Disappearance of pili annulati following an episode of alopecia areata

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Summary

Pili annulati is a distinctive autosomal dominant hair shaft disorder that produces alternating light and dark bands that can give a spangled appearance to the hair. The literature contains three case reports of patients in whom the condition has disappeared following recovery from alopecia totalis. None of these reports contain a direct microscopic comparison of pre- and post-regrowth hairs. We report a 6-year-old girl who was first noted to have pili annulati at the age of 2 years and who developed alopecia totalis at the age of 3 years. When the hair regrew spontaneously, 18 months later, the pili annulati was no longer visible. Hair samples obtained before and after the episode of alopecia areata were compared by normal and cross-polarized light microscopy. While not apparent on careful clinical examination, banding was present on light microscopy in 20% of the hairs. Eighty per cent of the affected hairs displayed banding throughout their entire length. In contrast, prior to the episode of alopecia totalis, when the pili annulati was clearly visible, 50% of the hair obtained was banded on microscopy and 90% of the affected hairs showed banding throughout their microscopic length.

Report

Pili annulati (PA) is a rare disorder of the hair shaft that causes a banding pattern without affecting hair strength.[1] The pattern is more obvious in those with fair hair and also with age-related greying of the hair. The banding is caused by cavities located in the cortex of the hair shaft both within and in between cortical cells. These spaces reflect scattered light and are thus darker than normal sections of the hair shaft when viewed by transmitted light.[2] Inheritance is autosomal dominant but some cases are sporadic.

There are at least five previous case reports of alopecia areata and PA occurring in the same patient. Three describe a loss of banding after regrowth of the alopecia areata.

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We report a further case of coexpression of alopecia areata and PA with apparent disappearance of the PA following recovery from alopecia areata and, for the first time, provide direct comparison of pre- and post-regrowth hair. This demonstrates that the PA becomes subclinical, but does not disappear.

Our patient had striking banding of her hair noted from the age of 2 years. There was no associated hair fragility and she is otherwise well. At the age of 3 years, she developed patchy alopecia areata of the scalp that progressed to alopecia totalis over the next 6 months. There was associated total loss of eyebrows and near total loss of eyelashes, as well as nail pits. At the age of 5 years there was complete spontaneous regrowth of her hair, however, the PA was no longer visible. Follow-up at 6.5 years of age shows no recurrence of alopecia areata and no visible banding of the hair.

Scalp hairs obtained prior to the development of alopecia totalis were compared with hairs that regrew following recovery from the episode of alopecia areata. With the naked eye, banding is clearly visible in the pre-alopecia areata hair, but absent from the post-regrowth hair (Fig. 1).

Figure 1 Comparison of gross features of PA banding in pre-alopecia totalis hair (left) with post-alopecia totalis hair (right).

Approximately 20 hairs from each sample were fixed in Ultramount™ (Fronine Pty Ltd) and viewed with both normal and cross-polarized light. The number of hairs along the slide was counted. In accordance with previously described protocols[3] the features of PA were described in three categories according to: (i) presence or absence of PA; (ii) whether affected hairs had features of PA throughout their full length; (iii) whether the defect occupied the full or partial width of hairs. The results are shown in Table 1 and Fig. 2.

Eighteen members of the patient's extended family were examined and PA was diagnosed in nine of them. The pattern of inheritance in this family is clearly autosomal dominant. There is no family history of alopecia areata.

PA is a rare disorder of the hair shaft that is characterized by alternating light and dark bands. Hair strength is normal.[1,2]

Table 1 Features of PA in our patient according to: the percentage of affected hairs; features of PA throughout the full length of affected hairs; features of PV throughout the full width of affected hairs.

	Pili annulati (%)*	Complete length (%)†	Full width (%)‡
Pre alopecia areata	50	90	85
Post alopecia areata	30	80	50

*The percentage of hairs with PA bands visible by light microscopy. †The percentage of PA hairs affected by banding throughout their length. ‡The percentage of PA hairs with full width bands (defined as affecting ≥ 80% of the width of the hair viewed by light microscopy).

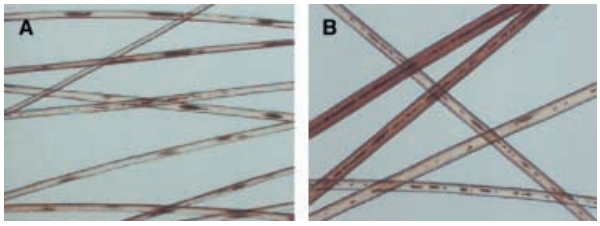

Figure 2 Comparison of microscopic features of PA banding pre- and post-alopecia totalis.(A) Pre-alopecia totalis: the PA banding is clearly visible. (B) Post-alopecia totalis: there is much less PA banding, however, the medulla in several hairs is much more prominent.

There are at least five previous reports of patients with PA developing alopecia areata. In one of these, banding remained visible following regrowth of the hair.[4] In one case it is unclear as no follow up is mentioned. In three cases the PA was no longer visible following regrowth.[5–7]

In addition to alopecia areata, PA has been associated with blue naevi[8] leukonykia[9] and melanoderma, syndactylia and polydactylism.[10]

Although it is a genetically autosomal condition the phenotypic expression of this disorder is extremely variable. In this family 60% of affected individuals are subclinical and the degree of banding depends on from where on the scalp the sample is taken and which part of the hair is sampled. The onset of the banding is also quite variable: some individuals displaying striking banding at birth with others first displaying grossly apparent banding after puberty or in adulthood.

This case demonstrates that an episode of alopecia areata has altered this patient's PA. Alopecia areata is a chronic relapsing autoimmune disorder of the hair follicle that causes anagen hairs to enter catagen prematurely. PA is a disorder of hair matrix function leading to cavity formation in the hair cortex that produces hair banding. The intermittent nature of the hair pathology associated with a lack of hair fragility possibly suggests that this might not be a disorder of a gene involved in making up the substance of the hair shaft, i.e. keratins or keratin associated proteins, but rather a gene related to control of hair growth dynamics. In contrast with PA, monilethrix, a disease of type II (basic) human hair keratin genes, has not been reported to be altered by alopecia areata. Given that alopecia areata leads to aberrant hair cycling, it is possible that the immune insult to the hair follicle could lead to a permanent change in the balance of controlling factors in hair shaft assembly, thus altering the frequency of hair shaft defects seen in PA. It is highly unlikely that the marked improvement in PA banding in this patient

was caused by the natural history of this condition. Although, in our experience, the banding does seem to lessen over a long period of time and tends to be milder in older affected adults, the rate at which the change occurred in this patient and the temporal relationship to an episode of alopecia totalis makes it much less plausible that this was merely a coincidence. Genetic studies will help unravel the reason why the pathology of alopecia areata, in this case and those previously reported, lead to lessening of the pathology in PA.

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