Atypia predicting prognosis for intracranial extraventricular neurocytomas.

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Abstract

OBJECT: The literature, at present, provides limited information about extraventricular neurocytomas (EVNs) and is almost exclusively composed of case reports or small case series. Treatment for EVNs has largely been guided by results from central neurocytoma outcome studies. The authors present an analysis of all reported intracranial EVN cases to establish if tumor histopathological features can substratify EVN into groups with differing prognosis and help guide treatment decisions.

METHODS: The authors identified studies reporting histology, treatment modality, and outcomes for patients with intracranial EVN. The rates of recurrence and survival for patients were compared using Kaplan-Meier analysis. Atypical tumors, defined by MIB-1 labeling index exceeding 3% or atypical histological features, were compared with typical tumors, and patients 50 years of age or older were compared with those younger than 50 years of age.

RESULTS: Eighty-five patients met the inclusion criteria, and 27% of them had an atypical histology. Typical EVNs had a better prognosis than atypical EVNs after primary treatment, with a 5-year recurrence rate of 36% compared with 68% (p < 0.001), and a 5-year mortality rate of 4% compared with 44%, respectively (p < 0.001). Age younger 50 years was associated with a better prognosis than age equal to or greater than 50 years, with a 5-year recurrence rate of 33% and 74%, respectively (p < 0.001), and a 5-year mortality rate of 4% and 52%, respectively (p < 0.001). Multivariate analysis demonstrated that atypical EVNs carried significantly increased risk for recurrence (hazard ratio [HR] 4.91, p < 0.001) and death (HR 22.91, p < 0.01). Gross-total resection was superior to subtotal resection (STR) alone in tumor control rates for typical EVNs (95% and 68%, p < 0.05), and there was a trend for adjuvant external-beam radiotherapy to benefit STR. There was suggestion of similar trends in patients with atypical EVNs.

CONCLUSIONS: There are at least 2 distinct histological subtypes of EVN, with different prognostic significances. Atypia or MIB-1 labeling index greater than 3% is a significant predictor of poor prognosis for EVNs. Complete resection or more aggressive attempts at providing adjuvant therapy following STR appear to improve the prognosis for patients with EVNs. Although the authors' results are informative, there are limitations to their analysis. Given the relatively modest total number of cases reported, as well as the nature of the disaggregated analysis, the authors were not able to use formal meta-analytical methods to limit the impact of between center heterogeneity. Additionally, they were not able to control for individual differences in data analysis and presentation across the different studies included in their analysis.