Co-occurrence of Monilethrix and Type 1 Diabetes Mellitus

Sir,

A 14-year-old girl child, a known case of type I diabetes mellitus (DM), presented with complains of sparse hairs over scalp since birth that gradually became denser with age. Her younger brother had similar complains with no history of type 1 DM in any of the other family members. On examination, the patient had sparse lusterless hairs, which was more prominent over the frontal, temporal, and parietal regions of the scalp [Figure 1]. Sparse axillary and pubic hairs were present. Multiple follicular papular lesions were present all over the scalp. Dermoscopic examination (Dino-Lite premier AM3013T nonpolarized, Contact type; ×50 magnification, connected to a computer with inbuilt camera for taking photographs) showed few hairs with uniform spindle-shaped nodes and intermittent constrictions (internodes) [Figure 2a]. Few hairs showed translucent areas at regular intervals [Figure 2b]. Fractured hairs were seen. Patient was diagnosed to have monilethrix with type I DM, and was started on topical minoxidil 5% and multivitamins, following which the patient showed little improvement.

The word monilethrix is derived from Latin "monile" meaning "necklace" and the Greek "thrix" meaning "hair." The first case of monilethrix was described by Walter Smith in 1879 but the term "monilethrix" was coined by Radcliff

Crocker.^[1] Several genetic studies have suggested that monilethrix is likely caused by a hair keratin mutation. The most common mutation is the *E413K* mutation in *hHb6*. Autosomal dominant monilethrix is caused by mutations in hair keratin genes *KRT81*, *KRT83*, or *KRT86*, whereas in autosomal recessive form, mutation in the desmoglein-4 gene (*DSG4*) has been reported.^[2]

Monilethrix occurs mainly on the scalp with eyelashes, evebrows, axillary, pubic, and limb hairs occasionally affected. Lanugo hairs are normal in the neonatal period. When terminal hairs start appearing, nodes begin to form along the hair shafts at regular interval of 0.5-1 mm. Unlike our case, the short and stubble brittle hairs are more prominent on the occipital region and the nape of the neck and are seen emerging from the top of the horny follicular papules. Apart from short, sparse, fragile, nongrowing hairs, affected patients may have keratosis pilaris, koilonychia, and rarely, systemic disturbances such as mental and physical retardation, syndactyly, cataract, teeth, and nail anomalies. ^[3] No association has been found between monilethrix and type 1 DM even after extensive literature search. The basic pathophysiology behind this association may be involvement of common genes that could be inherited with incomplete penetrance; therefore, further clinical studies would be needed for confirming definite association.



Figure 1: Lustreless hairs with follicular papules over the scalp

Dermoscopy shows hair shafts with uniform spindle-shaped nodes and intermittent constrictions (internodes); hairs bent at multiple locations and a tendency of shaft to fracture at the sites of constrictions. The nodes seem to represent normal growth; the internodes are characterized by the wrinkling of cortical cells leading to fragility of hair with an absence of medulla.

The course and outcome of this condition is very variable and unpredictable. It may persist throughout life, but spontaneous partial or complete remission can occur with age. Various activities causing hair trauma such as dyeing, bleaching, and curling should be avoided. Improvement has been reported with griseofulvin, iron supplementation, oral retinoids, and topical minoxidil in isolated cases.^[4]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.



Figure 2: (a) Dermoscopy showing uniform spindle-shaped nodes and intermittent constrictions (internodes) of hair shaft (\times 50). (b) Dermoscopy showing hairs with translucent areas along the hair shafts at regular intervals (\times 500)

Conflicts of interest

There are no conflicts of interest.

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