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Monilethrix

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Abstract

Monilethrix is a genetic disorder of hair shaft structure characterized by increased fragility. We report a 6 year old boy with monilethrix in view of rarity of this disorder.

Introduction

Monilethrix is a rare autosomal dominant disorder of hair. There is an alternating widening and narrowing of the hair shaft. The narrow portions of the hair shaft are prone to fracture easily resulting in varying degrees of alopecia.

Case report

A 6 years old boy born of non-consanguineous marriage presented to the outpatient department with poor growth of hair starting after a few months of normal hair growth in infancy. His parents noticed that the hair did not grow beyond a certain length. There was no history of this disorder in any of the family members.

His mental and physical growth was normal. The scalp showed diffuse hair loss with scattered short stubby hair of few centimeters in length. There were prominent horny follicular papules over the vertex and nape of the neck with hair emerging from the summit giving the scalp a rough feel **(Fig.1)**. His eyebrows and eyelashes were sparse and short **(Fig.2)**. Hair was absent on the rest of the body and multiple skin colored horny papules were seen on the upper back. The light microscopic examination of the hair shaft revealed the pathogonomic finding of alternating node (widening) and internode (narrowing) confirming the diagnosis of monilethrix.

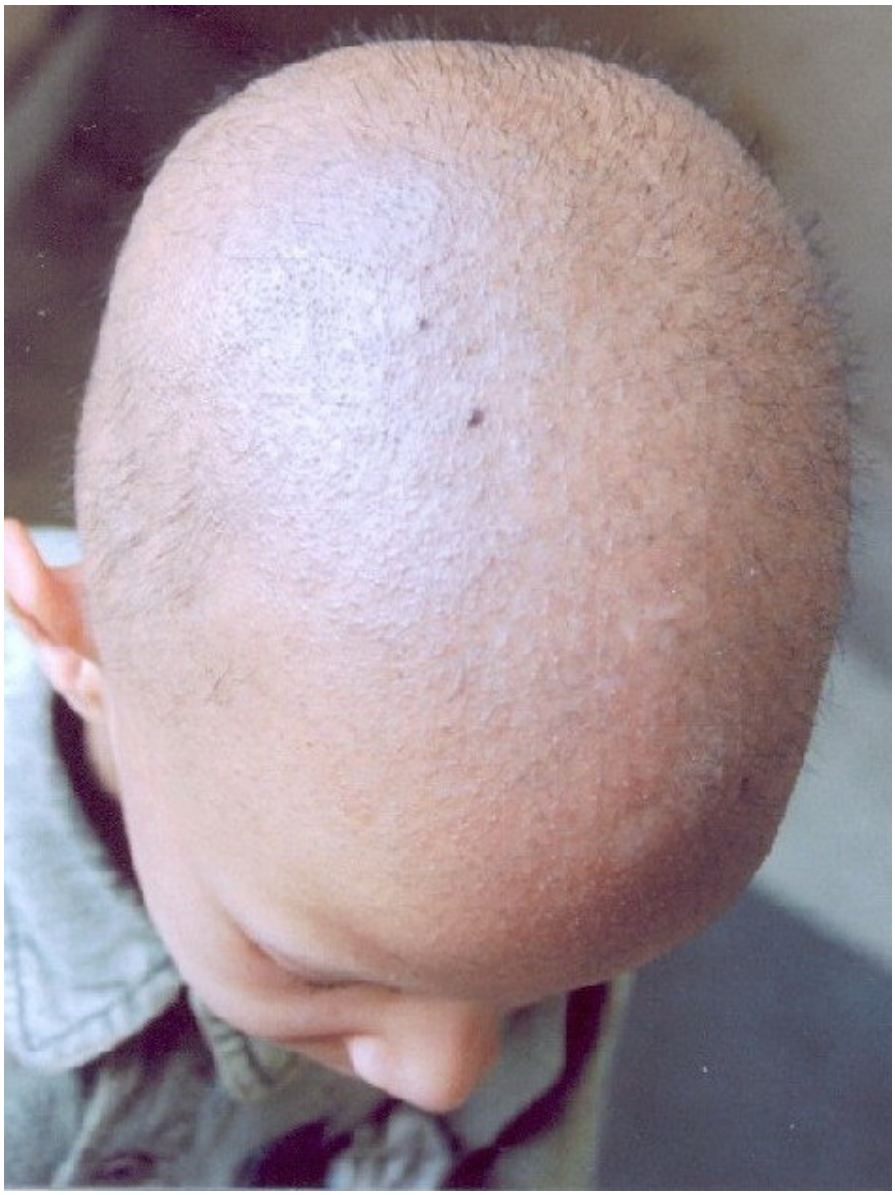

Fig 1: Diffuse alopecia with scattered short stubby hair and prominent horny follicular papules over the vertex of the scalp.

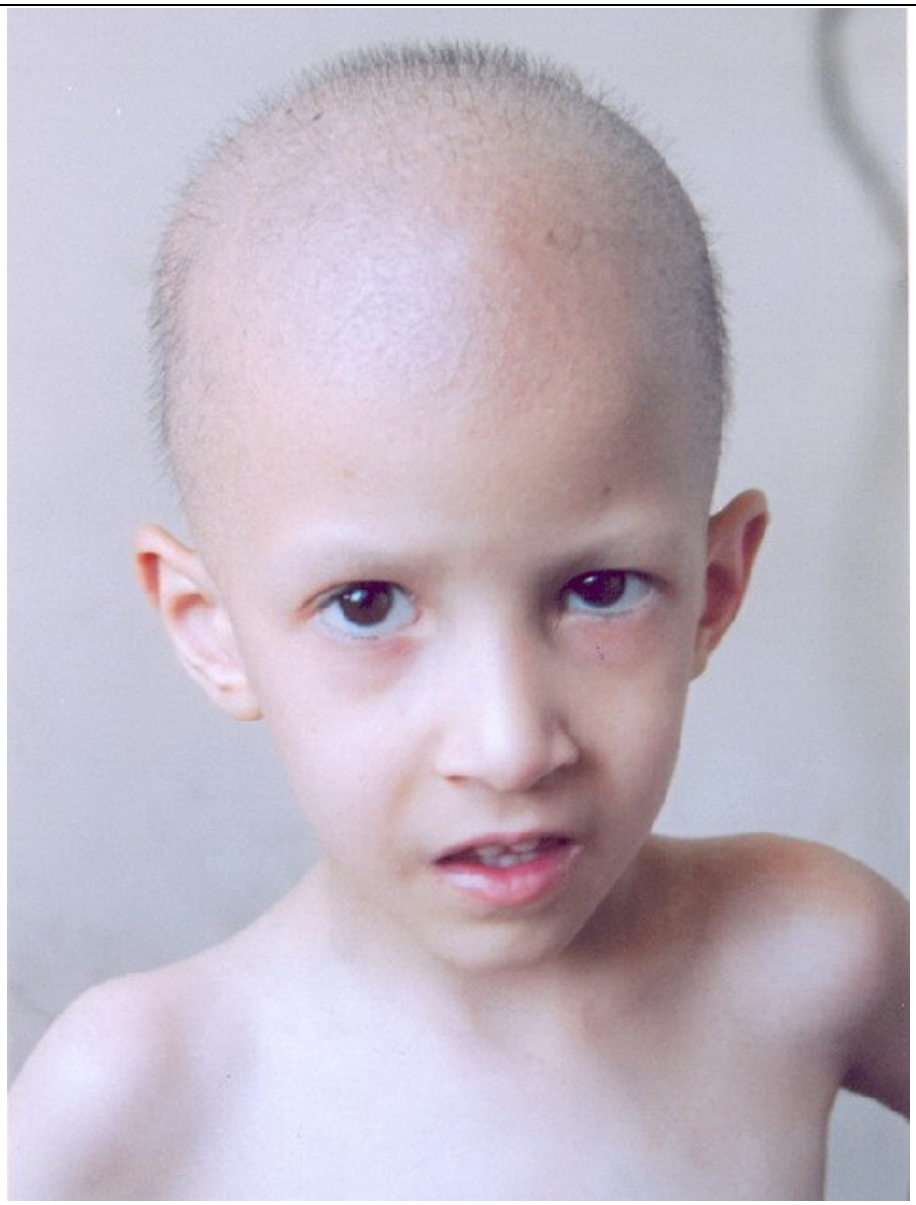

Fig 2: Short and sparse eyebrows and eyelashes.

Discussion

This genetic disorder of hair shaft structure with increased fragility was first described by Walter Smith in 1879 as "a rare nodose condition of the hair" [1]. Radcliffe Crocker suggested the term monilethrix which is derived from the Greek meaning necklace hair.

This developmental abnormality of the hair shaft is inherited in an autosomal dominant manner with high penetrance and variable expressivity. The hair growth is normal at birth and is replaced by short brittle hair during the first few months of life. The short stubble brittle hairs are more prominent on the occiput and the nape of the neck and are seen emerging from the top of the horny follicular papules. The eyebrows, eyelashes, axillary and pubic hair and the hair on the rest of the body may also be affected. The susceptibility to premature weathering results in its inability to attain

normal length. The condition has been known to persist throughout life but in some cases it may resolve spontaneously [2,3].

The light microscopic examination reveals alternating wider elliptical nodes and narrower internodes. The premature weathering and breakage is seen at the fragile internodes. Transmission microscopy of the internode showed deviation in the axis of the macrofibrils within the cortical cells and disorganized globular inter-macrofibrillar cystine rich material [4]. The uneven transverse distribution of this matrix protein is thought to contribute for the asymmetrical tension and subsequent fragility. The internodes occur at the site of a greater cortical defect due to the effect of compression by the inner root sheath during keratinization process and is seen as invagination of the cuticle into the weakened cortex [1]. The regular elliptical beading of the hair shaft occurs as a secondary feature due to the abnormal intermittent activity of the basal cells producing the cortical protein.

Other associated abnormalities seen with monilethrix include trichorrhexis nodosa, juvenile cataracts, syndactyl, nail and teeth abnormalities.

There is no specific treatment for this condition, although some improvement has been seen with oral retinoids, griseofulvin, iron supplementation and topical minoxidil [5,6,7,8].

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