ABSTRACT

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# Characteristics of Children ≤36 Months of Age with Diffuse Intrinsic Pontine Glioma (DIPG): A Report from the International DIPG Registry.

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BACKGROUND: Children ≤36 months with Diffuse Intrinsic Pontine Glioma (DIPG) have increased long-term survival (LTS, overall survival (OS) ≥24 months). Understanding distinguishing characteristics in this population is critical to improving outcomes.

METHODS: Patients ≤36 months at diagnosis enrolled on the International DIPG Registry (IDIPGR) with central imaging confirmation were included. Presentation, clinical course, imaging, pathology and molecular findings were analyzed.

RESULTS: Among 1183 patients in IDIPGR, 40 were eligible (median age: 29 months). Median OS was 15 months. Twelve patients (30%) were LTS, 3 (7.5%) very long-term survivors ≥ 5 years. Among 8 untreated patients, median OS was 2 months. Patients enrolled in the registry but excluded from our study by central radiology review or tissue diagnosis had median OS of 7 months. All but 1 LTS received radiation. Among 32 treated patients, 1-, 2-, 3-, and 5-year OS rates were 68.8%, 31.2%, 15.6% and 12.5%, respectively. LTS had longer duration of presenting symptoms (p=0.018). No imaging features were predictive of outcome. Tissue and genomic data were available in 18 (45%) and 10 patients, respectively. Among 9 with known H3K27M status, 6 had a mutation.

CONCLUSIONS: Children ≤36 months demonstrated significantly more LTS, with an improved median OS of 15 months; 92% of LTS received radiation. Median OS in untreated children was 2 months, compared to 17 months for treated children. LTS had longer duration of symptoms. Excluded patients demonstrated a lower OS, contradicting the hypothesis that children ≤36 months with DIPG show improved outcomes due to misdiagnosis.



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