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The impact of changes in gadolinium-enhancement on disease progression in children with neurofibromatosis type 1-associated optic pathway glioma: a retrospective analysis

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Abstract

Purpose: There has been limited investigation of imaging features associated with visual acuity (VA) decline and initiation of treatment for patients with neurofibromatosis type 1 (NF1) and optic pathway glioma (OPG).

Methods: To evaluate the association of increased gadolinium enhancement with decline in VA, initiation of chemotherapy, and tumor growth, we performed a retrospective cohort study of children diagnosed with NF1-OPG between January 2006 to June 2016. Two cohorts were defined: a new diagnosis and a longitudinal cohort. Outcomes were examined at 1 and 2 years from initial diagnosis, and 1 and 2 years from initial increase in enhancement in the longitudinal cohort.

Results: Eighty patients were eligible; all 80 contributed to the new diagnosis cohort and 73 to the longitudinal cohort. Fifty-six patients (70%) demonstrated enhancing NF1-OPG at diagnosis. 39% of patients in the new diagnosis cohort and 45% of those in the longitudinal cohort developed increased enhancement during the study period. There was no significant association between increases in enhancement and VA decline in the newly diagnosed or longitudinal cohorts, as well as with initiation of treatment in the longitudinal cohort. Although there was an association of enhancement increase with treatment in the new diagnosis cohort, this association was not maintained when stratified by concurrent change in tumor size.

Conclusion: Increased gadolinium-enhancement independent of a concurrent increase in tumor size on MRI should not be used as a marker of NF1-OPG progression and does not appear to be associated with visual decline or initiation of chemotherapy.

Keywords: Cancer epidemiology; Magnetic resonance imaging; Neurofibromatosis type 1; Optic pathway glioma.