Trichoepithelioma Multiplex and Dystrophia Unguis Congenita: A New Syndrome?

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Abstract. A simultaneous appearance of trichoepitheliomas and nail dystrophy is described. Only the nail dystrophy was known in the family. Dermabrasion, together with electrosurgery, proved to be a satisfactory treatment for the trichoepitheliomas.

Key words: Trichoepithelioma multiplex; Nail dystrophy

Trichoepithelioma is a rare tumour differentiating towards hair follicles, usually multiple and often familial, with an autosomal dominant mode of inheritance in which there is a lessened expressivity and penetrance in males (1). In 1975 a syndrome of trichoepitheliomas, milia and cylindromas was published (3). Cylindromas may be similarly inherited and associated with or be a different expression of the same disease (2).

Trichoepitheliomas range in size from 2 to 30 mm and usually develop during puberty and chiefly around the nasolabial folds and eyelids, but also on the scalp and trunk.

CASE HISTORY

The patient was a 28-year-old, previously healthy man, who in 1978 had undergone appendectomy. The operation revealed a diverticulosis of the small bowel. Histological examination of diverticula showed pancreatic cells. During puberty a solitary tumour on the nose had appeared, followed gradually by more and more small pink, firm nodules arising in the nasolabial folds, on the eyelids and forehead. (Fig. 1). A biopsy from a tumour on the nose revealed cystic spaces filled with keratin, the marginal cells being flattened basophilic cells, and the presence of solid islands of the same kind of cells. The epidermal and surrounding tissue was normal.

Since birth the patient had had dystrophy of the nails. On both thumbs half of the nail was only partly developed and the index showed typical koilonychia (Fig. 2). His mother and his maternal grandmother and maybe his grandfather also had nail dystrophy but with onychia only of the thumbs (Fig. 3). The patient was without children.

In February 1980 a dermabrasion concluded with electrocautery was performed on the nose and the surrounding areas. Three months later the result was acceptable.

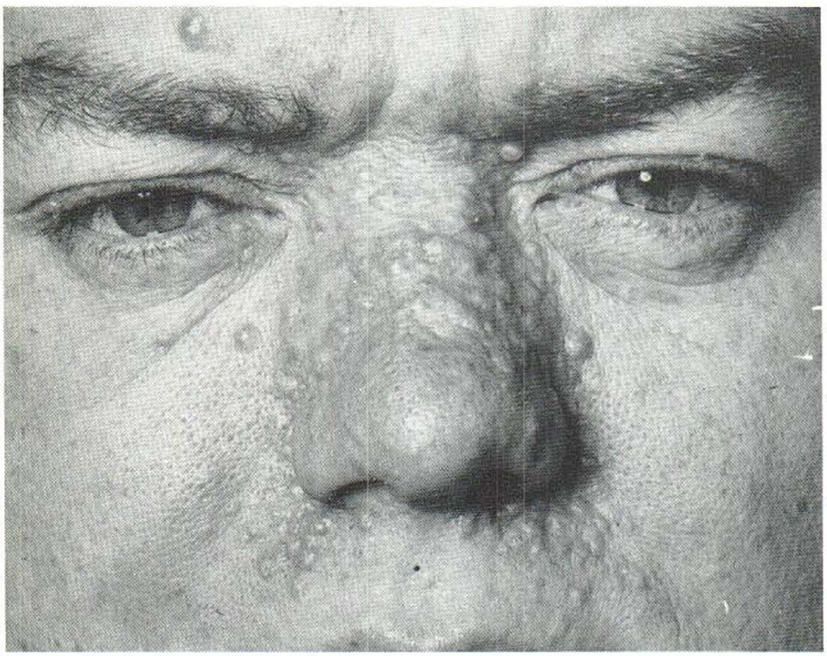

Fig. 1. The nose and surrounding areas of the patient, a 28-year-old male with trichoepithelioma.

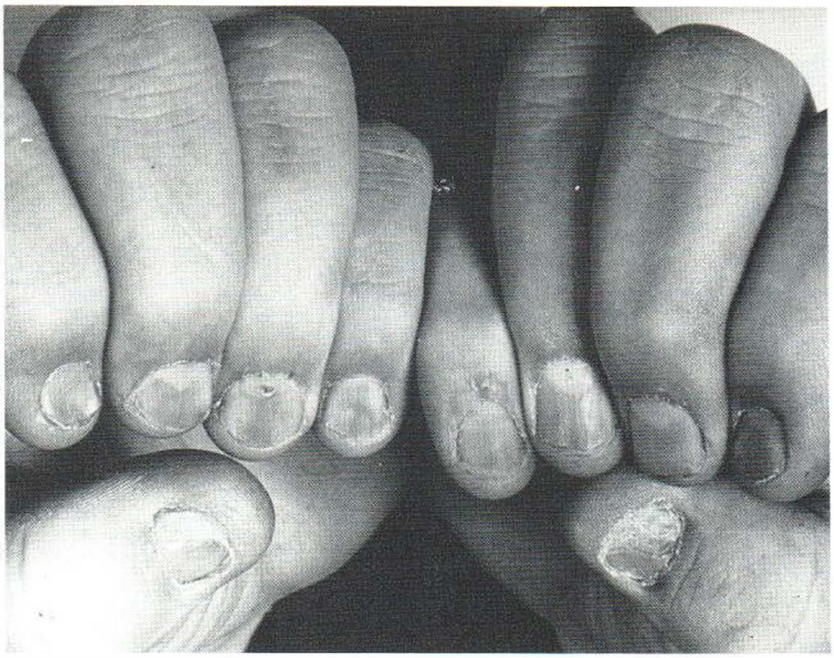

Fig. 2. The patient's nails.

DISCUSSION

Trichoepithelioma multiplex is often familial. Our patient, however, had no family history of trichoepithelioma on his mother's side. His father was unknown. The nail dystrophy seems most likely to have been inherited from his mother. Her mother had had the same kind of dystrophic nails.

The simultaneous appearance of trichoepitheliomas and nail dystrophy may be an example of coincidental genetic linkage or it may be a phenotypic expression of the same genotype of genetic defect.

The treatment with dermabrasion and electrocautery was deemed satisfactory.

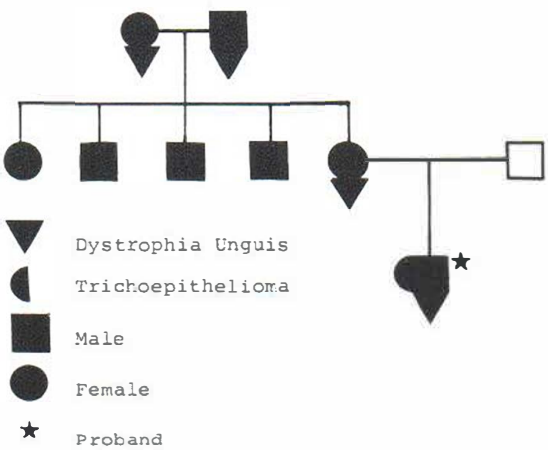

Fig. 3. Pedigree of the patient, showing the hereditary pattern of trichoepitheliomas and nail dystrophy.

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Megalopinna in Naevus Uniuslateris: A Case Report

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Abstract. A female patient, aged 10 years, with naevus uniuslateris limited to the left half of head and neck includ-