Monilethrix: A Report of Three Cases in Children Confirmed with Dermoscopy

Abstract

Monilethrix is a rare hereditary disorder affecting hair resulting in hair fragility and alopecia. We report three patients of monilethrix who presented with complaints of sparse and brittle hair from early childhood. All three patients had multiple discrete hyperkeratotic papules over the scalp. Dermoscopy revealed beaded appearance of hair with the presence of elliptical nodes and intermittent constrictions on the hair shafts as well as broken hairs, which were confirmed with routine microscopic examination of hair. Dermoscopy helps in easier and faster diagnosis of monilethrix.

Keywords: *Beaded hair, dermoscopy, monilethrix*

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Introduction

Monilethrix is a rare hereditary disorder affecting hair resulting in hair shaft anomaly. Although it is an autosomal dominant disorder with variable expression, few cases of autosomal recessive inheritance have also been reported in the literature.[1] It is characterized by short, brittle, and sparse hair, usually over the scalp, and at times involving eyebrows, axillary, and pubic hair. Follicular keratosis of the affected scalp and keratosis pilaris are also typical.

Case Reports

Case 1

A 3-year-old girl born out of second-degree consanguineous marriage was brought by her mother with complaints of sparse and fragile hair since 1 year of age. History of repeated tonsuring was present once every 6 months in anticipation of stronger and thicker hair growth afterward, with the last tonsuring 2 months prior to the hospital visit. Examination revealed short, rough, lusterless, and sparse hair, easily breakable over the scalp and eyebrows with multiple discrete keratotic papules all over the scalp [Figure 1a]. Dermoscopic examination (HEINE DELTA20®, 10× magnification) under nonpolarized contact dermoscopy over vertex, frontal, bitemporal and occipital scalp showed regular variations in the diameter of the hair shaft with elliptical nodes separated by internodes, broken hair [Figure 1b (blue arrow)], and areas with very much thinned out internodes on the verge of breakage (red arrow). Dermoscopy of keratotic papules over the occipital scalp revealed broken and beaded hair emerging from the surface of papules [Figure 2a]. Routine microscopic examination of hair from the scalp revealed regularly beaded hairs with constriction and fraying at the internodal junction suggesting monilethrix [Figure 2b].

Case 2

A 9-year-old girl born out of second-degree consanguineous marriage presented with complaints of sparse hair over the scalp and eyebrows. She had multiple hyperkeratotic papules localized to the occipital scalp [Figure 3a]. Dermoscopy of multiple sites over the scalp revealed regularly arranged nodes and internodes with broken hairs [Figure 3b].

Case 3

The third patient was a 13-year-old girl who attained secondary sexual characteristics 1 year before, presenting with similar complaints. On examination, hair was short and sparse over the scalp and eyebrows and almost absent over the axilla and pubic area with keratotic papules over the occipital scalp [Figure 4a-c]. Microscopic and dermoscopic findings were similar to the

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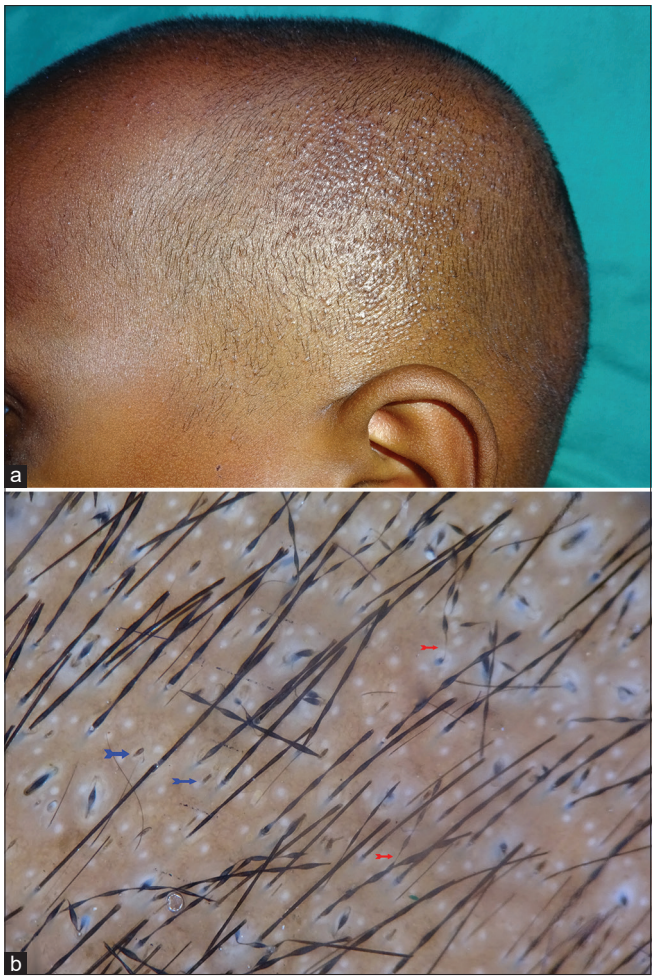

Figure 1: (a) Short and sparse hairs with multiple discrete hyperkeratotic papules over the temporal area of the scalp. (b) Dermoscopy (HEINE DELTA20® nonpolarized contact dermoscope, 10 × magnification) showing regular variations in the diameter of the hair shafts with elliptical nodes separated by internodes, broken hairs (blue arrows), and areas with very much thinned out internodes on the verge of breakage (red arrows)

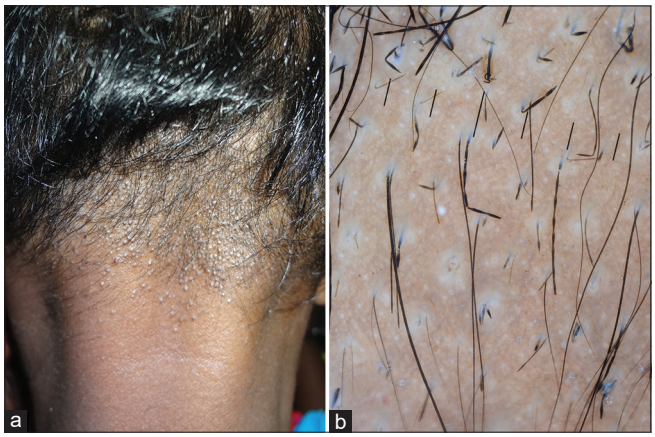

Figure 3: (a) Multiple keratotic papules localized over the occipital scalp with sparse hair. (b) Dermoscopy demonstrating regularly arranged nodes and internodes with broken hairs

first two patients [Figures 4d and 5]. Biopsy was not done in any of the three cases.

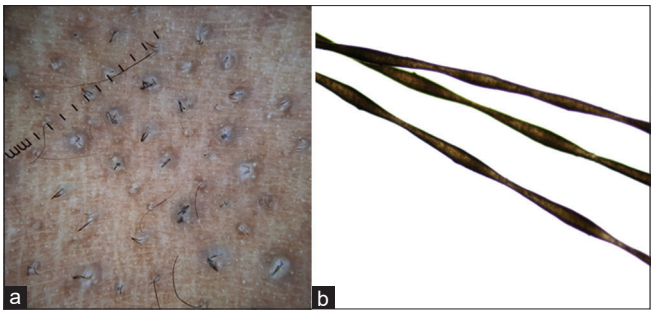

Figure 2: (a) Dermoscopy of keratotic papules showing broken and beaded hair emerging from the surface of papules. (b) Routine microscopic examination of scalp hair showing regularly beaded hairs with constriction at the internodal junction (40×)

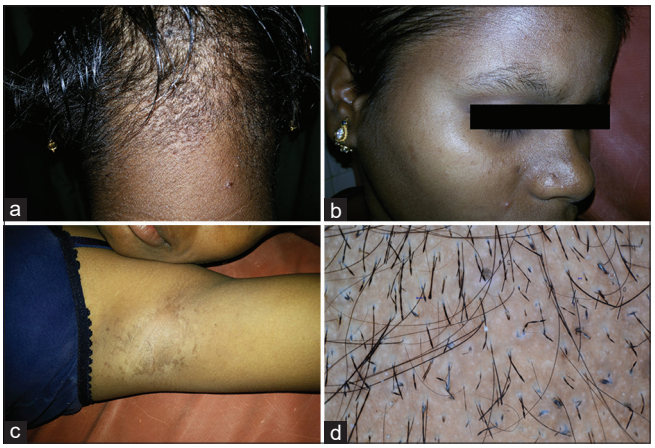

Figure 4: (a) Keratotic papules over the occipital scalp along with sparse hair. (b) Sparse hairs over the eyebrows. (c) Almost absent hair over the axilla. (d) Dermoscopy demonstrating regularly arranged nodes and internodes, broken hairs, and exclamation hair-like hairs (blue arrows)

Examination of skin, teeth and nails was normal for all three patients. There was no evidence of any other associated systemic abnormality. Parents were counseled about the course and prognosis of the condition. The second and third patients were started on topical minoxidil and systemic acitretin after necessary investigations. Patients are currently under follow-up without any significant improvement.

Discussion

Monilethrix was first described by Smith in 1879, and the term "monilethrix" was coined by Radcliff Crocker from Latin word "monile" meaning "necklace" and Greek word "thrix" meaning "hair." It is caused by mutations in genes coding for hHb1 and hHb6, the human basic hair keratins, in which a highly conserved glutamic acid residue Glu413 is substituted by lysine or aspartic acid residue. The autosomal recessive forms have been reported to have mutations in desmoglein 4.[2]

Affected children have normal lanugo hair on birth. Symptoms begin when terminal hair characteristics begin to develop.[3] The hair is dull and fragile and breaks easily, especially over sites of friction such as the nape and

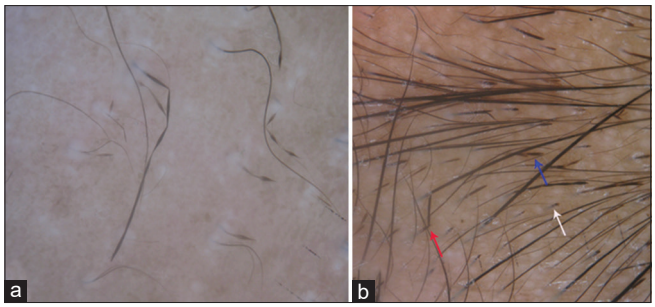

Figure 5: (a) Dermoscopy of axilla showing sparse axillary hair with alternate nodes and internodes, broken hair, and bend hairs. (b) Dermoscopy of eyebrows showing broken (white arrow), bend (red arrow), and split hair (blue arrow) in addition to nodes and internodes

occipital areas. Monilethrix is characterized by beaded appearance of hair due to the presence of elliptical nodes and intermittent constrictions on the hair shafts, resulting in hair fragility at those points. The elliptical nodes have the diameter of normal hair and are medullated with regular number and structure of cortical cells. These are regularly separated by internodes that are narrow and are devoid of medulla with lower number and wrinkling of cortical cells making them the site of fracture.[1,4] The thin internodes are considered to be the result of diminished anagen hair growth in a 2-day rhythm.[4]

Various follicular abnormalities are seen in association with monilethrix, ranging from perifollicular erythema and hyperkeratosis to horny follicular papule formation. It can also be associated with koilonychia, syndactyly, cataract, blepheritis, pannus, and dental abnormalities.[1]

In addition to beaded appearance, dermoscopic examination may reveal hair shafts bent regularly at multiple locations, with a tendency to curve in different directions. These findings have been described as "rosary bead appearance" or "regularly bended ribbon sign" by Tosti[5] and Rakowska *et al*.[6] Dermoscopy helps in easier and faster diagnosis of monilethrix and is more patient-friendly without any plucking and cutting, as well as rules out iatrogenic pseudomonilethrix, which may result in wrong diagnosis during the microscopic examination.[7] In addition, it is a helpful tool in examining siblings or parents with no obvious anomalies where diagnosis can be missed when the number of hairs with characteristic appearance are limited.

Management of monilethrix is still a challenge for dermatologists. Avoiding trauma to the hair is the most effective step in the management of this hereditary condition.[8] Spontaneous improvement has been noticed in adolescence and pregnancy suggesting a hormonal influence in the pathogenesis and course of this condition.[9] Various modalities such as topical minoxidil, systemic acitretin, griseofulvin, and N-acetylcysteine have been tried with reports of some cosmetic improvement.[8] Another promising option is oral minoxidil which was tried in two patients with significant improvement in hair density and growth and reduced hair shedding.[10] However, no permanent cure has been reported.

Conclusion

Monilethrix is a rare condition with characteristic findings. The diagnosis can be proven non-invasively using dermoscopy, which is easier and faster than the conventional light microscopic examination of hair.

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

Conflicts of interest

There are no conflicts of interest.

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