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[Fulltext](#) | [PDF \(3.43 M\)](#)**Gliomas in Neurofibromatosis Type 1: A Clinicopathologic Study of 100 Patients.****Original Articles**

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*Rodriguez, Fausto J. MD; Perry, Arie MD; Gutmann, David H. MD, PhD; O'Neill, Brian Patrick MD; Leonard, Jeffrey MD; Bryant, Sandra MS; Giannini, Caterina MD, PhD***Abstract:**

There are few pathologic studies of gliomas in patients with neurofibromatosis type 1. We analyzed clinical and pathologic features of gliomas from 100 neurofibromatosis type 1 patients (57 men; 43 women). The median age at tumor diagnosis was 13 years (range, 4 months to 68 years). Most tumors were typical pilocytic astrocytoma (PA) (49%) or diffusely infiltrating astrocytoma (DA) (27%) that included World Health Organization Grades II (5%), III (15%), and IV (7%); others were designated as low-grade astrocytoma, subtype indeterminate (LGSi; 17%). Two pilomyxoid astrocytomas, 1 desmoplastic infantile ganglioglioma and 1 conventional ganglioglioma, were also identified. The tumors in 24 cases arose in the optic pathways and included PA (n = 14), LGSi (n = 4), DA (n = 4), pilomyxoid astrocytoma (n = 1), and ganglioglioma (n = 1). The prognoses of the PA and LGSi gliomas overall were generally favorable; there were no survival differences between PA and LGSi groups based on site, tumor size, mitotic activity, or MIB-1 labeling index. In the combined PA and LGSi group, age younger than 10 years and gross total resection were associated with an increased overall survival rate (p = 0.047 and 0.002, respectively). Compared with the combined group (PA + LGSi), patients with DA at all sites had decreased overall and recurrence-free survival times (p < 0.001 and p = 0.003, respectively). This study emphasizes the wide histologic spectrum of gliomas that occur in patients with neurofibromatosis type 1. Classic PA and LGSi are the most common, and most have favorable prognoses. By contrast, DAs are more aggressive, similar to those that arise sporadically.

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