Acquired progressive kinking of the hair in an elderly woman: a case report

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Abstract

Acquired progressive kinking of the hair (APK) is a rare disorder that presents at any age and is infrequently presented in the literature. This diagnosis has been used to describe similar hair changes, though there are significant differences in presentation, population, and individual clinical features. Histologic analysis of hair shafts aids in diagnosis owing to commonly reported features such as irregular twisting, bending, and grooving of hair shafts with polygonal shapes on cross-section. We present a case with classic histologic features in an unusual patient to highlight this rare condition. Additional cases and studies are necessary to evaluate etiology, differentiate subsets of APK, and develop both treatment and prevention strategies.

Keywords: hair, acquired progressive kinking, wooly hair, pili torti, scalp

Introduction

Acquired progressive kinking of the hair (APK) is a rare hair morphologic disorder with only about 25 cases reported in the English literature [1]. This diagnosis has been used to describe similar hair changes, though there are significant differences in presentation, population, and individual clinical features [2, 3]. Many of the case reports are decades old and focus on phenotypic appearance and progression rather than on etiology and histologic analysis. Most commonly, patients present with coarse hair changes in an androgenetic pattern with sharp demarcation between normal and affected hairs, ultimately leading to alopecia [4]. We present a case of this rare condition in an atypical patient with classic histologic findings to support the diagnosis.

Case Synopsis

An 80-year-old female of Caucasian descent presented with several years of progressive undesired texture change of her posterior scalp. Beginning as a small patch, the coarse hair progressed to encompass the entire parieto-occipital scalp over an 8-year period by the time of presentation (**Figures 1, 2**). She denied any history of chemical treatment to her hair and reported no recent change in her hair care routine. Physical examination revealed a normal appearing scalp

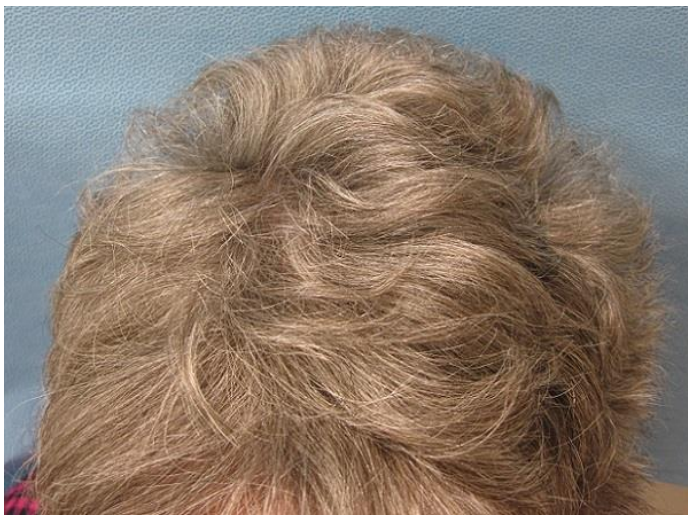

Figure 1. *Frontal, temporal, and parietal hairs retain smooth wavy appearance with luster.*

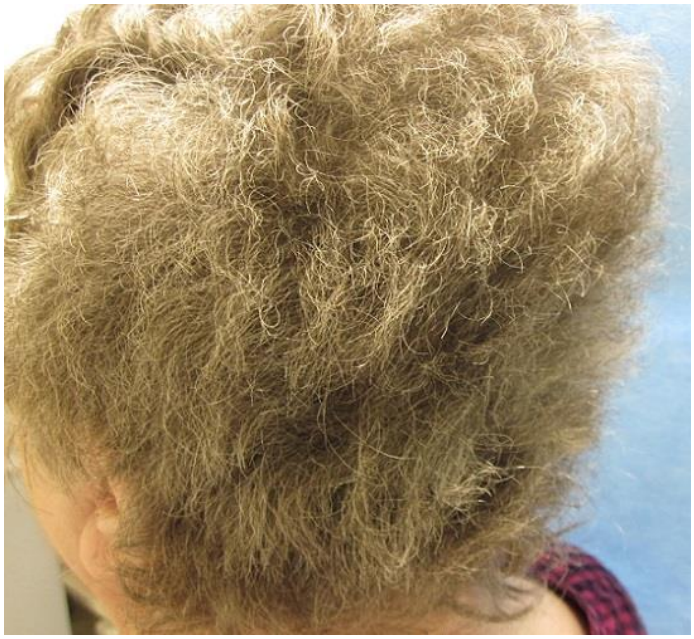

Figure 2. *Occipital hairs appear dull, unruly, and irregular.*

without overt inflammation or scarring. Biopsies from areas of both normal and kinky hair were taken and viewed under light microscopy. Horizontal cross-sections at the infundibular level were also examined with hematoxylin and eosin staining for histopathologic analysis. Examination revealed irregular twisting, bending, and grooving of the affected hairs as well as the presence of differing polygonal shapes of hair shafts in cross-section despite otherwise healthy appearing follicles (**Figures 3, 4**).

Case Discussion

Acquired progressive kinking of the hair has been described in the literature on rare occasions over the past several decades. The majority of patients are young men who present with fronto-temporal coarsening of the hair. These changes often precede androgenetic alopecia and although androgen involvement has been speculated, the precise pathway has not been clearly identified. Prior case reports of APK in an elderly woman have been difficult to locate, suggesting that this may be the first case report of its kind and may shed additional light in elucidating the pathophysiology. The presentation of APK in an elderly woman may also support the theory of androgenetic involvement related to known increasing of androgen/estrogen ratios in postmenopausal females.

Although some consensus regarding observable features of APK has been described, specific diagnostic criteria for APK have not been firmly established. This is likely related to rarity and variability in presentation between patients. Currently supported features include adult onset dull or lusterless woolly hair in a limited area, absence of previous trauma or hair chemical use, and irregular twists and kinks (pili canaliculi) of affected hairs [5-7]. Others suggest that diagnostic criteria include pigment changes and sharp demarcation between normal and affected hairs [8]; however, a case review by Tran highlights significant variability between cases [3]. Phenotypically, APK is similar to other conditions involving differences in scalp hair appearance and texture such as woolly hair nevus, Menkes syndrome, congenital pili torti syndrome, and uncombable hair syndrome [3, 9]. However, these conditions are present very early in life [3]. APK typically occurs later in life as a progressive change to hair texture and appearance over varying durations [2]. Currently, the oldest patient described is a 52-year-old woman [10].

In previous reports of APK, histology was reported as either unremarkable or more often not included. Interestingly, Sakamoto described a case that shares many features of APK, yet he reported a diagnosis of acquired pili torti-like hair defects. In Sakamoto's case, the cross-sectional appearance of hair shafts was reported as heart-shaped, triangular, or ellipsoidal [11]. In contrast, cross sectional

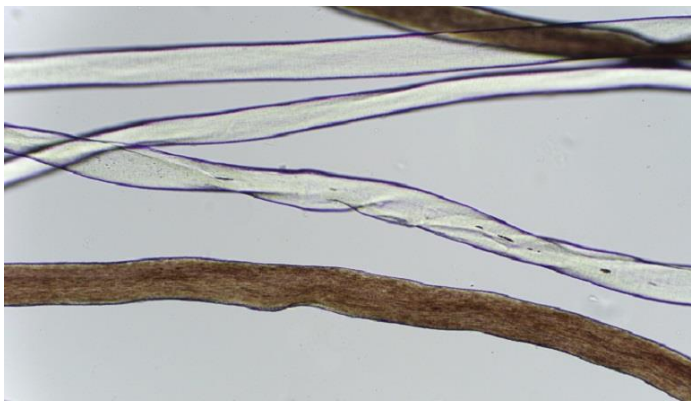

Figure 3. *Light microscopy showing irregular grooving, twisting, and bending of effected hair shafts, 100×.*

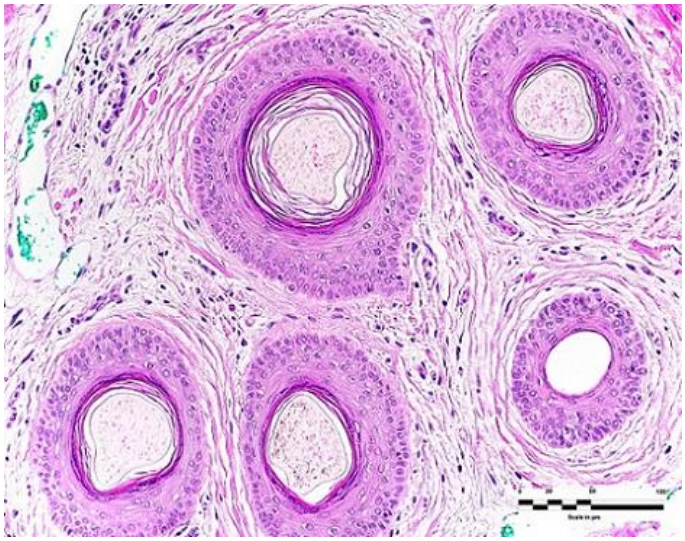

Figure 4. *Light microscopy showing a horizontal section at the infundibulum with irregular, polygonal, shaft contours. H&E, 200×.*

appearance of hairs from African-American individuals are more uniform and ellipsoidal with an eccentric position relative to the follicle [12]. Given similar findings in our case and that reported by Sakamoto, irregular bending and twisting of the hair shafts as well as diverse polygonal shaft contours on cross section should be included as diagnostic criteria for APK. Further exploration at multiple levels for unique histopathologic features such as pigment casts, concentricity, or cellular distribution in future cases could provide additional diagnostic criteria as well.

Conclusion

Microscopic findings described in our case are consistent with changes seen in previously described cases of APK. This condition most commonly presents in healthy adolescent and young adult individuals without an identified etiology, however, androgenetic involvement seems plausible. Histologic and light microscopic appearance of hairs in both longitudinal and cross section may help differentiate APK from phenotypically similar disorders like pili torti-like hair defects. Additional cases and investigations are needed to support these findings. Further examination at varying tissue levels with advanced techniques such as immunostaining or electron microscopy may also expose previously unknown factors in the pathogenesis of APK. We present a case of this rare disorder for the unique histological images that will contribute to the body of evidence that is lacking about APK.

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