Case Report: Hemangioblastoma of the Cerebellum in an Octogenarian

Louis J. Kim, MD
Peter H. Maughan, MD
Robert Evans, PA†

In the elderly, symptomatic posterior fossa lesions are usually suspected to be metastatic disease. In this report, we describe the rare presentation of a symptomatic posterior fossa hemangioblastoma in an 83-year-old woman. Familiarity with the classic radiological and clinical features allows clinicians to diagnose these lesions correctly, even in the unusual case of an elderly patient.

Key Words: cerebellum, elderly, hemangioblastoma

Abbreviations Used: CT, computed tomography; MR, magnetic resonance; PICA, posterior inferior cerebellar artery; SCA, superior cerebellar artery

Division of Neurological Surgery, Barrow Neurological Institute, St. Joseph's Hospital and Medical Center, Phoenix, Arizona, and †Scottsdale Memorial Osborn Hospital, Scottsdale, AZ

The differential diagnosis for a homogenously enhancing posterior fossa lesion in the elderly population is relatively limited. Most clinicians suspect metastatic disease unless proven otherwise. We report a rare case of a symptomatic cerebellar hemangioblastoma in an octogenarian.

Clinical Presentation

Over several months, an 83-year-old woman had experienced progressive confusion, memory deficits, and gait difficulties. During this time, the possibility of dementia was raised. At admission the patient suffered from severe nausea, vomiting, confusion, and headache. Laboratory studies revealed a hematocrit of 53%. On examination, she demonstrated lateral nystagmus, left-sided dysmetria, and dysdiadochokinesia. Noncontrast head CT revealed significant transependymal edema and obstructive hydrocephalus (Fig. 1). A large left cerebellar mass was identified. CT of the chest, abdomen, and pelvis showed an adrenal cyst (negative needle biopsy) and no evidence of a primary malignancy. At this time, a bedside ventriculostomy was placed, with an opening pressure of 18 mm Hg. After the procedure the patient's acute nausea and vomiting subsided.

MR imaging showed a brightly contrast-enhancing tumor of the left cerebellum, extending along the ipsilateral tonsil, which herniated below the foramen magnum. Two large peritumoral cysts were also evident. Mass effect on the adjacent brainstem and fourth ventricle produced obstructive hydrocephalus (Fig. 2). Due to the radiographic

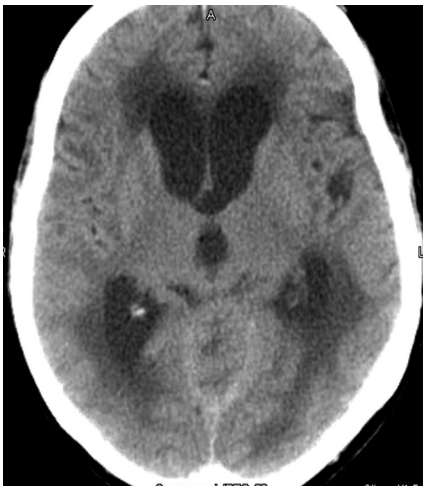

Figure 1. Nonenhanced axial head CT shows significant hydrocephalus associated with transependymal edema.

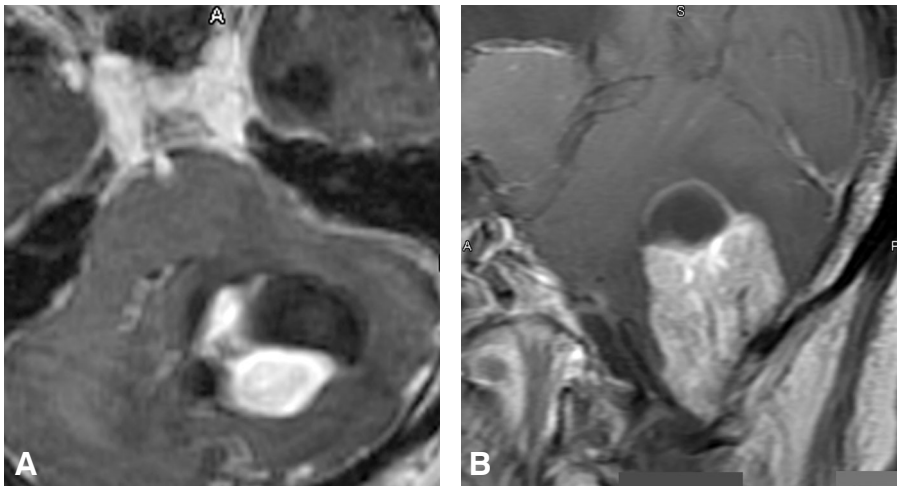

Figure 2. (*A*) Axial and (*B*) sagittal gadolinium-enhanced MR images show a large, brightly enhancing mass in the left cerebellar hemisphere extending down below the foramen magnum. Peritumoral cysts are evident. Note the presence of obstructive hydrocephalus.

features, polycythemia, and clinical history, hemangioblastoma was the leading diagnosis.

The patient underwent cerebral angiography and an attempt at tumor embolization. However, due to the patient's severely tortuous arterial anatomy along the left subclavian and vertebral arteries, embolization could not be performed. Diagnostic imaging indicated that the primary feeding arteries were from the PICA and SCA (Fig. 3A). Angiographically, the arterial blush demonstrated the extraordinary vascular nature of this tumor (Fig. 3B).

Given the radiographic mass effect and dramatic clinical symptoms, the patient underwent a far-lateral craniotomy and gross total resection of the lesion (Fig. 4A and B). Histological analysis confirmed the diagnosis of hemangioblastoma. Except for her baseline cerebellar signs, the patient's postoperative course was benign. Postoperative imaging of the spinal cord revealed no further lesions.

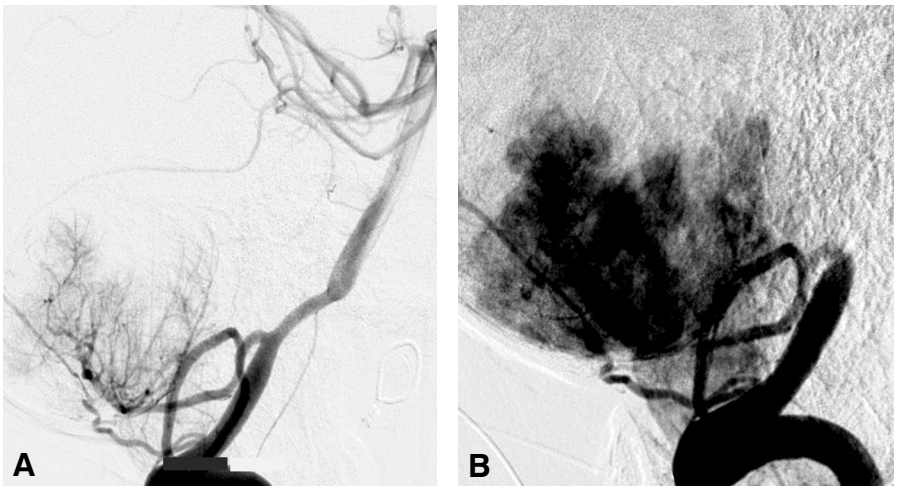

Figure 3. (*A*) Early arterial phase of this left vertebral angiogram shows a large feeding branch from the PICA. (*B*) Capillary phase of the same angiogram shows a dense vascular blush of the tumor mass consistent with, but not pathognomonic for, hemangioblastoma.

Discussion

The differential diagnosis for an intraaxial enhancing mass of the posterior fossa in the elderly is a metastasis until proven otherwise. This diagnosis corresponds with a poor overall prognosis. However, as this case demonstrates, clinicians should be aware of hemangioblastomas as a rare cause of brightly enhancing posterior fossa masses in the elderly.[2] Features such as intense enhancement, flow voids, and peritumoral cysts can be radiographic indicators of hemangioblastomas. Extremely vascular metastatic tumors, such as renal cell carcinoma hypernephroma, can mimic hemangioblastomas radiographically.[7] Therefore, it is reasonable to rule out a primary carcinoma as a potential etiology in the elderly or in patients with risk factors for cancer. Other rare causes of intraaxial tumors in the posterior fossa include glioblastomas, ependymomas, choroid plexus carcinomas, medulloblastomas, and lymphomas.[11]

Hemangioblastomas are benign vascular tumors. If suspected clinically, aggressive treatment may be warranted based on the patient's clinical symptoms and radiographic evidence of mass effect. Surgical management is based on a case-by-case analysis. Preoperative embolization, while not mandatory, can be useful for large tumors and for lesions adjacent to the brainstem or fourth ventricle.[9] Surgical extirpation is curative. Radiosurgery, although supported by

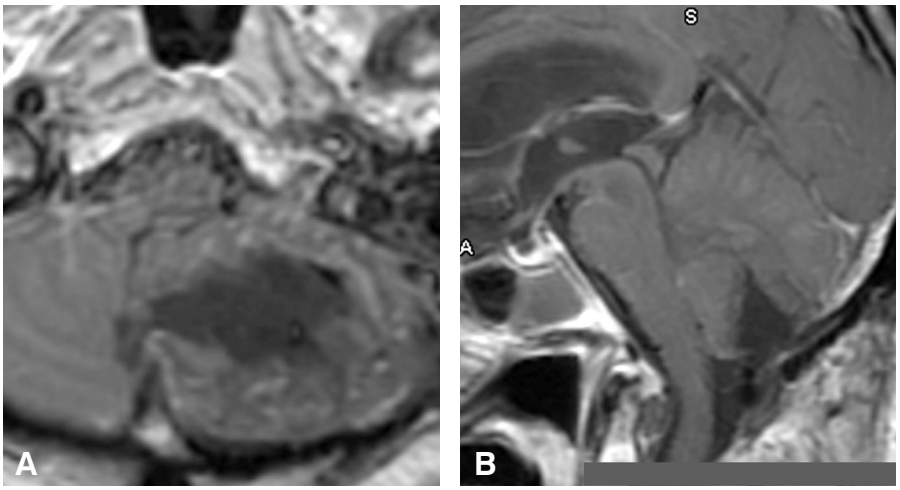

Figure 4. (*A*) Axial and (*B*) sagittal gadolinium-enhanced MR images confirm gross total resection of the tumor. Frozen and permanent sections (*not shown*) confirmed the diagnosis of hemangioblastoma.

some authors,[5,8] has no proven role in the treatment of this disease.

Hemangioblastomas compose 1 to 2.5% of all intracranial tumors, with about 85% occurring in the cerebellum. Less commonly, they are found in the spinal cord, medulla, and cerebrum.[3,6,12] Histologically, hemangioblastomas are composed of three cell types: pericytes, endothelial cells, and stromal cells.[4] Due to the vascular nature of these tumors, histopathological sections frequently catch branched arterioles in longitudinal sections, creating the classic "staghorn" feature. Approximately 60% of hemangioblastomas are solid tumors, while 40% can have cystic components. Cysts contain clear, straw-colored fluid, and the cyst wall does not need to be removed during resection. Enhancing mural nodules and solid components, however, must be treated with gross total resection.

Hemangioblastomas usually become symptomatic in the third to fourth decades of life. Polycythemia can occur in 10 to 20% of patients. Eighty percent are sporadic, and 20% can be associated with von Hippel-Lindau syndrome. The latter is characterized by an autosomal dominant heritance pattern; multiple hemangioblastomas; and other systemic tumors such as renal cysts, renal cell carcinomas, pheochromocytomas, and retinal angiomatoses.[1]

The advanced age of our patient is noteworthy. This case report demonstrates that a hemangioblastoma can be a rare cause of a primary intraaxial tumor in the cerebellum in elderly individuals. Given the classic appearance of these lesions on MR imaging and angiography and the absence of any primary source of tumor, the diagnosis of hemangioblastoma was correctly suspected preoperatively. Because the patient was symptomatic from both mass effect and cerebellar signs, we believed that surgical resection was a reasonable option in this otherwise healthy octogenarian.[2,10]

References

1. Conway JE, Chou D, Clatterbuck RE, et al: Hemangioblastomas of the central nervous system in von Hippel-Lindau syndrome and sporadic disease. **Neurosurgery 48:**55-62, 2001
2. Gnanalingham KK, Apostolopoulos V, Chopra I, et al: Haemangioblastoma: A rare cause of a cerebellar mass in the elderly. **Br J Neurosurg 17:**461-464, 2003
3. Ho VB, Smirniotopoulos JG, Murphy FM, et al: Radiologic-pathologic correlation: Hemangioblastoma. **AJNR Am J Neuroradiol 13:**1343-1352, 1992
4. Ishizawa K, Komori T, Hirose T: Stromal cells in hemangioblastoma: Neuroectodermal differentiation and morphological similarities to ependymoma. **Pathol Int 55:**377-385, 2005
5. Park YS, Chang JH, Chang JW, et al: Gamma knife surgery for multiple hemangioblastomas. **J Neurosurg 102 Suppl:**97-101, 2005
6. Prieto R, Roda JM: Hemangioblastoma of the lateral ventricle: Case report and review of the literature. **Neurocirugia (Astur) 16:**58-62, 2005
7. Rosenthal G, Israel Z, Umansky F: Metastatic brain adenocarcinoma masquerading as recurrent haemangioblastoma. **Acta Neurochir (Wien) 140:**1207-1208, 1998
8. Tago M, Terahara A, Shin M, et al: Gamma knife surgery for hemangioblastomas. **J Neurosurg 102 Suppl:**171-174, 2005
9. Takeuchi S, Tanaka R, Fujii Y, et al: Surgical treatment of hemangioblastomas with presurgical endovascular embolization. **Neurol Med Chir (Tokyo) 41:**246-251, 2001
10. Van Velthoven V, Reinacher PC, Klisch J, et al: Treatment of intramedullary hemangioblastomas, with special attention to von Hippel-Lindau disease. **Neurosurgery 53:**1306-1313, 2003
11. Yong RL, Kavanagh EC, Fenton D, et al: Midline cerebellar medulloblastoma in a seventy-one-year old patient. **Can J Neurol Sci 33:**101-104, 2006
12. Zhou LF, Du G, Mao Y, et al: Diagnosis and surgical treatment of brainstem hemangioblastomas. **Surg Neurol 63:**307-315, 2005