A 24-year-old male Caucasian presented to our outpatient clinic for multiple nevi on his trunk. His nevi were examined by dermoscopy and those suspected to be dysplastic were totally excised for histopathologic examination. On clinical examination, the patient was observed to have blonde, unruly and unmanageable hair. His mother stated that his hair had always had that texture since first months of his life and that his hair not only seemed to grow slowly, but also was difficult to comb. He was born to a consanguineous couple. Family history did not reveal similar hair findings. On dermatologic examination, the entire scalp was affected with yellowish, dry, frizzy hair that projected outward (Fig. 1a and b). Neither alopecia nor hypotrichosis was present and no evidence of scalp abnormalities was detected. Eyebrows, eyelashes and body hair were not involved. Mucosae, teeth, nails and sweating of the patient were normal. Systemic and laboratory examinations were also within the normal limits. The hair pull test demonstrated that hairs could not be easily extracted. Light microscopic examination of the hairs revealed pathognomonic characteristics of UHS, canal-like longitudinal...
depressions and on trichoscopy a longitudinal grooving were also detected (Figs. 2 and 3). To confirm the diagnosis, hair samples were also investigated by SEM and shallow grooving was found along the entire length of the hairs examined (Fig. 4). Further work-up demonstrated bilateral juvenile cataract on biomicroscopic examination (Fig. 5).

Uncombable hair syndrome or pili trianguli et canaliculi is a dysmorphic hair disorder of unknown origin. It can be a sporadic or an inherited hair disorder, isolated finding without associated motor or cognitive developmental abnormalities. However, rarely, retinal dysplasia/pigmentary dystrophy, juvenile cataract, ectodermal dysplasia, digit abnormalities, tooth enamel anomalies, oligodontia, and phalangeopiphysseal dysplasia have been reported. Schena et al. report a case of UHS and neurofibromatosis associated with mental retardation, single palmar crease and arched palate (7). Bork et al. defined a new entity of ectodermal syndrome consisting of “congenital hypotrichosis with uncombable hair, juvenile cataract, retinal pigmentary dystrophy, oligodontia and brachymetacarpia” in a family (10). Our case was investigated in terms of these abnormalities and was found to have bilateral cataract.

Although most disorders of hair shaft can readily be visualized and diagnosed by light microscopy, UHS requires SEM evidence for diagnosis. Scanning electron microscopy is the gold standard, which reveals the distinct feature of shallow grooving along the entire length of the hair in at least 50% of the hairs examined.

Trichoscopy is a noninvasive procedure allowing for rapid and magnified (X10) in vivo observation of the skin with visualization of morphological features not visible to the naked eye (3). It was initially used to assess pigmented lesions. Recent reports have shown that trichoscopy is a helpful, easy and rapid way in the diagnosis of many hair disorders. It has been used for rapid diagnosis of some hair shaft disorders such as monilethrix, trichorrhexis nodosa, uncombable hair, and also other hair diseases such as alopecia areata, androgenetic alopecia, traction alopecia and tinea capitis (4, 6, 11).

On admission, the patient was aware of having a hair shaft abnormality that had been overlooked for years and he had not been investigated in this respect. On detailed examination, he was also found to have bilateral cataract as a component of UHS and as a re-

**Figure 2.** The appearance of longitudinal depression on light microscopy.

**Figure 3.** The appearance of longitudinal grooving on trichoscopy.

the latter with an autosomal dominant or recessive pattern and variable levels of penetrance (7). Scalp hairs are usually silver or blond, wiry, frizzy, rough, and described as unmanageable. The hair may grow normally or at a slower rate. Hair cannot be combed flat. It can present from infancy to 12 years of age, although it occurs typically between the ages of 3 months and 3 years. It usually resolves spontaneously by adolescence (8, 9). Uncombable hair syndrome is usually an
As a result of early diagnosis, it will be followed by ophthalmology periodically. After suspecting, the diagnosis was easily based on the characteristic features seen by light microscopic examination and SEM. Microscopic examination requires numerous hair samples to be removed. Therefore, using trichoscopy, which is an easier way, the diagnosis was made by demonstrating hairs with longitudinal grooving. Although the recommended working magnification is 20- to 160-fold (12), in our case, analysis of the low-magnified dermoscopic image of the hairs is presented.

Trichoscopy may provide an alternative to standard light microscopy and to electron microscopy for examination of hairs and to diagnose hair shaft anomalies as well as to rule out the possibility of other associated abnormalities, so it should be used widely.

References

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