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Predicting disease progression and the need for tumor-directed treatment in tectal plate gliomas

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Abstract

Objective: Tectal plate gliomas are rare, slow-growing tumors of the midbrain that are discovered predominantly in the pediatric population. Because of their indolent nature, treatment mainly consists of observation and management of hydrocephalus. Unfortunately, a subset of tectal gliomas may exhibit tumor enlargement and disease progression. Currently, there are no established guidelines for predicting future progression of tectal gliomas or the need for tumor-directed treatment. In this paper, the authors present a large case series of tectal plate gliomas with the aim of determining early indicators of tumor progression and the need for tumor-directed treatment in a pediatric population, along with providing their experience in treating progressive tumors.

Methods: A retrospective chart review of 170 patients diagnosed with tectal plate glioma from a single institution, of whom 67 were pediatric patients (≤ 18 years of age), was performed. Univariate analysis was used to determine statistically significant predictors of symptomatic disease progression requiring eventual tumor-directed therapy.

Results: The median patient age of the full cohort was 24 years (range 0-73 years). Compared with the pediatric population, the adult population had more instances of incidental lesions ($p < 0.001$) and lower rates of hydrocephalus (50% vs 84%, $p < 0.001$). Of the pediatric patients who had ≥ 5 years of follow-up (n = 51), 12 (24%) experienced radiological progression and 13 (25%) required treatment for their tumor. The 1-year, 5-year, and 10-year radiographic progression-free survival (PFS) rates were 98%, 90%, and 86%, respectively. In univariate analysis, lesion involvement of the pons, moderate T1 hypointensity, and moderate contrast enhancement on baseline radiology were significantly associated with worse radiographic PFS. Alternatively, significant predictors of requiring tumor-directed treatment included extraocular eye movement abnormalities at presentation, involvement of the lesion beyond the tectum on baseline radiology, moderate T1 hypointensity, moderate contrast enhancement, and an increase in total lesion size during progression. At the most recent follow-up, 94% of the patients had stable/nonprogressive disease, 2% had progressive disease, and 4% died of tumor progression.

Conclusions: Patients who demonstrate radiographic progression may not necessarily experience clinical/symptomatic progression or require tumor-directed treatment. Certain patient presentation characteristics and baseline radiographic features may be predictive of worse radiographic PFS or the need for future tumor-directed treatment in the pediatric population. Typically, the natural history of these lesions lends to excellent long-term survival, even in patients who experience clinical

progression, should appropriate treatment be initiated.

Keywords: midbrain glioma; midbrain tumor; tectal glioma; tectal plate glioma; tectal tumor.