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[Childs Nerv Syst.](#) 2009 Nov 25. [Epub ahead of print]

Neurofibromatosis type 1 and high-grade tumors of the central nervous system.

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PURPOSE: Neurofibromatosis type 1 (NF1), a common genetic disorder, predisposes patients to the development of both benign and malignant tumors. Although the most common central nervous system (CNS) tumor is a low-grade pilocytic astrocytoma of the optic pathway, there have been sporadic reports of NF1 patients with more aggressive CNS lesions. We investigated the incidence of aggressive CNS lesions in NF1 patients at our institution. **METHODS:** We conducted a retrospective review of all patients with NF1 and any CNS tumor being followed in the Children's Memorial Hospital NF1 Clinic. **RESULTS:** Seven hundred forty patients with a diagnosis of NF1 were identified. Of these, 145 (20%) patients had CNS tumors, 99 (68%) of whom had optic pathway tumors (OPTs). Five patients (3%) were identified as having high-grade tumors, which consisted of anaplastic medulloblastoma (n = 1) and high-grade glioma (n = 4). The mean age at diagnosis of NF1 was 2 years. Three of the five patients had a history of an OPT prior to the development of their high-grade lesions. The clinical courses and treatment of these five patients varied. Currently, two patients are alive and receiving therapy at a mean of 10 months following diagnosis. **CONCLUSION:** High-grade CNS tumors may occur in children with NF1. Although tumors in NF patients are generally benign, clinicians should have a high index of suspicion of malignancy in patients whose tumors are in an unusual location or behave in an uncharacteristically aggressive manner.

PMID: 19937438 [PubMed - as supplied by publisher]

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