

Beaded necklace hair: diagnosing monilethrix in a pediatric patient

Cabelo em colar de contas: diagnosticando monilethrix em um paciente pediátrico

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Keypoints

What is known

- Monilethrix is an inherited genodermatosis of unknown incidence, given its rarity and limited documentation.

What is added

- Due to its infrequency, it is crucial to review and share such cases, aiming to ensure diagnostic clarity and early management.

A nine-year-old female patient, with no significant medical or surgical history, was referred for limited hair growth and easy breakage since birth, resulting in permanently sparse and short hair. There was no significant family history.

On physical assessment, decreased hair density with intense hypotrichosis and small hyperkeratotic papules were observed. The hair was only a few centimeters long, with a dry and lusterless appearance (Figs. 1 and 2).

The tug test was positive, indicating hair fragility, while the pull test was negative, showing no active hair shedding. No abnormalities were observed in the nails or teeth. Trichoscopy revealed narrow areas of constriction periodically separated by elliptical nodules on the hair shaft, creating an appearance similar to a “beaded necklace/rosary beads” (Fig. 3). Based on the clinical and dermoscopic findings, the diagnosis of monilethrix was established. Topical 5% minoxidil was prescribed, to be applied twice daily.

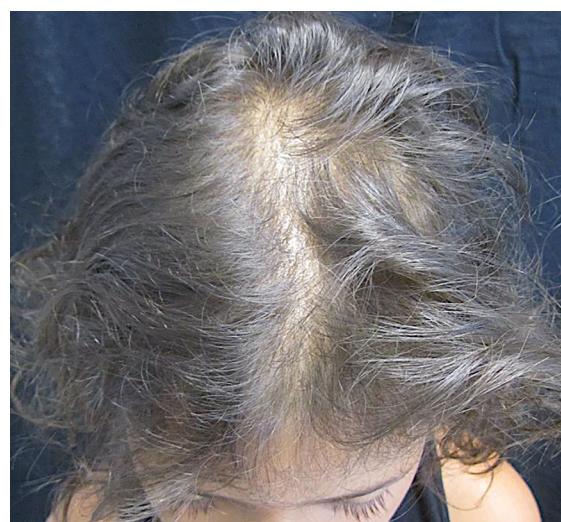


Figure 1. The scalp of a nine-year-old girl exhibits diffuse hypotrichosis with sporadic coarseness, with no impact on the eyebrows and eyelashes.

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Figure 2. Hyperkeratotic follicular papules.

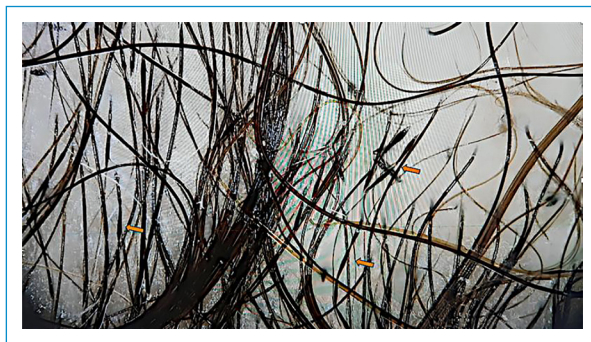


Figure 3. Trichoscopic image of the parietal scalp, showing uniform elliptical nodes, interspersed by intermittent constrictions, a pathognomonic sign of monilethrix.

Discussion

Monilethrix is a rare genodermatosis of autosomal dominant inheritance¹ that primarily affects the hair shaft and typically presents during early childhood. It is characterized by dysplasia of the hair shaft, leading to progressive hair follicle thinning and the development of fragile, short, and lackluster hair. The clinical expression of monilethrix exhibits significant variability² and can ultimately result in hypotrichosis due to hair fragility^{2,3}.

Diagnosis can be easily established by assessing the patient's clinical history and conducting trichoscopy, which can reveal intermittent constrictions along the hair shaft, resulting in a distinctive rosary bead-like appearance³.

As of now, there is no established successful treatment for this hair condition⁴. However, topical or low-dose oral minoxidil (0.25-2.5 mg/day)⁵ has shown promise as a well-tolerated and potentially effective treatment option⁵.

Author contributions

S. Santos do Vale: Conception and design of the study, report, review or other type of work or paper; Acquisition of data either from patients, research studies, or literature; Drafting the article. L. Santos Silva: Conception and design of the study, report, review or other type of work or paper; Critical review of the article for important intellectual content; Final approval of the version to be published; Agreement to be accountable for the accuracy or integrity of the work.

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Conflicts of interest

None.

Ethical considerations

Protection of humans and animals. The authors declare that no experiments involving humans or animals were conducted for this research.

Confidentiality, informed consent, and ethical approval. The authors have followed their institution's confidentiality protocols, obtained informed consent from patients, and received approval from the Ethics Committee. The SAGER guidelines were followed according to the nature of the study.

Declaration on the use of artificial intelligence. The authors declare that no generative artificial intelligence was used in the writing of this manuscript.

References

1. Winter H, Rogers MA, Langbein L, Stevens HP, Leigh IM, Labrèze C, et al. Mutations in the hair cortex keratin hHb6 cause the inherited hair disease monilethrix. *Nat Genet.* 1997;16(4):372-4.
2. Bindurani S, Rajiv S. Monilethrix with variable expressivity. *Int J Trichology.* 2013;5(1):53-5.
3. Baltazard T, Dhaille F, Chaby G, Lok C. Value of dermoscopy for the diagnosis of monilethrix. *Dermatol Online J.* 2017;23(7):13030/qt9h-f1p3xm.
4. Randolph M, Tosti A. Oral minoxidil treatment for hair loss: A review of efficacy and safety. *J Am Acad Dermatol.* 2021;84(3):737-746.
5. Darchini-Maragheh E, Moussa A, Wall D, Meah N, Sinclair R. Low-dose oral minoxidil for the treatment of monilethrix: A retrospective review. *J Eur Acad Dermatol Venereol.* 2024 Sep 18. doi: 10.1111/jdv.20329. Epub ahead of print.