GENETICS AND GENODERMATOSES

NEUROFIBROMATOSIS TYPE 1 : EMERGENCE OF NEW TYPE OF TUMORS

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Introduction: Neurofibromatosis type 1 (NF1) is the most common genodermatosis. Its incidence, prognosis and genetic mechanisms have been the subject of many publications. This autosomal dominant disease predisposes to the development of benign tumors (neurofibromas) but also malignant tumors. Cancers are the leading cause of premature death in patients with NF1. Frequently observed tumors are melanoma, pheochromocytoma, medullary thyroid carcinoma, glioma of the optic pathway, leukemia and breast cancer. Sarcomatous transformation remains exceptional.

Objective: The aim of our work is to reporte the peculiarities of owr 24 patients with NF1 and especially to share the new type of tumors complicating this disease found in owr series.

Material and methods: We reviewed all the files of patients hospitalized or who consulted for an NF1 between June 2015 and June 2018. The selected files were analyzed according to a grid including epidemiological, clinical, paraclinical, therapeutic and evolutionary data.

Results: Twenty four patients were enrolled, with a mean age of 33 years (13-68 years), with a sex ratio F / H of 0.62. Second degree consanguinity was found in 28% of cases. The average age of the diagnosis was 14 years old. School reflection was the tell-tale symptom of NF1 in 28% of cases, visual acuity (BAV) was decreased in 14. Tumor complications were diagnosed in 2 patients, the first tumor was an anterior abdominal wall synovial sarcoma developed during pregnancy and the second was a metastatic cutaneous sarcoma leading to the death of the patient.

Conclusion: The data of our study are similar to those of the literature, excluding the type of malignant tumors found. Adequate clinical follow-up must be coordinated to enable early detection of malignant tumors .