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Clinical, Histological and Molecular Features of Gliomas in Adults with Neurofibromatosis Type 1

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Abstract

Background: People with NF1 have an increased prevalence of CNS malignancy. However, little is known about the clinical course or pathologic features of NF1-associated gliomas in adults, limiting clinical care and research.

Methods: Adults (≥18 years) with NF1 and histologically confirmed non-optic pathway gliomas (non-OPGs) at Johns Hopkins Hospital, Memorial Sloan Kettering Cancer Center, and Washington University presenting between 1990-2020 were identified. Retrospective data were collated, and pathology reviewed centrally.

Results: Forty-five patients, comprising 23 females (51%), met eligibility criteria, with a median of age 37 (18-68 years) and performance status of 80% (30-100%). Tissue was available for 35 patients. Diagnoses included infiltrating (low-grade) astrocytoma (9), glioblastoma (7), high-grade astrocytoma with piloid features (4), pilocytic astrocytoma (4), high-grade astrocytoma (3), WHO diagnosis not reached (4) and one each of gliosarcoma, ganglioglioma, embryonal tumor, and diffuse midline glioma. Seventy-one percent of tumors were midline and underwent biopsy only. All 27 tumors evaluated were IDH1-wild-type, independent of histology. In the 10 cases with molecular testing, the most common genetic variants were NF1, EGFR, ATRX, CDKN2A/B, TP53, TERT, and MSH2/3 mutation. While the treatments provided varied, the median overall survival was 24 months [2-267 months] across all ages, and 38.5 [18-109] months in individuals with grade 1-2 gliomas.

Conclusions: Non-OPGs in adults with NF1, including low-grade tumors, often have an aggressive clinical course, indicating a need to better understand the pathobiology of these NF1-associated gliomas.

Keywords: Neurofibromatosis type 1; Non-optic pathway glioma; glioma; overall survival; treatment outcome.

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