MONILETHRIX

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ABSTRACT

Monilethrix is a rare condition characterised by beading of hairs. It is a structural defect of hair with increased fragility, inherited as an autosomal dominant condition. It shows a considerable variation in the age of onset, severity and course. Various treatments have been tried with varying results.

We are presenting a case report of 18 years old female patient with monilethrix. She complained of diffuse hair fall from the whole of scalp from last 10 years. Multiple papules over the nape of neck, back and extremities were present. Microscopy and histopathology confirmed the diagnosis. The patient was put on iron and conditioning shampoo.

KEYWORDS

Monilethrix, Autosomal Dominant, Fragility, Conditioning.

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INTRODUCTION

Smith initially called this condition 'a rare nodose condition of the hair.[1] Radcliffe Crocker, subsequently suggested the term monilethrix. Nevertheless, some early reports and even some more recent ones, confuse monilethrix with other shaft defects (e.g. Trichorrhexis Nodosa) when 'weathering' is severe.

Autosomal dominant transmission has been demonstrated in numerous large pedigrees.[2] The alleged occurrence of normal carriers of the dominant gene has not been proven, as a parent with only 5% of abnormal follicles is easily passed as normal.[3] The abnormality is attributed to mutations in the genes coding for the human hair keratins hHb1 and hHb6.[4] There is no clear correlation between the severity of the phenotype and the mutation responsible.[5] A variant of monilethrix with scalp erosions and the appearance of congenital alopecia is attributed to a mutation in the gene for desmoglein 4, which can be recessively inherited.[6]

The diagnosis depends on the typical history, clinical features, histological and microbiological examination. There is no definitive treatment for this condition, though many modalities have been tried.

CASE DESCRIPTION

An 18 years old female student, came with complaints of diffuse hair fall from scalp and difficulty in combing from last 10 yrs. The onset was gradual and progressive in nature. On examination, scalp showed varying degree of alopecia more prominent in occipital and fronto-parietal areas. Hairs were dry, lustreless, fragile, black in colour and of varying length. No seborrhoea was appreciated on the scalp surfaces. Hair pull test was positive. Multiple, well-defined, small-sized keratotic papules over the nape of neck, back and extensors of extremities were present. Nails showed koilonychia and were dry, lustreless. Mucosa was normal.

Hair specimens were sent for histological and microbiological examination. Histologically, the follicle shows wide and narrow zones corresponding to the nodes and internodes. Microscopic examination of hair revealed the characteristic beaded hair. Patient was explained the nature, course and prognosis of the disease. There is no definite treatment for this condition. The patient was put on:

1. Tab Biotin BD.
2. Tab Iron BD.
3. Conditioning shampoo for L/A OD.

The above mentioned treatment was given for 2 months.

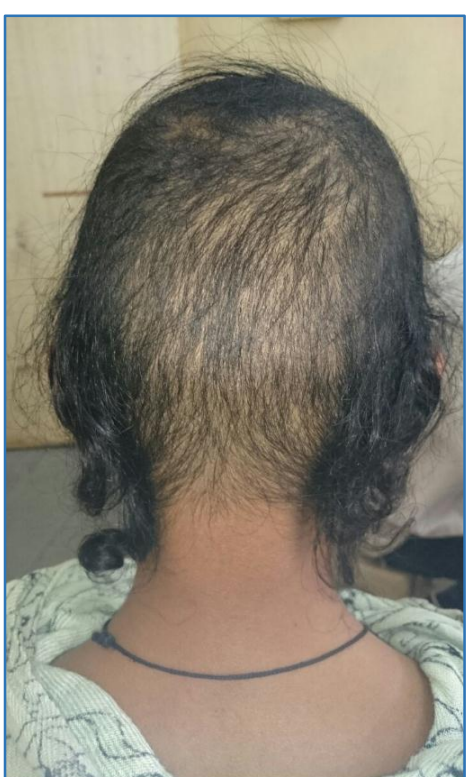

FIGURE 1:DIFFUSE ALOPECIA IN OCCIPITAL REGION AND HYPERKERATOTIC PAPULES ON NAPE OF NECK

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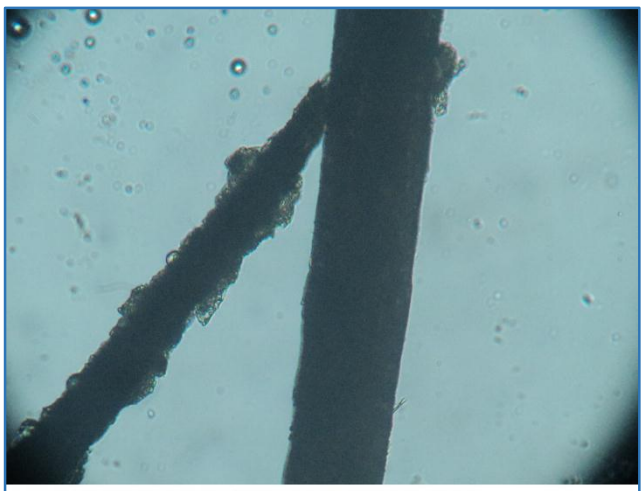

FIGURE 2:MICROBIOLOGICAL EXAMINATION SHOWING BEADING OF HAIR

FIGURE 3:MICROBIOLOGICAL EXAMINATION SHOWING BEADING.

DISCUSSION

Monilethrix is a rare autosomal dominant condition. It is a structural defect of the hair characterised by increased fragility and beaded hair. The hair shaft is beaded and breaks easily. Elliptical nodes 0.7–1 mm apart are separated by narrow internodes with a form resembling the body and neck of a skittle. The widths of the nodes and the distances between them vary between the hairs of an individual and between members of the same family. The nodes and some of the internodes show a normal imbricated scale pattern, but most internodes show longitudinal ridging.[7,8] True monilethrix must be distinguished from pseudo-monilethrix, an artefact produced by tweezers or compressing overlapping hairs between two glass slides. Histologically, the follicle shows wide and narrow zones corresponding to the nodes and internodes.[9] Attempts have been made to investigate the mechanism of node formation and to relate it to the diurnal rate of hair growth, but with no overall conclusion.[7,10,11] Intermittent administration of an antimitotic agent can give rise to zones of constriction alternating with zones of normal diameter.[12]

Monilethrix shows considerable variation in age of onset, severity and course.[13] Hair loss or broken hair is accompanied by follicular keratoses most commonly on the nape and occiput. In some cases, the eyebrows and eye lashes, pubic and axillary hair and general body hair may be affected. In many patients, the condition persists with little change throughout life.[2] Spontaneous improvement or complete recovery has occurred.[1] and has been reported during pregnancy.[14]

Three reports support the use of oral retinoids, providing a reversible benefit.[15–17] Improvement has been attributed to the hormone changes of menarche in one report.[18]; male and female cases may both improve at puberty. Griseofulvin, also has temporarily restored normal hair growth.[19] Iron supplementation has been reported to be of value in the presence of iron deficiency.[20] Reduction of hairdressing trauma may be followed by some improvement, by lessening the 'weathering' from chemical and physical insults.

CONCLUSION

Diagnosis of monilethrix depends upon clinical and microbiological examination. There is no definitive treatment for the condition. This case is being presented for the rarity and the characteristic clinical presentation.

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