Long-term natural history of neurofibromatosis Type 2-associated intracranial tumors.

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Abstract

OBJECT: Neurofibromatosis Type 2 (NF2) is a heritable tumor predisposition syndrome that leads to the development of multiple intracranial tumors, including meningiomas and schwannomas. Because the natural history of these tumors has not been determined, their optimal management has not been established. To define the natural history of NF2-associated intracranial tumors and to optimize management strategies, the authors evaluated long-term clinical and radiographic data in patients with NF2.

METHODS: Consecutive NF2 patients with a minimum of 4 years of serial clinical and MRI follow-up were analyzed.

RESULTS: Seventeen patients, 9 males and 8 females, were included in this analysis (mean follow-up 9.5±4.8 years, range 4.0-20.7 years). The mean age at initial evaluation was 33.2±15.5 years (range 12.3-57.6 years). Patients harbored 182 intracranial neoplasms, 164 of which were assessable for growth rate analysis (18 vestibular schwannomas [VSs], 11 nonvestibular cranial nerve [CN] schwannomas, and 135 meningiomas) and 152 of which were assessable for growth pattern analysis (15 VSs, 9 nonvestibular CN schwannomas, and 128 meningiomas). New tumors developed in patients over the course of the imaging follow-up: 66 meningiomas, 2 VSs, and 2 nonvestibular CN schwannomas. Overall, 45 tumors (29.6%) exhibited linear growth, 17 tumors (11.2%) exhibited exponential growth, and 90 tumors (59.2%) displayed a saltatory growth pattern characterized by alternating periods of growth and quiescence (mean quiescent period 2.3±2.1 years, range 0.4-11.7 years). Further, the saltatory pattern was the most frequently identified growth pattern for each tumor type: meningiomas 60.9%, VSs 46.7%, and nonvestibular schwannoma 55.6%. A younger age at the onset of NF2-related symptoms (p=0.01) and female sex (p=0.05) were associated with an increased growth rate in meningiomas. The identification of saltatory growth in meningiomas increased with the duration of follow-up (p=0.01).

CONCLUSIONS: Neurofibromatosis Type 2-associated intracranial tumors most frequently demonstrated a saltatory growth pattern. Because new tumors can develop in NF2 patients over their lifetime and because radiographic progression and symptom formation are unpredictable, resection may be best reserved for symptom-producing tumors. Moreover, establishing the efficacy of nonsurgical therapeutic interventions must be based on long-term follow-up (several years).

Comment in

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