An intracranial aneurysm on the feeding artery of a cerebellar hemangioblastoma

Case report

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A number of investigators have described the association of aneurysms and cerebral lesions such as tumors\textsuperscript{13,23} or vascular malformations\textsuperscript{4,8,10,20} in the same patient. Different theories about the causes, pathophysiological mechanisms, and treatment modalities of these coexistent pathological entities have been suggested.\textsuperscript{2} To the best of our knowledge, however, only two cases of coexisting aneurysm and cerebellar hemangioblastoma—other than the one presented here—have been reported in the literature\textsuperscript{18,25} and neither report describes an aneurysm located on the main feeding artery.

In the present article, we report on a patient with an aneurysm of the distal anterior inferior cerebellar artery (AICA), which was the main feeding vessel of the hemangioblastoma. Successful total excision of the hemangioblastoma and clipping of the AICA aneurysm achieved in a one-stage operation was demonstrated on postoperative angiography.

Key Words • cerebellar hemangioblastoma • aneurysm • angiography

A case of cerebellar hemangioblastoma with a coexistent arterial aneurysm on the feeding artery of the tumor is reported. The patient presented with an acute onset of headache, loss of consciousness, and left-sided hemiparesis due to a posterior fossa hemorrhage found adjacent to a hemangioblastoma. Four-vessel angiography revealed an aneurysm on the anterior inferior cerebellar artery (AICA), which was the main feeding vessel of the hemangioblastoma. Successful total excision of the hemangioblastoma and clipping of the AICA aneurysm achieved in a one-stage operation was demonstrated on postoperative angiography.

Case Report

History. This 53-year-old right-handed man maintained a good state of health until 8 months prior to admission, at which time he noted headaches, vertigo, and occasional losses of balance.

Examination. On the day of admission, the patient experienced a sudden onset of headache with loss of consciousness and left-sided hemiparesis. He was referred to another hospital where computerized tomography (CT) scanning revealed a left cerebellar hemorrhage with significant surrounding edema. The patient was given Solu-Medrol and transferred to the Harborview Medical Center, where additional CT and magnetic resonance (MR) imaging were performed. The radiological examinations revealed an intensely enhancing, cystic, left posterior fossa mass, with an adjacent hemorrhage just lateral to the fourth ventricle, as well as obstructive hydrocephalus (Fig. 1). Four-vessel cerebral angiography confirmed the presence of a 3-cm-wide area of tumor neovascularity in the left cerebellar hemisphere that was consistent with a hemangioblastoma and was predominantly supplied by the left AICA (Fig. 2A). Proximal to the hemangioblastoma was a 5-mm AICA aneurysm that appeared to be the likely cause of the posterior fossa hemorrhage. Based on the tortuosity of the AICA, the location of the aneurysm, and the blood supply to the tumor, embolization could not be performed.

Operation. On the day of admission the patient was taken to the operating room for acute treatment of the obstructive hydrocephalus. A ventriculostomy that resulted in initial stabilization was performed first. Subsequently, the patient underwent a left suboccipital craniectomy with removal of the tumor. Following resection of the tumor, the intracerebral hemorrhage, which was located in the posterior fossa, was unroofed and old clotted blood emulated under pressure. This was accompanied by sudden onset of bleeding. The intraoperative rupture of the aneurysm was controlled by placement of three straight clips. Frozen sections of a tumor biopsy specimen were used to confirm the diagnosis of a hemangioblastoma (Fig. 3).

Postoperative Course. Postoperative cerebral angiography demonstrated no residual tumor blush or drainage veins within the left side of the posterior fossa and no evidence of residual aneurysm or vasospasm of the left AICA (Fig. 2B).
Coexistence of intracranial aneurysm and hemangioblastoma

**Discussion**

Hemangioblastomas are tumors of the central nervous system (CNS) that account for approximately 1 to 2% of all intracranial tumors and 7 to 12% of all infratentorial tumors. They can develop in any portion of the CNS, but the most common site is the cerebellum. Approximately 75% of hemangioblastomas are found as solitary lesions, whereas 25% are a manifestation of the hereditary von Hippel–Lindau disease. Hemangioblastomas are characterized by a dense capillary network separated by stromal cells and by a high frequency of cyst formation. When cystic, the solid tumor is relatively small and is incorporated at some point in the cyst wall as a mural nodule. As a result of these cystic lesions' slow expansion to a large size, obstructive hydrocephalus commonly occurs. Histologically, hemangioblastomas consist of three cell types: vascular endothelial cells that form the capillary network, pericytes adjacent to the endothelial cells, and epithelioid or stromal cells that are often laden with lipids and may give the tumor a yellow appearance on gross sections.

The association of an intracranial aneurysm and a posterior fossa hemangioblastoma is a rare occurrence. To find a comparative pathophysiological situation, we looked at the association of cerebral aneurysm and arteriovenous malformation (AVM), which has been reported to occur in approximately 10% of AVM cases. Three principal theories have been proposed to explain the pathogenesis of coexisting AVMs and aneurysms: 1) an augmented flow in the feeding vessel of the AVM; 2) a common congenital defect of the vessel walls; and 3) coincidental occurrence. Similar to that observed with AVMs, the feeding arteries of hemangioblastomas show high flow velocity because of missing capillary bed resistance. This can be seen by the early appearance of tumor blush on angiography. The high flow velocity in the feeding artery of the hemangioblastoma could contribute to aneurysm formation, as proposed in the first theory regarding AVMs. Vascular endothelial growth factor (VEGF), also known as vascular permeability factor, is a potent polypeptide regulator of blood vessel function that acts on two endo-

![Fig. 1. A: Preoperative coronal T1-weighted MR image revealing a cystic posterior fossa lesion (arrow) and dilated lateral ventricle. B: Contrast-enhanced CT scan demonstrating an intensely enhancing, cystic left posterior fossa mass (arrow), with an adjacent acute hemorrhage (arrowheads).](image1)

![Fig. 2. Lateral vertebral artery angiograms. A: Preoperative angiogram revealing a large 3-cm-wide area of intense tumor blush in the left cerebellar hemisphere (large arrow), which is predominantly supplied by the left AICA (open arrowhead) with some supply from the left posterior inferior cerebellar artery (closed arrowhead). A 5-mm-wide aneurysm on the AICA is identified projecting posteriorly and inferiorly toward the tumor (small arrow). B: Postoperative angiogram revealing a good blood supply to the posterior fossa and no evidence of residual AICA aneurysm or vasospasm. The three straight aneurysm clips can be identified (large arrow).](image2)
endothelial junctions and the secondary activation of pathways thought to initiate changes favoring leakage at intercellular membrane barriers. Vascular endothelial growth factor is expressed in vessel walls, whereas no expression of VEGF was found in non-aneurysmal cerebral vessels. In our case, the hemorrhage was most likely due to aneurysm rupture.

References


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