Teaching Neuro*Image*

Benign and malignant pathology in neurofibromatosis type 1

Melanie Walker, MD; and Patrik Gabikian, MD

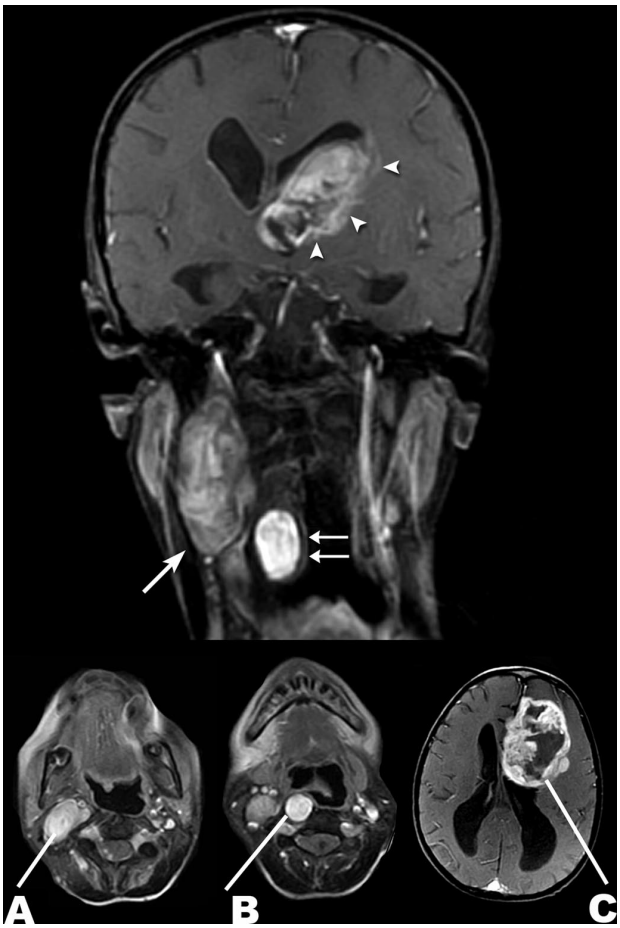

Figure. (Top) Gadolinium-enhanced coronal MRI of the brain revealed ventriculomegaly, right carotid sheath (arrow) and retropharyngeal nerve sheath tumors (double arrows), and a large heterogeneous left frontal mass with intraventricular extension (arrowheads). The extracranial lesions represent benign tumors seen in neurofibromatosis type 1. The left frontal mass was determined to be a gliosarcoma. (Bottom) Gadolinium-enhanced axial MRI reveals further detail of carotid sheath tumor (A), retropharyngeal nerve sheath tumor (B), and frontal gliosarcoma (C).

A 25-year-old man presented for evaluation of aphasia and generalized seizures. Examination revealed café au lait macules, axillary freckling, and bilateral Lisch nodules. Surgical history included excision of an upper extremity plexiform neurofibroma and optic glioma resection followed by whole-brain radiation 20 years previously. His mother and maternal grandmother had also been diagnosed with neurofibromatosis type 1 (NF1). Recent onset signs and symptoms were attributed to the frontal mass, biopsy proven to be gliosarcoma. Malignant gliomas can arise as a late consequence of whole-brain radiotherapy or result from malignant degeneration of previously benign tumors in patients with NF1[1,2] figure.

References

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From the Departments of Neurology (M.W.) and Neurological Surgery (P.G.), University of Washington School of Medicine, Seattle.

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Address correspondence and reprint requests to Dr. M. Walker, Neurology Box 359775, Harborview Medical Center, University of Washington School of Medicine, 325 9 Ave., Seattle, WA 98104; e-mail: walkerm@u.washington.edu

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