Dermatology Practical & Conceptual

Sparse Brittle Hair and Bilateral Temporal Alopecia in a Child

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Case Presentation

A healthy 2-year-old boy, the second child of nonconsanguineous parents, presented with sparse, brittle hair and alopecia affecting the bilateral temporal and parietal scalp since he was 2 months old (Figure 1). Both parents had normal hair; however, his 9-year-old sister experienced a similar condition that appeared to resolve spontaneously. Dermoscopy revealed regular constrictions in the hair shafts, giving a beaded appearance. Hair microscopy showed beaded nodes at regular intervals with a reduced central dark medulla in the constricted areas. A diagnosis of monilethrix was made.

Teaching Point

Monilethrix is a rare hair shaft disorder, typically inherited in an autosomal dominant pattern. It is caused by mutations in genes encoding hair shaft proteins (*KRT81*, *KRT83*, *KRT86*, and desmoglein), leading to abnormal medulla formation, which results in hair thinning, fragility, and breakage [1]. Clinically, monilethrix is characterized by brittle, fragile hair and often sparse hair density, accompanied by patchy alopecia with variable distribution. The clinical presentation is remarkably polymorphic, even among members of the same family carrying the same genetic mutation. While some individuals may exhibit only fine, sparse hair, others may develop extensive alopecia from an early age. Additional features such as follicular keratosis may be observed, particularly on the scalp, neck, and extremities. The severity of the condition can vary over time, often being more pronounced during childhood and improving with age. In many cases, symptoms tend to improve or even resolve spontaneously in adulthood. There is no definitive treatment; however, supportive measures and avoiding trauma to the hair may help reduce breakage.

Figure 1. Sparse, brittle hair and patchy alopecia in a 2-year-old boy with monilethrix, showing involvement of the temporal and parietal scalp.

Reference

1. Ortner-Tobider D, Trafoier T, Moosbrugger-Martinz V, Tollinger S, Gruber R, Schmuth M. Keratin variants in monilethrix. *Br J Dermatol*. 2024 Nov 18;191(6):863-864. DOI: 10.1093/bjd/ljae340. PMID: 39192774.