Metastatic brainstem pheochromocytoma in a patient with von Hippel–Lindau disease

Case illustration

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Patients with von Hippel–Lindau (VHL) disease frequently harbor cerebellar hemangioblastomas that enhance densely on magnetic resonance (MR) images.1 This 63-year-old woman with VHL presented with a hemangioblastoma in the orbit and multiple enhancing cerebellar tumors (Fig. 1). She had previously undergone an enucleation of her left eye for an ocular hemangioblastoma, an adrenalectomy for a pheochromocytoma, and multiple resections of cerebellar and spinal cord hemangioblastomas. In addition, she was known to have disseminated pheochromocytomas.

The patient was admitted for resection of the orbital hemangioblastoma; however, during the immediate preoperative period she experienced severe hypertension and, thus, surgery was postponed. Workup included a 99Tc/I meta-iodobenzylguanidine (MIBG) scan that revealed MIBG uptake into an enhancing brainstem lesion (Fig. 2). A guanethidine analog, MIBG competes with norepinephrine for uptake into adrenergic neurons. The uptake of MIBG into an enhancing lesion that had the typical appearance of a brainstem hemangioblastoma, a tumor we have commonly found in hundreds of patients with VHL, indicated that this tumor was instead a metastatic pheochromocytoma (98.9% specificity of the method).2 The records of eight patients with hemangioblastomas and pheochromocytomas were retrospectively examined and no evidence of MIBG uptake was found in any of their hemangioblastomas. Because patients with VHL have a higher incidence of both hemangioblastomas and pheochromocytomas, which appear similar on MR images, it is important to consider the possibility that a central nervous system (CNS) mass that appears to be a hemangioblastoma may be a metastatic pheochromocytoma, especially in a patient with hypertension. The distinction may affect operative strategy.

References