MONILETHRIX

(Report of two cases in a family)

By

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Monilethrix (beaded hair) is a rare hereditary disorder which was first described by W. G. Smith in 1879 as "A rare nodose condition of the hair". In this condition, the hair shaft shows nodes or swellings with intervening constructions, thus hairs have beaded appearance. It is thought that node or bulbous part of the affected hair is normal and the thin internode segment is deficient in hair matrix (Low, 1910 and Solomon & Green, 1963). The hair usually breaks off at the constriction when it is a few millimeters long (Solomon and-Green, 1963). Usually it fails to grow longer than 5-10 mm (Baker, 1962) and rarely exceeds 2-5 cms. in length (Summarly & Donaldson, 1962). The disease is probably transmitted as simple autosomal dominant without sex limitation (Butterworth & Lyon, 1962).

The condition usually appears after the normal first hair of the infant had fallen about the sixth week of life, but cases have been recorded in which it began in youth and even in adult life. Sabauraud found seventeen cases in five generations of one family, McCall Anderson reported fourteen cases in six generations (Savill & Warren, 1962). Dr. Godwin Tomkinson (1932) found twenty-two cases in five generations of one family. Solomon (1963) reported twenty eight cases in seven generations of one family. Bakers (1961) reported four cases in one family. Summerly and Donaldson (1962) reported five patients from a family with nine affected over five generations.

Periodic inhibition of keratin synthesis probably explains the production of the constriction, however the cause of rhythmic action is not known. Pillsbury et al (1956) suggested abnormality of the amino-acid metabolism as the underlying cause. Summerly and Donaldson (1962) did not come across any biochemical defect in this genitically determined disorder. Grosfield et al (1965) however demonstrated intermittent arginino succinic aciduria in two unrelated patients which may still suggest a metabolic disorder.

The abnormal hair may grow all over the scalp or only in one or more localised patches. Some individuals begin to show the abnormality in small areas on the neck or frontal region and later it may involve the entire scalp. The hair of the eyebrows, eye lashes, axillae, pubic region and even that of the trunk and

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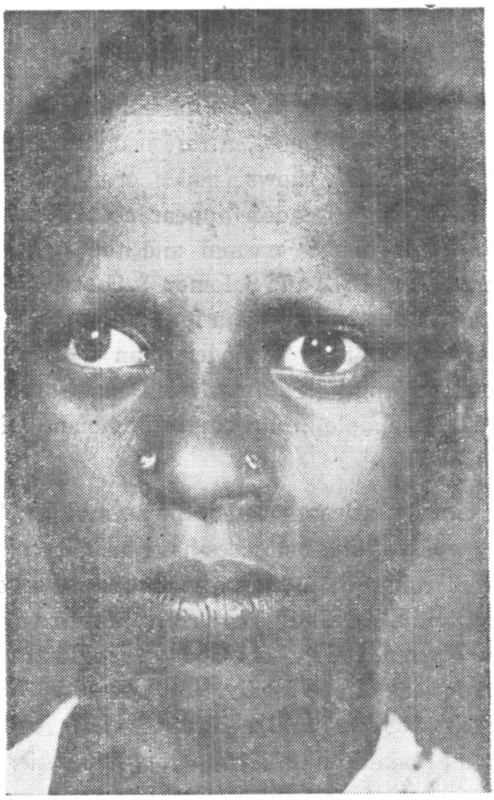
extremities may be similarly altered. The distribution of changes varies greatly even among members of the same family. In one case the alterations were limited only to the hairs of the legs (Butterworth and Strean, 1962).



Photograph 1: S, 4, case No. II showing prominance of the hair follicles over scalp with hyperkeratosis and gritty feel.
Note that there are no hairs over the area, which have all fallen off.

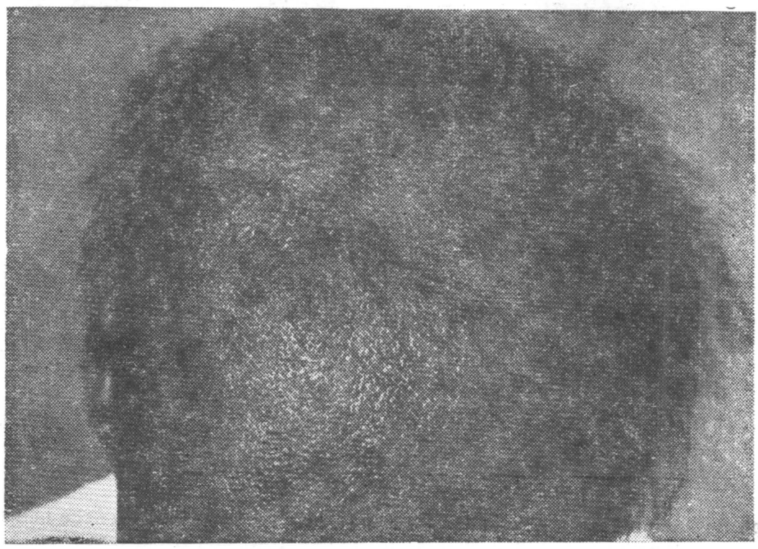
Keratosis pilaris or follicular hypekeratosis accompanies the hair changes in most cases and imparts "gritty" feeling (Baker, 1962). Among affected families, there may be individuals who only exhibit keratosis pillaris and transmit monilethrix to their children (Mackee and Rosen, 1916).

Apart from temporary improvement each summer, a considerably temporary improvement may occur in pregnancy Some cases may recover completely in later childhood or adult life (Rock et al, 1968). There is also naturral tendency of the disease to disappear about the age of puberty.

We came across a patient of monilethrix in a female patient. From the study of the family history, we could find out the disease in three more members of the same family i. e., her brother, whom we could examine and investigate (case II), her younger sister who died at an early age and her uncle (whom we are trying to persuade for investigations). However the parents of the children had consanguinous marriage i. e., the father having married the cousin sister. The two cases are reported below :

Case I : R., 11 years old, female presented with the chief complaints of sparseness of hairs over the scalp since birth. The patient's father stated that the child had since birth very small hairs which also disappeared after a couple of months, leaving behind prominent hair follicles. There is absence of hairs over the eyebrows, axillae and groins.

Family History : In the family, there was history of consanguinity of marriage amongst the parents, the father having married his first cousin (i. e., uncle's daughter). The younger brother of the patient is having the same disease and his case report is presented below as case No. II. The younger sister of the patient had also similar trouble but she died at an early age (cause of death unknown). Patient's uncle is having the same disease.



Photograph 2: Same case. Close-up from top of the scalp showing prominence of the hair follicles.

Examination revealed a young girl, average built and moderately nourished. There were few, small hairs over her scalp. There was absence of hairs over the eye-brows, axillae, pubis. The hair follicles over the scalp, eyebrows, axillae, pubis as well as on the arms were prominent and typically gritty on feel.

There was no other abnormality except tranmatic right lenticular opacity. Patient had not yet attained menarche. Her IQ was below normal.

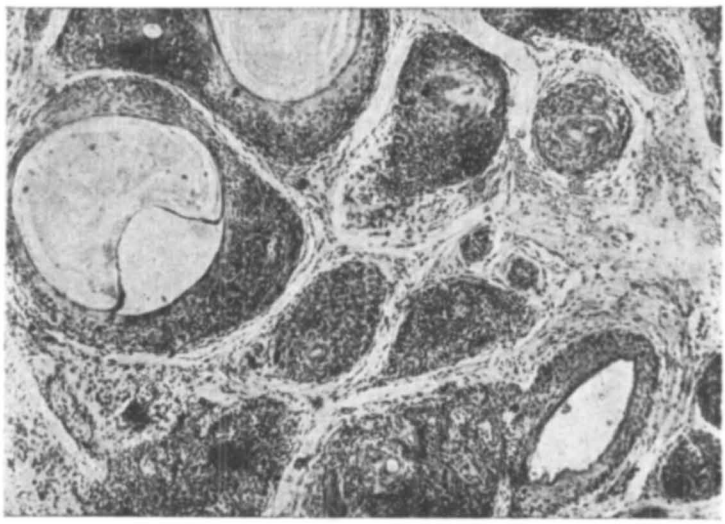
An examination of hair under the microscope showed bulbous thickening in some hairs. Biopsy taken from the scalp and arm revealed hyperkeratotic follicles.

Case II : S., 4 years old, male reported with the complaints of sparseness of hairs over the scalp and eyebrows ever since birth. There was prominence of hair follicles over the scalp, eyebrows, axillae and groins as well as over the arms and legs

The patient is the younger brother of the above patient.

There was hardly any hair to be examined. Biopsy of the prominent skin the scalp and arms showed predominance of hair follicles.

Discussion : Two cases of Monilethrix in a family of four affected, are reported as interesting clinical entity of academic interest. Apart from the sparseness of hairs in both the patients from the very beginning, there was follicular hyperkeratosis or keratosis pilaris over the usual hairy sites. The examination of hair in case I showed typical constriction and swellings alternating with each other. There are no special histological features attributed to the disease. The family shows involvement of three children and their paternal uncle. Further work on the family regarding pedigree study and chromosomal abnormalities is in progress and shall be reported in due course.



Photograph 3 : Same Case. View of fore-head showing the prominence of the hair follicles of the glabrous area as well as of the Eyebrows and Eyelashes.

Summary : Two clinical cases of Monilethrix in a family of four affected are presented. The literature on the subject is reviewed. The pedigree and chromosomal studies are in progress.

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