A Case of Pili Annulati Following Resolution of Alopecia Areata

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Abstract
Pili annulati (PA) is a primarily inherited condition that has been reported to occur concurrently with alopecia areata (AA) and following resolution of AA. PA represents a non-fragile hair disorder that becomes more evident in adulthood, allowing it to be missed in many patients. Microscopic examination of affected hairs reveals a characteristic banded, alternating dark-and-light appearance. There is no formal treatment for PA, but proper hair care can prevent harm from external factors.

Introduction
Pili annulati (PA) is a rare, non-fragile hair disorder, often inherited in an autosomal-dominant fashion, though there are reports of sporadic cases. PA clinically presents as light spangled hair. Microscopically, it is characterized by alternating dark and light bands initiating from the hair sheath that are present in a random fashion along the length of the upper three quarters of the hair shaft.1,2 The condition may be diagnosed in the beard, scalp, pubic and axillary regions.3-5 The alternating dark and light bands appear as such due to the presence of air-filled pockets within the cortex of the hair shaft at varying locations.6 Upon microscopic examination, these pockets scatter light and appear dark due to the decreased transmission of light; however, clinically they correspond to the light strands, because under reflective light the "microscopically dark" areas actually reflect more light.6 PA has been reported concomitantly in patients with alopecia areata (AA) and following the resolution of AA.

Case Report
A 6-year-old male presented to the dermatology office with the primary complaint of "hair not growing the way it used to." Hair loss had been occurring in a non-discrete pattern for two months' duration. The patient reportedly had very thick hair, and changes were particularly notable after a recent haircut prior to presenting to the office. The patient had a past medical history of hay fever. Past surgical and family histories were non-contributory.

On physical examination, the patient had round patches of non-scarring alopecia with mild scaling. A fungal culture was performed for suspected tinea capitis, and the patient was prescribed 2% ketoconazole topical lotion to be applied daily, pending cultures. Baseline laboratory examination revealed a mildly elevated white blood cell count, while the remainder of the exam was within normal limits. Fungal cultures did not reveal any growth, at which point ketoconazole therapy was discontinued and triamcinolone 0.1% cream with twice daily application was initiated for suspected alopecia areata. Further follow-up at two months revealed hair regrowth with only textural changes, and triamcinolone therapy was discontinued (Figure 1). A hair sample was sent for pathological examination to rule out a structural hair-shaft deformity. Examination of the hair shaft under polarized light revealed alternating dark and light bands with sharply cut edges indicative of a diagnosis of pili annulati (Figure 2).

Discussion
Hair abnormalities are subtle and therefore require extensive investigation. Pili annulati is a rare, autosomal-dominant hair shaft disorder, discovered in 1866, that has an unknown pathogenesis.1,3 Possible explanations for development of PA include a structural protein defect within the extra cellular matrix, a basement membrane zone defect, a cytokeratin abnormality and a genetic defect on the long arm of the 12th chromosome.12,9 Immunohistochemical examination for cytokeratin anomalies have not revealed any differences between PA and normal hair specimens.1 However, antibodies to aspects of the lamina densa, lamina lucida, and anchoring fibrils have been noted on immunohistochemical studies, which is supported by transmission electron microscopy of PA hair specimens that demonstrate reduplication of the lamina densa in the hair root bulb.8 Linkage analysis within families with autosomal-dominant PA has also been performed, revealing a gene locus on 12q with specific linkage between 12q24.32–24.33 regions.8,10 Although no known mutations have been isolated, having this specific locus provides an opportunity for further exploration.

The diagnosis of PA is often made by light microscopy, with increased sensitivity when a liquid medium is used to fix the hair.11 The differential diagnosis of PA includes pseudopili annulati and monilethrix.

Pseudopili annulati, a twisted-hair phenotype, characteristically appears in an elliptical shape with a normal hair shaft under light microscopy, but clinically it displays banding.12 Monilethrix is an inherited, autosomal-dominant condition that has an alternating node and constricted appearance on microscopy, resembling PA, but unlike PA it is a fragile hair disorder.13,14 PA has been documented to appear in conjunction with AA and post AA, and has also been reported to disappear after resolution of AA. One European study examined the presence of PA concomitant with AA and found a statistically significant 9% prevalence of concurrent AA in the PA group.15 The study, however, suggested that the concurrence was coincidental, as the genes identified in AA do not appear to have a relationship to the gene locus identified in PA.15 However, the study size was too small to determine an actual relationship. Our patient, on
the other hand, had PA after resolution of AA. A similar case has been reported by Cruz et al., in which a 31-year-old female with severe AA in locations of the scalp as well as other hair-bearing areas had resolution of her AA after being treated with several courses of intramuscular triamcinolone. Hair shaft examination of this patient’s regrown hair revealed PA. In contrast, a case has been reported of a patient with a past history of PA who presented with hair loss, and after treatment with intramuscular triamcinolone and topical minoxidil had re-growth of normal, non-PA hair.

PA has also been identified in a patient with a complex condition of autoimmune disease comprised of IgA deficiency, thyroid disease, and AA. Although there may be no association between PA and AA, the presence of autoimmune diseases may relay some connection to antigenic changes within the hair root bulb and the polygenic nature of AA as well as the distinct locus of PA.

Although there is no treatment for PA, patients with co-existing hair loss may benefit from topical minoxidil therapy, for it is thought to allow for normal matrix production, which in turn could theoretically repair the structural defect.

Conclusion

Pili annulati is a rare hair phenotype that has been documented to occur with AA, after resolution of AA, and in combination with other autoimmune disorders. Due to its inheritable nature, a family history of PA may warrant examination and education of patients affected with this condition. Although no treatment exists, patients should be reassured and taught proper hair care management for this non-fragile condition.

References


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