A Case of Monilethrix Treated with Etretinate

A.	Sivasundram
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Department of Dermatology, General Hospital, Klang, Malaysia

Dr. A. Sivasundram, Department of Dermatology, General Hospital, Klang, 412000 (Malaysia)

Monilethrix usually appears early in life with dryness, fragility and sparse hair growth. It occurs mainly on the occiput and nape of the neck, but it can affect the entire scalp. This inherited entity may be associated with physical retardation, cataract, nail and tooth abnormalities and extensive keratosis pilaris. The course of the disease is variable. Remissions during puberty, pregnancy and summer may occur. Several treatments have been used to alleviate the condition but with little success. Only scarce information exists on the effect of oral retinoids.

A 9-year-old otherwise healthy Indian boy presented with a life-long history of generalized sparse scalp hair. Three other siblings are normal. There is no family history of hair loss.

The scalp hair revealed broken off beaded hair with a length of 2-5 cm associated with follicular hyperkeratosis. Microscopic examination showed regularly spaced fusiform swellings in the hair shaft separated by narrow segments. There was extensive keratosis pilaris involving the trunk, upper and lower extremities.

The patient was treated with 0.5 mg/kg/day of etretinate. After about 6 months there was hair growth, especially of the vertex and temporal regions, but it was sparse in the occipital region. The keratosis pilaris completely regressed. The patient was given etretinate for a further period of 1 year, but there was no further significant improvement.

The only side-effect of the drug is dryness and cracking of the lips which ceased after a period of time. The therapy was stopped at this stage to see whether hair growth would be maintained after cessation of therapy. The

patient returned 2 months later complaining of loss of hair and relapse of the keratosis pilaris. He was restarted on etretinate, and soon there was hair growth with loss of beading as before. The overall cosmetic appearance was fair. The patient is still on a maintenance dose of 25 mg etretinate daily.

Oral retinoids have been used with success in the treatment of disorders of keratinization. The exact mechanisms of their mode of action is not known. It is also not known for how long the treatment should be continued.

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