

Images/Instrument in Dermatology/Dermatosurgery

## Monilethrix: Beaded hair and hypotrichosis in a child

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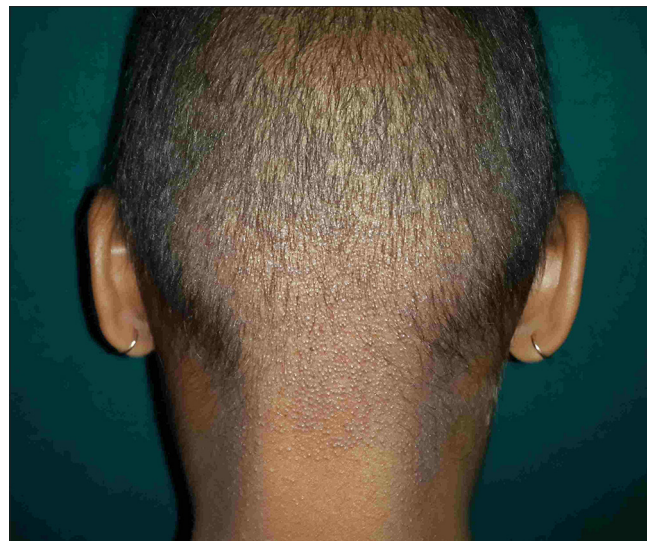
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A 9-year-old girl, with a history of scalp hair thinning and breakage since early childhood, presented with diffuse scalp hypotrichosis accentuated at the occiput. Examination revealed short, sparse, lustreless hair with follicular keratotic papules at the nape [Figure 1]. Light microscopy of plucked hair shafts demonstrated regularly spaced fusiform nodes separated by narrow, atrophic internodes—findings characteristic of monilethrix<sup>[1]</sup> [Figure 2]. This rare hereditary hair shaft disorder is typically autosomal dominant, due to mutations in type-II hair keratin genes *KRT81*, *KRT83*, or *KRT86* on chromosome-12. The internodes lack a medulla and are prone to fracture.<sup>[1]</sup>

Although dermoscopy classically shows a “rosary-bead” or “regularly bent ribbon” appearance, in this case, light microscopy was already available and clearly demonstrative. It should be differentiated from pseudomonilethrix, a debated entity showing irregular shaft indentations rather than the uniform beading of monilethrix, lacking the typical clinical picture, and generally attributed to optical artifacts from sampling, cosmetic products, or mechanical trauma rather than genetically determined true periodic shaft narrowing.<sup>[2]</sup>

Management is supportive, focusing on minimizing hair trauma. Topical minoxidil, oral acitretin, and biotin have been tried with inconsistent benefit.<sup>[3]</sup>



**Figure 1:** Hypotrichosis accentuated at the occipital scalp with dry, sparse, short hair and follicular keratotic papules at the nape.

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**Figure 2:** Light microscopy showing beaded hair shafts with fusiform nodes and narrow internodes.

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