Cerebellar Hemangioblastoma Manifesting as Hearing Disturbance
—Case Report—

Toshiyuki AMANO, So TOKUNAGA, Tadahisa SHONO, Masahiro MIZOGUCHI, Kenichi MATSUMOTO, Fumiaki YOSHIDA, and Tomio SASAKI

Department of Neurosurgery, Graduate School of Medical Sciences, Kyushu University, Fukuoka

Abstract

A 49-year-old man presented with a rare case of cerebellar hemangioblastoma manifesting as only hearing disturbance. He had suffered from hearing difficulty in the right ear for a few months. Magnetic resonance imaging revealed a cystic mass lesion with an internal fluid level and surrounding flow voids in the right cerebellopontine (CP) angle. Cerebral angiography disclosed a vascular-rich tumor fed by both the superior cerebellar and anterior inferior cerebellar arteries. En bloc resection of the tumor was planned under a preoperative diagnosis of cerebellar hemangioblastoma. The tumor protruded into the CP cistern and compressed cranial nerve VIII. The feeding arteries were meticulously coagulated and the tumor was successfully removed. The histological diagnosis was hemangioblastoma. After the operation, the patient’s hearing acuity improved dramatically. Cerebellar hemangioblastoma should be considered in the differential diagnosis of CP angle tumors associated with hearing disturbance.

Key words: cerebellar hemangioblastoma, acoustic tumor, cerebral angiography, hearing disturbance, magnetic resonance imaging

Introduction

The prevalence of hemangioblastomas has been estimated at approximately 1 to 2% of intracranial tumors and 7 to 8% of posterior fossa tumors.1,4,5) Hemangioblastomas most commonly arise from the cerebellum, followed by the spinal cord and brainstem,1) and usually manifest as headache or nausea, associated with elevated intracranial pressure (ICP) or cerebellar ataxia.2,7) Hearing disturbance is a common symptom of acoustic tumors or meningiomas expanding in the cerebellopontine (CP) cistern, but is a rare clinical manifestation of hemangioblastomas, especially as an initial symptom. We report a rare case of cerebellar hemangioblastoma manifesting as only hearing disturbance.

Case Report

A 49-year-old male suffered hearing difficulty in the right ear for a few months. Neurological examination on admission revealed no signs of cerebellar ataxia or elevated ICP, such as headache, nystagmus, hypotonia, tremor, dysmetria, wide-based gait, and scanning speech. Neither the heel-shin test nor the nose-finger-nose test showed signs of cerebellar dysfunction. Neuro-otological examination demonstrated marked increase of pure tone average (96.7 dB; Fig. 1A) and absence of the second to fifth waves of the auditory evoked brainstem responses (ABR) on the right side (Fig. 1B).
Computed tomography (CT) with contrast medium showed a ring-like enhanced mass lesion in the right CP angle (Fig. 2A, B). Bone window CT detected no enlargement of the internal auditory canal (Fig. 2C). T₁-weighted magnetic resonance (MR) imaging showed the lesion as isointense with ring-like enhancement after intravenous administration of gadolinium (Fig. 3A, B). T₂-weighted MR imaging disclosed a fluid level in the cystic lesion and surrounding flow voids, a feature of vascular-rich tumors such as hemangioblastoma, which can be associated with intratumoral hemorrhage (Fig. 3C). Cerebral angiography was thus performed and showed the tumor was fed by both the superior cerebellar artery and anterior inferior cerebellar artery (Fig. 4). Based on these observations, the preoperative diagnosis was cerebellar hemangioblastoma.

Surgical resection of the tumor was performed through the right suboccipital retrosigmoid approach. The tumor was covered with thinned cerebellar cortex, protruded cerebellar artery (Fig. 4). The tumor was removed completely. The pure tone average audiogram showing the successful hearing acuity recovery of the right ear is shown in Fig. 7.
into the CP cistern, and compressed cranial nerve VIII (Fig. 5). Minimal retraction of the right cerebellar hemisphere exposed the surface of the tumor and associated vessels, and the feeding arteries were cauterized and sharply transected. After completion of circumferential dissection of the tumor, the draining veins were transected and en bloc resection of the tumor was performed. Histological examination revealed tumor cells with clear, vacuolated, or eosinophilic cytoplasm proliferating diffusely or in nests, associated with massive hemorrhage and capillary vessels (Fig. 6). The histological diagnosis was hemangioblastoma.

Postoperatively his hearing acuity successfully recovered with a pure tone average of 31.7 dB (Fig. 7) and speech discrimination score of 100%. Postoperative ABR on the right side revealed that the fifth wave was reproducible.

Discussion

The most common clinical symptom of cerebellar hemangioblastoma is headache or nausea associated with elevated ICP found in 85% of reported cases. However, hearing disturbance can occur if the tumor involves the cochlear nucleus in the brainstem or compresses cranial nerve VIII in the cistern. The incidence of hearing disturbance in patients with cerebellar hemangioblastoma is estimated to be less than 5%.\(^2\) In our patient, the cranial nerve VIII was compressed by the tumor in the CP cistern, and hearing acuity recovered dramatically after total removal of the tumor. These observations clearly indicated that our patient’s symptoms were caused by the cerebellar hemangioblastoma protruding into the CP cistern.

In contrast, hearing disturbance is the most common initial symptom of extra-axial CP angle tumors such as acoustic tumors and meningiomas. CT and MR imaging usually provide useful information for the differential diagnosis of these tumors, such as enlargement of the internal auditory canal or dural attachment, neither of which was evident in our patient. Conversely, intratumoral hemorrhage is rare in acoustic tumors or meningiomas.\(^9\) We found a fluid level within the cystic lesion in our patient, consistent with previous intratumoral hemorrhage, which is one of the reported radiological features of cerebellar hemangioblastoma.\(^3\) In our opinion, the most important differential diagnosis of hemangioblastoma arising near the cerebellar flocculus is medial-type acoustic tumor,\(^4\) which usually causes no enlargement of the internal auditory canal and may be vascular-rich.

Patients with acoustic tumors and meningiomas often do not undergo preoperative cerebral angiography because of recent advances in CT and MR imaging. However, cerebral angiography can provide invaluable information, such as identification of feeding arteries and draining veins. The differential diagnosis of hemangioblastomas from acoustic tumors, especially medial-type tumors, and meningiomas is clinically important, because the surgical strategies for these tumors are quite different. Internal decompression is usually performed during surgery for acoustic tumors and meningiomas, but this strategy is potentially very dangerous in highly vascularized hemangioblastomas. Removal of hemangioblastomas must be performed by en bloc resection after interruption of the feeding arteries and draining veins. Angiographic information allows us to plan safer surgery. In addition, preoperative embolization of the feeding arteries can be performed if required.

The present rare case of hemangioblastoma manifesting as only hearing disturbance was caused by compression of cranial nerve VIII by the tumor in the CP cistern. CT and MR imaging are helpful for the diagnosis of cerebellar hemangioblastomas, but cerebral angiography is essential for confirmation of the preoperative diagnosis,\(^5\) and is also very useful and necessary to plan a safe surgical strategy. Therefore, we strongly recommend preoperative cerebral angiography in patients with CP angle tumors if cerebellar hemangioblastoma is included in the differential diagnosis.

References


Address reprint requests to: Toshiyuki Amano, M.D., Department of Neurosurgery, Graduate School of Medical Sciences, Kyushu University, 3–1–1 Maidashi, Higashi-ku, Fukuoka 812–8582, Japan.

E-mail: amano@ns.med.kyushu-u.ac.jp