Tumor recurrence and malignant progression of gangliogliomas.

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BACKGROUND: Most gangliogliomas (GGs) are benign tumors, but tumor recurrence and malignant progression are observed in some patients. METHODS: The authors analyzed their experience with 4 recurrent/progressive GGs (World Health Organization [WHO] grade I), 21 tumors with atypical features (WHO grade II), and 5 tumors with anaplastic histologic features (WHO grade III). Histopathologic findings (23 patients) were reviewed. The mean follow-up was 5.9 years (median, 4.5 years; range, 0.5-14.7 years). RESULTS: The 5-year survival rates were only 79% for patients who had tumors with atypical features and 53% for patients who had WHO grade III tumors. Secondary glioblastomas were diagnosed in 5 of 11 patients (45%) who underwent surgery for tumor recurrence. Age at surgery <40 years (P = .007) was associated significantly with better overall survival (OS), but it was not associated with better progression-free survival (PFS). Clinical presentation (drug-resistant epilepsy vs all other patients with seizures vs no seizures) was associated significantly with better OS (P = .005) and PFS (P < .001). Patients who had extratemporal tumors had a significantly shorter PFS (P = .01) but not OS. A complete resection was correlated strongly with both OS (P = .002) and PFS (P = .001). Neuropathologic examination revealed the presence of a gemistocytic cell component (PFS, P = .025), a lack of protein droplets (OS, P = .04; PFS, P = .05), and focal tumor cell-associated CD34 immunolabeling (OS, P = .03) as significant predictors of an adverse clinical course. CONCLUSIONS: The current data supported a 3-tiered GG histopathologic grading system that included an intermediate diagnostic category (atypical GG, WHO grade II). Careful attention to histopathologic findings and clinical parameters usually will identify patients who are at risk for an adverse clinical course.

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