

DERMABRASION IN A GIRL WITH CONGENITAL POIKILODERMA (Syndrome Rothmund-Thomson)

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SUMMARY

A case of congenital poikiloderma in a 6-year-old girl is reported. The lesions on the face, ears and gluteal area appeared during her first year. Pathohistology of a biopsy taken from the face revealed an atrophic epidermis, dilated capillaries in the papillary dermis and a mild perivascular infiltrate composed of lymphocytes, histiocytes and a few melanophages. A test-dermabrasion was carried out in an area of fully developed lesions on the face. Six months later no lesions could be observed in the dermabraded area, a second biopsy displayed a normal histologic structure of the skin including hair follicles, sebaceous and sweat glands.

In view of such observations the author raises the question at what time in the embrional development do the malformations start in this condition.

KEY WORDS

congenital poikiloderma, syndrome Rothmund-Thomson, dermabrasion, 6-year-old girl

INTRODUCTION

Poikilodermas may be considered as a special group inside the broader concept of congenital atrophies. They appear soon after birth or during the early infancy, the main symptoms being skin atrophy, hyper- and hypopigmentation, teleangiectasia and occasionally also a pityriasiform scaling. Congenital poikiloderma or Thomson's syndrome was described in 1923 and later in 1936 (1). It is inherited in an autosomal recessive mode, the familial occurrence being rare, consanguinity of the parents is mostly not detectable. The sites of predilection are the face, ears, extremities and the gluteal region. The

lesions usually start to appear during the first year of life as a patchy erythema of the face. Hypotrichosis, a triangular shape of the face with a high front, hypertelorism and a small chin may also be expressed.

Certain authors believe that the syndromes of Thomson and Rothmund represent the same pathologic entity, although some signs may be missing (2). Rothmund who described the symptom in 1868 mentioned additionally to the skin symptoms, consanguinity of parents, a juvenile cataract, shortened extremities, photosensitivity, hypoplasia of the genitals as well as occurrence of malignancies (1,3,4,5,6,7).

