

MONILETHRIX IN PEDIGREE

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A 30-year-old female patient presented with tiny papular scalp lesions with sparse, very short, curly hair of only 8 to 12 mm. Microscopic examination of hair revealed typical beaded or moniliform appearance. She also had typical lesions of psoriasis for past 2 months. Her father, 2 sisters and her 6-year-old daughter had similar types of beaded scalp hair.

Key Words : Monilethrix, Pedigree

Introduction

Monilethrix is a rare familial disorder of hair shaft of unknown cause.¹ It was first described by Smith in 1879.² It is an autosomal dominant disorder, occurring in either sex. Rarely, it is autosomal recessive.

Monilethrix is usually confined to the scalp. It is characterised by localised or widespread dystrophy of hair shafts and hair follicles which causes patchy or diffuse hair loss. The hair shafts show regular variations in diameter producing alternate "nodes" and "internodes" which confer a striking beaded or moniliform appearance on the hair. At the internodal portion, the medulla is diminished or absent and the hair breaks easily at this point. Therefore, hair fails to grow longer than 5 to 10 mm. These changes are usually associated with a follicular hyperkeratosis or keratosis pilaris. Under light microscope, hairs show typical beaded or moniliform appearance and elliptical nodes are 0.7 to 1 mm apart separated by narrow internodes.

Case Report

A 30-year-old female patient presented with diffuse hair loss and tiny papular lesions on the scalp since childhood. She had also

erythematous scaly plaques on the scalp, trunk, both upper and lower extremities for past 2 months. The patient's 2 sisters and her father had history of similar lesions on the scalp, trunk, both upper and lower extremities for past 2 months. The patient's 2 sisters and her father had history of similar lesions on the scalp since childhood. The patient's 2 daughters had similar complaints starting at the age of 3 years.

On examination, the scalp revealed sparse hair with hyperkeratotic tiny papular lesions. 8-12 mm sized hairs were projecting from the hyperkeratotic tiny papules. The hairs were easily breakable or fragile. Hairs were examined under light microscope. Typical beaded or moniliform appearance of hair was seen under light microscope. Her other hair bearing areas like eyebrows, eyelashes, public, axillary hairs and other body hairs were normal. She had erythematous scaly plaques of psoriasis on her scalp, trunk and on both extremities.

Her two daughters, two sisters and her father had similar types of keratotic papules with short fragile hair involving the scalp. Typical beading of hairs was seen under light microscope. We differentiated monilethrix from other hair shaft deformities by its classical beaded or moniliform appearance.

Discussion

Monilethrix is a rare hair shaft

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deformity. Its pedigree presentation is suggestive of hereditary transmission. One case of monilethrix with pedigree was reported by Solomon and Green.³ Many therapies like oral steroids, retinoids, immunosuppressive drugs are suggested.³ Avoidance of trauma may be helpful. In present case, we have not tried any systemic therapy as patient was not co-operative. Systemic steroids were avoided due to associated psoriatic lesions.

References

1. Deraemaeker R. Monilethrix : report of a family with special reference to some problems. *Am J Hum Genet* 1957; 9: 195-201.
2. Smith WG. A rare nodose condition of the hair. *BMJ* 1879; 11: 291-6.
3. Solomon IL, Green OC. Monilethrix. *N Engl J Med* 1963; 269: 1279-85.