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Natural history and management of low-grade glioma in NF-1 children.

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Abstract

Pediatric neurofibromatosis type 1 (NF-1) patients are prone to developing low-grade glioma (LGG). The HIT-LGG study 1996 aimed to observe the natural history of pediatric LGG and to postpone irradiation in younger children by using carboplatinum and vincristine in case non-surgical treatment was required. A total of 109 of 1,044 (10.4%) protocol patients had a genetic NF-1 trait [57 female patients; median age 5.1 years (range 1-15.4 years)]. Eighty-three patients (76%) suffered from an optic pathway tumor. Neuroimaging only allowed diagnosis in 67 patients. Histology revealed pilocytic astrocytoma WHO grade I in 38 of 42 biopsied patients. Sixty-five (60%) patients received non-surgical treatment, either chemotherapy (n = 55) or irradiation (n = 10). The overall survival rate of 96% after a median observation time of 5.25 years contrasts with an event free survival rate (EFS) of 0.24 at 5 years. Progressive LGG were observed even in children older than 11 years. Chiasmatic/postchiasmatic localization was a univariate risk factor for progressive disease. In the chemotherapy group, we observed a 5-year progression-free survival (PFS) rate of 0.73. Similarly, the PFS rate in the irradiation group was 0.78. Multivariate analysis revealed surgical intervention and localization within the optic pathway as factors that increased the risk of tumor progression. In this large prospective multinational study, LGG in NF-1 patients did progress in 75% of patients. Chemotherapy yielded acceptable PFS. The biological factors determining progression remain poorly understood.

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