Clinical study

BASAL CELL NEVUS SYNDROME. TUMOR SITES AND THERAPY
Analysis of 56 cases

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ABSTRACT

In 56 patients with basal cell nevus syndrome, clinical manifestations and therapy in various tumor sites were reviewed and contrasted with data from literature. For differential diagnosis, it is essential to ascertain a complete history and to search for further symptoms of the syndrome. The most important alternatives in therapy are surgery and cryosurgery. The latter is particularly suitable in cases with large numbers of superficial basal cell carcinomas located on the trunk and extremities and for treatment during early growth stages. Cryosurgery sessions are short and the method can be employed in out-patient care, which is of special significance as basal cell nevus syndrome patients require life-long attention.

KEY WORDS

basal cell nevus syndrome, 56 patients, accompanying symptoms, tumor sites, therapy

INTRODUCTION

The basal cell nevus syndrome (BCNS, more accurate: nevoid basal cell carcinoma syndrome) is a genetically determined, polysymptomatic disease. By some authors it has therefore been labelled „fifth phacomatosis" (1). Musger (1964) (2) defined „phacomatosis" as heterogeneous alterations which are capable of progression and originate in dysplasias or early embryonic differentiation disorders. Apart from the skin, the basal cell nevus syndrome can involve to a various extent the skeletal system, the central nervous system, the eyes, and the endocrine organs. The disease shows an autosomal dominant pattern of inheritance with high penetrance and varying expressivity. The defect has recently been located to chromosome 9q in section q22-23 (3,4).

Since the close of the 19th century, cases of multiple basal cell carcinoma, osseous anomalies, mandibular cysts and retardation have been reported. Binkley and Johnson (5) described the cases of a mother and her daughter who both showed more than 1000 basal cell carcinomas and various growth defects. More publications, predominantly individual case reports, followed (6,7,8,9,10). Thies et al. (11), and a little later Gorlin and Goltz (12) identified the disease as a separate syndrome.

Characteristically, the tumor growth is initially very slow (nevoid growth stage) and becomes invasive later (oncotic growth stage) (13). It is not yet certain what causes this development. The multiple