HAIR DISORDERS

SAME FAMILY, SAME DISEASE, BUT TWO DIFFERENT PRESENTATIONS IN MONILETHRIX

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Background: Monilethrix is a rare autosomal dominant disease of the hair shaft with high penetrance but variable expression. It is characterized by alopecia as a result of hair breakage. A moniliform aspect of the hair develops within the first months of life, hence the denomination “monilethrix”. Perifollicular erythema and follicular hyperkeratosis are also common. In extensive cases, there may be eyebrow, eyelash, and nail involvement. Herein, we report a case of two sisters that highlights inter-individual variability of clinical features of this rare alopecia in the same family.

Observation: Two sisters aged 18 and 16 year-old presented to our dermatology department for a history of congenital thin and short hair. They were born from a consanguineous marriage. Their hair was normal at birth then suffered from breakage after few months. Their brother had also the same symptoms. Dermatologic examination revealed mild hypotrichosis of the scalp with sparse, short, and fragile hair that seemed to be broken in the elder girl. However, severe hypotrichosis with extensive alopecia was observed in her sister. Small keratotic papules were also noted on their occipital region. There were no eyebrows nor eyelashes nor nail abnormalities. Dermoscopy of the scalp showed regular variations in the diameter of the hair shaft with elliptical dilations (nodes) and constrictions (internodes) for both of them. Congenital hair shaft disorder, in particular monilethrix, was diagnosed. Topical minoxidil 5% was prescribed but poor improvement after three months was reported in the two sisters.

Key message: This case is being reported not only for the rarity of the disease but also to highlight the inter-individual variability of this rare genodermatosis easily diagnosed using dermoscopy.