular density, uniform miniaturization of the follicles and patchy perifollicular inflammation

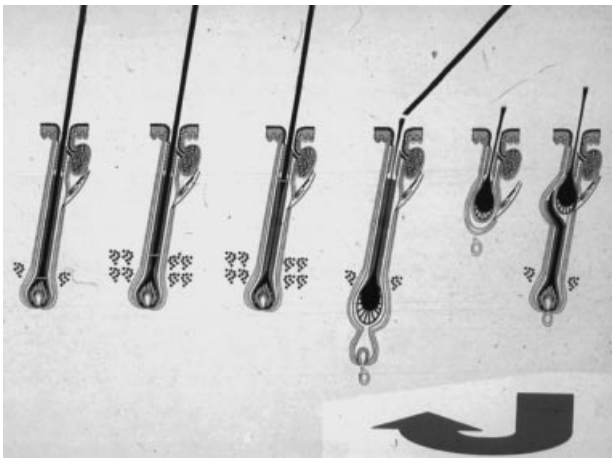

Figure 5 Pathogenesis of alopecia areata. The anagen hair follicles on the left are surrounded by inflammatory lymphocytes leading to the creation of the exclamation mark hair and, on the right, truncation of the hair cycle

Follow up over 4 years has not shown any change in his hair growth and he continues to have a fine down of depigmented hairs, uniformly 5 mm in length over his entire scalp that fail to grow any longer. He has not required a hair cut throughout this period. One attempt to color the hair with a permanent dye failed to take.

Discussion

The pathophysiology of alopecia areata involves an immunological arrest of anagen hair growth at the anagen IV stage of development. The inflammation precipitates onset of catagen, which is followed by telogen. The inflammatory infiltrate targets anagen hairs specifically and spares telogen hairs (Fig. 5). This suggests that the as yet to be discovered antigen target is selectively expressed in anagen IV, V and VI hairs. It has been postulated that hair pigmentation beginning at anagen IV, and in particular a melanocyte-related protein, is a target for inflammation.[1,2]

Telogen hairs re-enter anagen, and apparently normal growth occurs until anagen IV. At this stage, a resumption of inflammation terminates anagen growth.[3] Occasionally barely visible fine downy hairs 1–2 mm in length are seen on the scalp[4] but the clinical presentation of our patient, which resembles a haircut with a number 1 razor, has not been previously reported.

Possible explanations for our patient's hair growth in the presence of active alopecia areata include a delay in the onset of the immunological attack beyond anagen 4 or an unusually slow effect of the inflammation in triggering the onset of catagen. The latter would be analogous to the dystrophic anagen hairs with tapered fractures seen with the anagen effluvium patients receiving chemotherapy.[1] A delay in the onset of the immunological attack would suggest there is more than one antigen target in the hair follicle involved in the pathogenesis of alopecia areata.

Acknowledgments

This research was supported by a grant from the Scientific Research Fund of the Australasian College of Dermatologists and we would also like to thank Dr Jill Magee for interpreting the histology.

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