MONILETHRIX

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Monilethrix in a family running for three generations is being presented

Key words : Beaded hair, Fragility, Alopecia, Monilethrix.

Introduction

Monilethrix is a rare hereditary condition with variable expressivity characterised by the presence of beaded or spindle shaped shaft of the scalp hair. Hair shaft shows beaded appearance due to alternate zones of spindle like thickening and thinning placed about 0.7to 1mm apart. Swelling represents normal part of the hair, whereas narrowing part is abnormal. It is usually inherited as an autosomal dominant trait and rarely can be autosomal recessive.[1] It shows considerable variation in age of onset, severity and course. Hair is usually normal at birth and progressively replaced by abnormal hair during the first few months of life. Hair loss is due to hair fragility and break occurs at internodal junction. All over the scalp mostly on occiput and nape of the neck, keratotic follicular papules may be seen, from where broken hair stumps are observed. In some patients, eyebrows, eye lashes, pubic hair, axillary hair and general body hair may be affected. This condition may improve in adulthood but usually persists with little change throughout life. It is rarely associated with mental retardation, metabolic disorders, tooth and nail defects, cataract, and ichthyosis.

Case Report

A male patient aged 7 years was brought with the complaint of deficient hair growth over the scalp since birth.

Examination showed that the scalp hair was sparse, short and dry. The hair follicles were prominent with keratotic papules and broken hair stumps were observed in many follicles. The scalp felt gritty on touch, and keratosis pilaris extended to the neck and back (fig.1).

History and examination of patient's father revealed that he also had the same prob-

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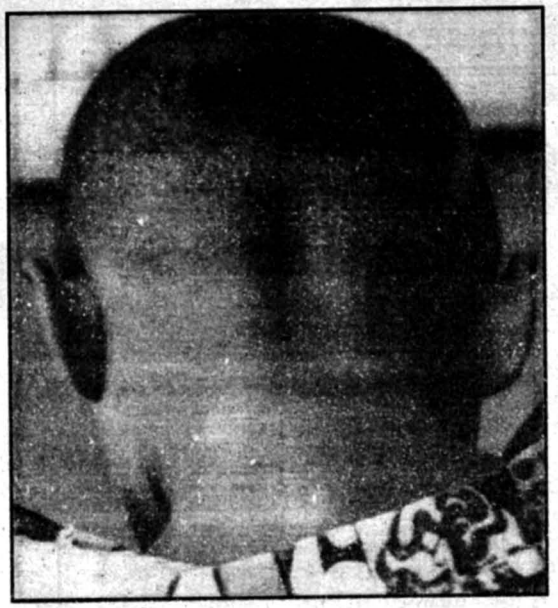

Fig.1. Monilethrix

lem, since childhood. Hair growth improved with advancement of age and he had prominent hair follicles over most of the area of the skin. Examination of a hair under light microscope showed bulbous thickening in many regions. Thorough family examination revealed that the patient's younger brother and paternal grandmother were also affected ; elder brother, mother and paternal aunt were not affected.

Discussion

Monilethrix is a rare condition and reported only once in this journal before.[2] Solomon reported twenty-eight cases in seven generations of one family.[3] Pattern of disease affliction in this family shows the autosomal dominant inheritance running in three generations. The exact cause for the narrowing is not known. Recent report denotes that internodal constrictions are abnormal usually without medulla.[4] Cortex and cuticle are defective.[5] In addition, abnormality of keratinizing zone of follicles also has been suggested.[6]

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Announcement

'Dr Bishnupriya Debi Award' for the best original article published in IJDVL for the years 1997 and 1998 will be awarded during the National Conference of IADVL to be held at Bangalore in January 2000.

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