

TWO CASES OF PROGRESSIVE ACQUIRED KINKING OF THE HAIR

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Summary

Progressive kinking of the hair is a rare acquired deformity of the hair shafts. It is believed to precede the development of androgenetic alopecia, especially in men. Besides, it has been observed in patients with chronic scalp dermatoses or taking certain medical preparations. We believe that its incidence is underestimated since not many patients seek medical advice because of the change in their hair structure. We present two cases of the disorder. Unfortunately little can be done in terms of treatment. Adequate styling agents and similar hair cosmetics can be of help.

Riassunto

La progressiva trasformazione in lanugine è una alterazione acquisita che colpisce la struttura esterna del capello.

Si pensa che preceda l'insorgere della alopecia androgenetica, soprattutto nell'uomo ed è stata anche osservata in pazienti affetti da dermatosi croniche del cuoio capelluto o che assumevano specifici medicinali.

Crediamo inoltre che questo tipo di incidenza sia sottostimata dato che molti pazienti non si giovano del consiglio medico quando avvengono modificazioni della struttura dei capelli.

Di seguito vengono presentati due casi di questo stato patologico.

Sfortunatamente scarse sono le possibilità di un intervento medico mentre alcuni prodotti cosmetici possono essere sicuramente di ausilio.

Acquired defects of terminal hair shaft structure in the majority of cases appear as the consequence of hair weathering and unfriendly influence of environmental factors. Damaged hair shafts are characterized by localized loss of cuticle cells, longitudinal splitting, increased fragility, loss of water, elasticity and shine. One or several of these features may be present at the same time. In light microscope the most commonly observed pathological changes in these cases are trichorrhexis nodosa, trichoklasia, trichoptilosis and trichonodosis, all of which are non-specific and can be caused by any exogenous factor. In the rare cases generalized nature of hair shafts abnormalities raise the possibility of systemic metabolism impairment.

The etiology of rare, acquired changes in hair texture and form is poorly understood. Scalp hair become coarse, twisted, loose shine and sometimes show increased pigmentation and resemble pubic hair macroscopically (1-4). Up to date, relatively few cases of progressive kinking of the hair have been described. Perhaps, this is due to the fact, that not many patients choose to see a doctor regarding this problem, some of them indeed prefer to consult their hairdresser. The other reason may be that this condition remains largely underdiagnosed.

Acquired progressive kinking of the hair usually begins after puberty and is more common in adults. At first, hair changes are localized, but slowly all scalp area may become involved. This condition is more common in males and sometimes it precedes androgenetic alopecia (3). The patients complain that their hair became curly instead of straight, resemble "wire" and style poorly. Light microscopic changes of the hair shafts include flattening and irregular partial twisting without the regular interval and constant twisting angle of 180° of the classical twisted hair (pili torti). Some authors with the use of scanning electron microscopy demonstrated the presence of longitudinal grooves resembling

those of pili trianguli et canaliculi in uncombable hair syndrome (2). Besides, in some patients with progressive kinking of the hair morphological changes of hair shafts may include pili torti and moniletrix.

The clinical and light microscopic findings in two new cases of acquired progressive kinking of the hair are presented.

Case 1. 23-year-old male presented with intensive pruritus and scaling of the scalp. During the previous two years he also noted patchy changes in his hair, distributed in a linear fashion in the frontal and vertex areas of the scalp. The affected hairs became curly and after each combing stood out as "cocks-comb", which along with the dandruff was the major concern of the patient (fig.1).



Fig. 1 Patient with progressive acquired kinking of the hair. Patchy linear changes.

Upon examination, diffuse erythema and thick scales on the scalp were consistent with the clinical diagnosis of psoriasis. Furthermore, there was marked pitting of the fingernails on both hands. Light microscopic investigation of the patient's hair revealed partial twists, occasional flattening of hair shafts and areas of irregular hair diameter, resembling morphological changes in moniletrix (fig 2-4).

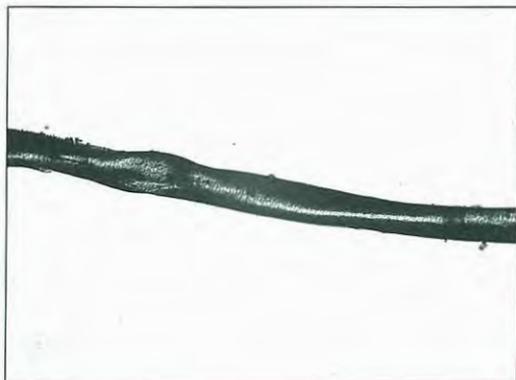


Fig. 2 Irregular twisting of the hair shaft.

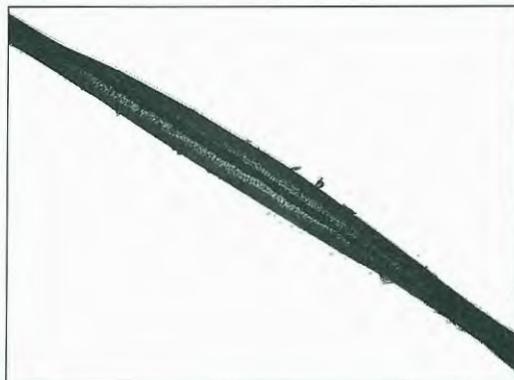


Fig. 3 Hair shaft abnormality resembling that of moniletrix.

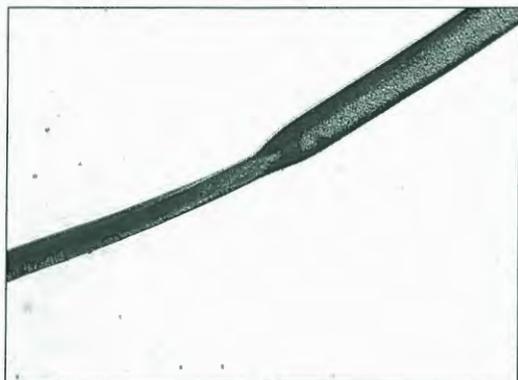


Fig. 4 Uneven diameter of the hair shaft.

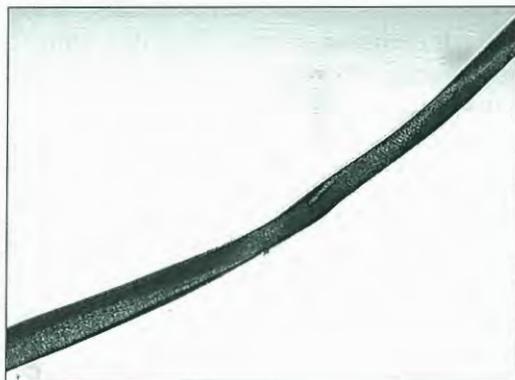


Fig. 5 Irregular partial twisting and uneven diameter of the hair shafts.

Case 2. 19-year-old female presented with complaints that during the period of one year her previously straight hair became tightly curly and in spite of her efforts to straighten them with hot blow-dryer the hair still looked very untidy like “wire”. Clinically the changes in hair texture were diffuse, but more pronounced in fronto-parietal area. Further questioning of the patient revealed that she had been long treated for the ovarian dysfunction and had laboratory confirmed hyperandrogenemia. Light microscopic investigation of the patient’s hair demonstrated abnormalities similar to those of the previously described patient.

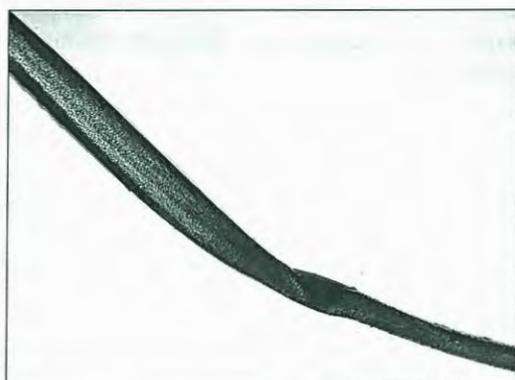


Fig. 6 Irregular partial twisting and uneven diameter of the hair shafts.

As already mentioned above, etiology and pathogenesis of this condition is unknown. The currently predominant point of view among trichologists is that progressive acquired kinking of the hair is an androgen-dependent process. Indeed, in many reported cases it progressed to androgenetic alopecia (3). However, in our particular patients only long-term observation will confirm or rule out this tendency. Abnormalities of ovarian function and predominance of androgens in our female patient makes progression to androgenetic alopecia a high probability. As for the male patient, chronic inflammatory process in his scalp skin does not exclude the possibility of development of male pattern baldness in the future. It is unclear, to what extent does chronic inflammation affects the structure of hair shafts in this patient.

In various reports etiological factors have been implicated, such as drugs, chemical, mechanical and thermal damage to the scalp (1-5). Progressive kinking of the hair has been reported in patients with seborrhoeic dermatitis as well as in patients with psoriasis undergoing treatment with retinoids (6). In latter case the authors consider retinoids to be the cause of hair alterations, rather than chronic inflammatory process in the skin. It is known that acquired pili torti occur in undernourished patients with anorexia nervosa taking high doses of vitamin A preparations (7). It's worth mentioning, that there are no strict criteria and definitions of this rare condition, therefore various authors differ in their classification of morphological hair changes that they observe in their patients. Thus, some compare their findings with wooly hair, others – with uncombable hair and a Menkes kinky hair syndrome (2,4).

The above-mentioned hair diseases are genetically determined and are usually diagnosed in early childhood. Therefore, careful anamnesis makes the differential diagnosis easy. However, inherited hair shaft abnormalities can be used as

prototypes for comparison.

Menkes kinky hair syndrome is a serious and potentially fatal X-linked recessive systemic disease, caused by copper deficiency due to its impaired intestinal absorption. Scalp hairs appears normal in newborns, but later are substituted by steel-colored, hypopigmented, short and curly hairs, that break easily. Hypotrichosis can develop. Morphological abnormalities of the hair shafts include pili torti, trichorrhexis nodosa and rarely - moniletrix. Interestingly, healthy female carriers of the pathological gene have similar structural changes of their scalp hair (8). Besides Menkes syndrome, pili torti in association with systemic abnormalities can be seen in a number of neurodegenerative diseases, so called neurotrichoses (9).

The uncombable hair syndrome (pili trianguli et canaliculi) is a rare autosomal-dominant condition. Structural abnormalities of the hair shaft make styling of the hair impossible and are caused by defect in keratinization process and pathological configuration of inner root sheath (triangular or kidney-shaped at cross-section). The changes become noticeable in children at about age 3 or later. The hair is light, quite thick, without any change of growth dynamics. In light microscope the hair shafts may appear normal. Investigation with the use of scanning or transmission electron microscope is required to demonstrate longitudinal shaft grooves or triangular or kidney-shaped form at cross-section (10).

Twisting of the hair also occurs in wooly hair, both diffuse and nevoid. Wooly hair nevus is usually found in association with epidermal nevi in patients with epithelial nevus syndrome (11,12).

Thus, it is reasonable to say that there are a lot of morphological similarities between certain hereditary and acquired abnormalities in hair shaft structure, at least as can be perceived at light microscopic examination of affected hairs.

However, it is of importance that such genetically determined parameters as hair-shaft form and thickness can be affected by a number of factors. Among those, hormonal factors predominate. It is a well-known fact that the earliest symptom of androgen-dependent alopecia is hair thinning due to hair follicles' miniaturization. Oral contraceptive drugs cause straightening of previously curly hair in some women, and a more common phenomenon is straightening of curly hair in women during pregnancy [personal observations]. On the other hand, progressive acquired kinking of the hair may precede the development of androgenetic alopecia. More cases with follow-up are required to make judgment regarding this seemingly heterogeneous group of patients.

However, for both of our patients aesthetic considerations that prompted them to see a doctor were of major importance. No treatment is currently available; though we may hypothesize that topical minoxidil could be of help. In organ culture of mouse hair follicles minoxidil prevented kinking of the hair shafts as well as other abnormalities observed in hairs cultured without minoxidil (13).

We feel that the best recommendations for hair care in such cases is use of high-quality protective styling agents specially indicated for hair straightening and avoidance of vigorous manipulations with hair and hot comb. We also always recommend oral vitamins and minerals supplements to support hair growth.

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