ABSTRACT

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Adult diffuse intrinsic pontine glioma: clinical, radiological, pathological, molecular features, and treatments of 96 patients.

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OBJECTIVE: Unlike its pediatric counterpart, adult diffuse intrinsic pontine glioma (DIPG) remains largely unelucidated. In this study, the authors examined the clinical, radiological, pathological, molecular, and clinical aspects of 96 adult DIPGs.

METHODS: The National Brain Tumor Registry of China (April 2013-December 2019) was used to collect data on radiologically diagnosed adult DIPG patients. Survival analysis was conducted using Kaplan-Meier curves and univariate and multivariate Cox regression. The chi-square test/Wilcoxon rank-sum test and multivariable logistic regression were used to examine the clinical and radiological characteristics of patients with long-term survival (LTS). Interaction analyses between clinical factors were also conducted.

RESULTS: The median age at symptom onset was 33.5 years, and the median duration of symptoms was 4.5 months. The frequencies of H3K27M and IDH1 mutations were 37.2% and 26.5%, respectively. All adult DIPG patients had a median overall survival (OS) of 19.5 months, with 1-, 2-, and 3-year survival rates of 67.0%, 42.8%, and 36.0%, respectively. The median OS of 40 patients who did not undergo treatment was 13.4 months. Patients with H3K27M-mutant tumors had a poorer prognosis than those with IDH-mutant tumors ($p < 0.001$) and H3K27M(-)/IDH-wild-type tumors ($p = 0.002$), with a median OS of 11.4 months. The median OSs of patients with H3K27M-mutant tumors who received treatment and those who did not were 13.8 months and 7.5 months, respectively ($p = 0.016$). Among patients with and without a pathological diagnosis, H3K27M mutation ($p < 0.001$) and contrast enhancement on MRI ($p = 0.003$), respectively, imparted a worse prognosis. Treatments were the predictive factor for patients with H3K27M-mutant tumors ($p = 0.038$), whereas contrast enhancement on MRI was the prognostic factor for the H3K27M(-) group ($p = 0.038$). In addition, H3K27M mutation and treatment were significant predictors for patients with symptom duration ≤ 4 months (H3K27M, $p = 0.020$; treatment, $p = 0.014$) and tumors with no contrast enhancement (H3K27M, $p = 0.003$; treatment, $p = 0.042$). Patients with LTS were less likely to have cranial nerve palsy ($p = 0.002$) and contrast enhancement on MRI at diagnosis ($p = 0.022$).

CONCLUSIONS: It is recommended that all adult DIPG patients undergo genomic testing for H3K27M and IDH mutations. Despite the low prevalence, additional study is needed to better characterize the efficacy of various treatment modalities in adults with DIPG.

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