

## Pial brainstem artery arteriovenous malformation with flow-related intracanalicular aneurysm masquerading as vestibular schwannoma: illustrative case

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**BACKGROUND** Lesions of the internal auditory canal presenting with partial hearing loss are almost always vestibular schwannomas (VSs). Intracanalicular anterior inferior cerebellar artery (AICA) aneurysms are extremely rare but can mimic VS based on symptoms and imaging. The authors report the case of a flow-related intracanalicular AICA aneurysm from a pial brainstem arteriovenous malformation (AVM) masquerading as VS.

**OBSERVATIONS** A 57-year-old male with partial left-sided hearing loss and an intracanalicular enhancing lesion was initially diagnosed with VS and managed conservatively at an outside institution with surveillance imaging over 3 years. When he was referred for VS follow-up, new imaging raised radiological suspicion for vascular pathology. Cerebral angiography revealed a small pial AVM located at the trigeminal root entry zone with an associated flow-related intracanalicular AICA aneurysm. The AVM was obliterated with open surgery, during which intraoperative angiography confirmed no AVM filling, preservation of the AICA, and no further aneurysm filling.

**LESSONS** Intracanalicular AICA aneurysms and other lesions, including cavernous malformations, can mimic radiographic features of VS and present with hearing loss or facial weakness. Modern vascular neurosurgical techniques such as endovascular intervention and open surgery in a hybrid operating room allowed definitive management of both lesions without untoward morbidity.

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**KEYWORDS** arteriovenous malformation; intracanalicular aneurysm; anterior inferior cerebellar artery; vestibular schwannoma; vascular neurosurgery

Vestibular schwannomas (VSs) are a pathology that is familiar to the practicing neurosurgeon. More than 3,000 cases of VS are diagnosed annually in the United States. With an incidence of about 1 per 100,000,<sup>1</sup> VS accounts for 8% of all intracranial tumors.<sup>2</sup> Ninety percent of patients with VS present with unilateral hearing loss,<sup>2</sup> and, in the absence of worsening symptoms, these benign tumors are often managed with yearly surveillance magnetic resonance imaging (MRI) or, when indicated, interventions including radiosurgery and surgical resection.

In contrast, an extremely rare cause of an internal auditory canal (IAC) lesion causing unilateral hearing loss is an intracanalicular aneurysm arising from the anterior inferior cerebellar artery (AICA). The

ubiquity of VS and its easily recognizable radiographic appearance may lead to delayed diagnosis of intracanalicular aneurysms. A missed diagnosis of an intracanalicular AICA aneurysm can have devastating consequences because almost all of these reported in the literature presented in the setting of subarachnoid hemorrhage (SAH).<sup>3</sup>

Here, we present a case of a flow-related intracanalicular AICA aneurysm associated with a pial brainstem arteriovenous malformation (AVM) located at the trigeminal root entry zone. The aneurysm was initially managed conservatively as a VS. We describe the initial imaging that led to this diagnosis, subsequent superselective angiography, and the ultimate definitive surgical management of both lesions.

**ABBREVIATIONS** AICA = anterior inferior cerebellar artery; AV = arteriovenous; AVM = arteriovenous malformation; CT = computed tomography; IAC = internal auditory canal; MR = magnetic resonance; MRI = magnetic resonance imaging; SAH = subarachnoid hemorrhage; VS = vestibular schwannoma.

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## Illustrative Case

### Background

A 57-year-old male with a 3-year history of presumed left-sided VS and left-sided partial hearing loss was referred for evaluation after follow-up MRI raised the question of a vascular lesion instead of a VS within the IAC (Fig. 1). The patient's VS had been managed conservatively with annual surveillance MRI with and without contrast. On his latest MRI, a magnetic resonance (MR) angiography sequence was performed, the results of which suggested a vascular lesion within the left IAC as well as a small AVM supplied by the distal AICA. A subsequent computed tomography (CT) angiogram revealed a 3 × 5-mm lesion within the left IAC, which was inseparable from the course of the AICA within the IAC, and a small AVM located at the level of the trigeminal root entry zone (Fig. 2). Due to its lobulated appearance on MRI, the IAC lesion was still favored to represent a VS, with a loop of the AICA coursing around the mass. However, an aneurysmal outpouching arising from the AICA could not be excluded.

### Diagnosis

The patient underwent diagnostic and superselective cerebral angiography, which confirmed the presence of arteriovenous (AV) shunting from a small pial AVM supplied by branches of the distal left AICA at the level of the trigeminal entry zone (Fig. 3). The AVM was noted to drain superficially along the left cerebellar hemisphere through cerebellar and mesencephalic veins toward the level of the torcula, with an associated flow-related aneurysm in the left IAC. The patient subsequently underwent superselective angiography with a plan for possible embolization of the AVM, but no intervention was performed, given the presence of an “*en passage*” distal left AICA branch supplying the left lateral aspect of the pons. Thus, the decision was made to proceed with open surgery to obliterate the AVM and the aneurysm.

### Surgical Management

An image-guided left-sided retrosigmoid craniotomy was performed, with the goal of disconnecting the AVM and clipping the aneurysm under direct visualization of the operating microscope. The dura was opened over the left lateral cerebellum, and the cisterna magna was opened to relax the brain. The arachnoid was opened over the seventh and eighth nerve complex, and a large AICA branch was seen coming out of the IAC, coursing toward the lateral brainstem at the level of the trigeminal root entry zone. Immediately apparent were dilated venous structures and small branches arising from the AICA shunting toward the small AVM.

These were cauterized and cut, disconnecting the AVM from its supply. Last, the local draining vein draining the AVM was then cauterized. Indocyanine green video angiography was performed, which showed good flow in the AICA vessel and no evidence of AV shunting to suggest residual malformation or fistula. Next, intraoperative diagnostic cerebral angiography was performed, which confirmed obliteration of the AVM. Interestingly, the intracanalicular aneurysm dome was no longer visualized; only the loop of the AICA within the IAC was seen (Fig. 4). Thus, further exposure and clipping were not performed. The patient was discharged 2 days later with no deficits, apart from baseline left-sided hearing loss, which remained unchanged from before surgery.

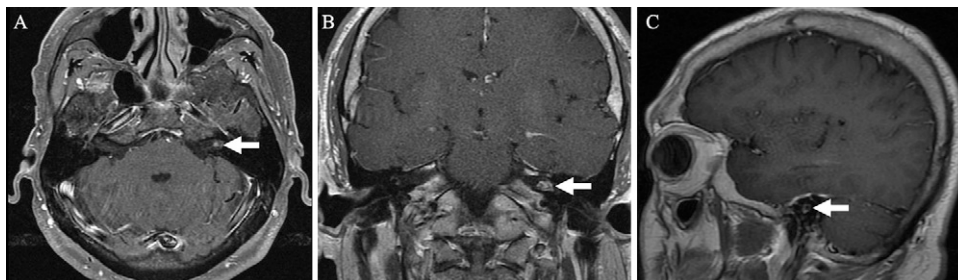
## Discussion

### Observations

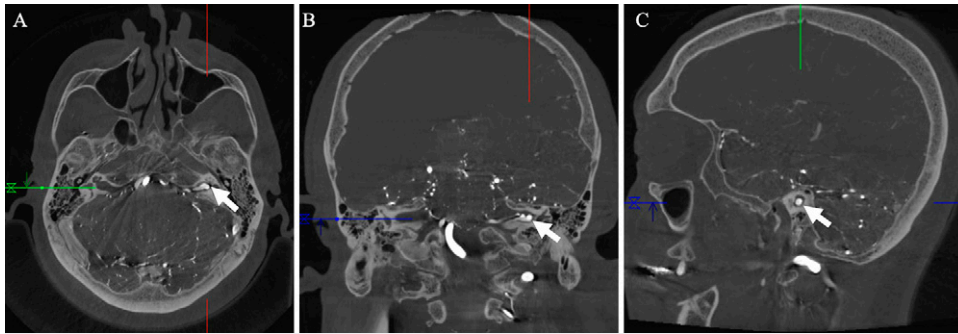
The patient's presenting symptom of partial hearing loss with an IAC lesion on MRI is commonly associated with VS. This clinical picture, combined with a mostly standard appearance on imaging, led to delay in diagnosis of the true pathology of AVM and flow-related intracanalicular AICA aneurysm. Only at 3-year maintenance imaging was a vascular lesion suspected, and subsequent cerebral angiography revealed a pial brainstem AVM with an associated flow-related intracanalicular AICA aneurysm. Obliteration of the AVM was sufficient to limit flow to the aneurysm and facilitate its closure. This case demonstrates a diagnostically difficult, complex vascular lesion and the efficacy of modern vascular neurosurgical management, which allowed definitive management with the full range of tools, including diagnostic and superselective angiography, embolization, and open microsurgical resection with intraoperative angiography.

Aneurysms of the AICA are rare, accounting for 0.1% to 0.5% of all intracranial aneurysms,<sup>4</sup> and an intracanalicular location is even more uncommon, with fewer than 10 cases reported in the literature.<sup>3</sup> A flow-related etiology from AV shunting is common, with a recent meta-analysis showing that 18% of patients with AVMs have associated intracranial aneurysms.<sup>5</sup> In almost all reported cases of intracanalicular AICA aneurysms, patients presented with SAH; the aneurysm and associated AVM were discovered by angiography as part of the management protocol for ruptured aneurysm.<sup>3</sup>

The natural histories of VS and intracanalicular AICA aneurysms are different, and thus accurate diagnosis can prevent worsening seventh/eighth nerve symptoms or more devastating SAH. On imaging, a T1 and T2 hypointense cerebellopontine angle lesion with postgadolinium enhancement is usually VS, but the differential diagnosis also includes the much less common meningioma or vascular lesion,<sup>6</sup> including aneurysm or cavernous malformation. Although the pretest probability of an



**FIG. 1.** Preoperative MRI. Axial (A), coronal (B), and sagittal (C) views of preoperative T1 with contrast MRI. White arrows denote location of aneurysm, initially thought to be vestibular schwannoma.

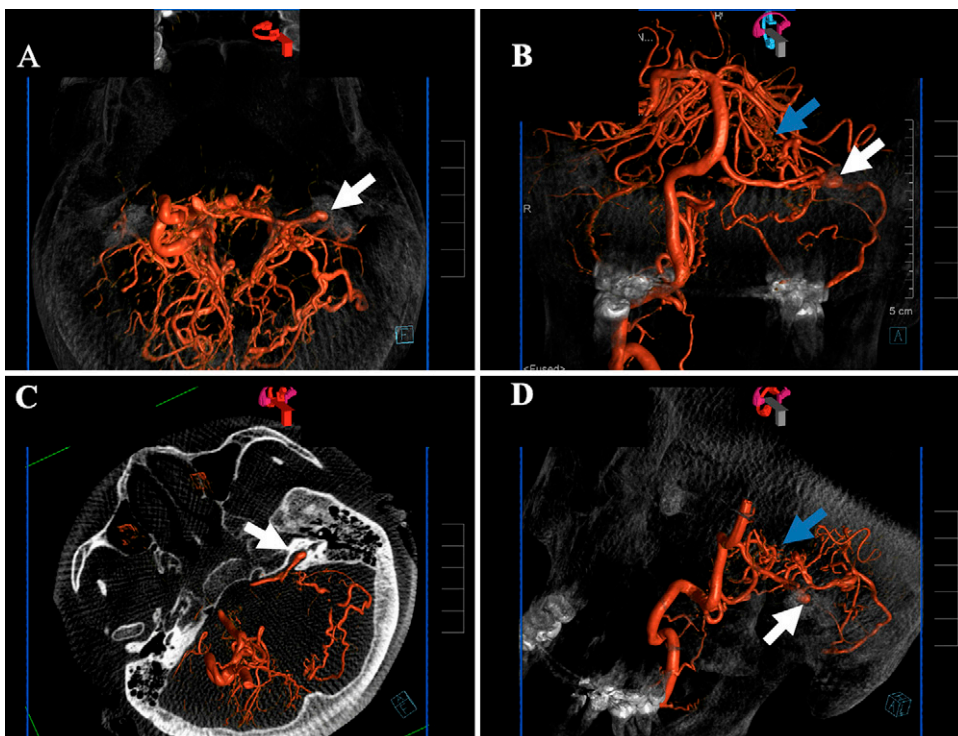


**FIG. 2.** Preoperative CT imaging. Axial (A), coronal (B), and sagittal (C) views of preoperative CT with contrast angiography. *White arrows* denote location of aneurysm, initially thought to be vestibular schwannoma.

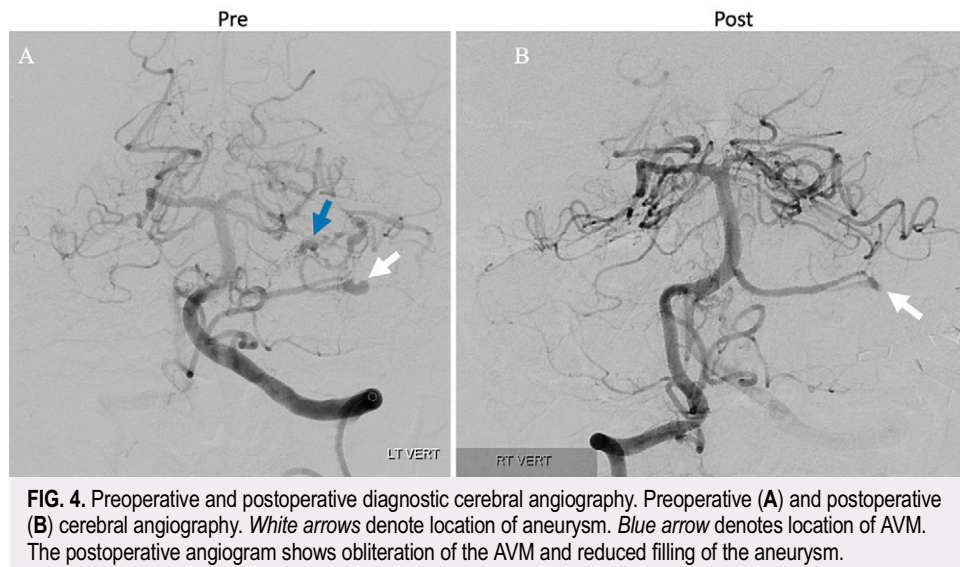
intracanalicular aneurysm is extremely low and thus not all such lesions necessitate angiography, some imaging features may offer clues that an IAC lesion is not a VS. For example, the absence of IAC remodeling (because generally only large vascular lesions or neoplasms tend to cause bony remodeling) and the “blurry dot sign” on T2 MRI may suggest a vascular lesion.<sup>7,8</sup> If a vascular lesion is suspected, the high spatial resolution of CT angiography can be used to examine the anatomical relationship of the AICA, IAC bone, and lesion.<sup>6</sup> In our case, we used MR angiography and CT angiography to evaluate for the presence of a vascular lesion, which led to definitive diagnosis with cerebral angiography.

Modern vascular neurosurgery is well equipped to manage this case of an AVM with an associated flow-related intracanalicular AICA aneurysm with minimal morbidity. We initially pursued embolization as

part of the diagnostic and superselective angiogram, which has previously been reported as definitive management if the collateral vasculature to the lateral pons is favorable.<sup>9,10</sup> Given the presence of an *en passage* distal AICA, endovascular treatment of the AVM was not possible. Therefore, direct microsurgical obliteration of the AVM was pursued as the treatment strategy. Some surgical reports have recommended additional bony exposure with petrosectomy to visualize and potentially clip the aneurysm. For example, a similar previous case in 1989 required direct visualization with a transcochlear approach, which sacrificed hearing.<sup>11</sup> Notably, in this previous case, the lesion was presumed to be VS until surgical exposure, and the aneurysm was not clipped due to lack of distal control. In contrast, our patient’s AVM and aneurysm were both completely obliterated through retrosigmoid craniotomy, with no morbidity aside from baseline partial hearing loss.



**FIG. 3.** A–D: Preoperative diagnostic cerebral angiography. Three-dimensional reconstructions of diagnostic cerebral angiography. *White arrows* denote location of aneurysm. *Blue arrows* denote location of AVM.



**FIG. 4.** Preoperative and postoperative diagnostic cerebral angiography. Preoperative (A) and postoperative (B) cerebral angiography. White arrows denote location of aneurysm. Blue arrow denotes location of AVM. The postoperative angiogram shows obliteration of the AVM and reduced filling of the aneurysm.

## Lessons

Intracanalicular AICA aneurysms are rare but may mimic the much more common VS on work-up in terms of symptoms, location, and imaging features. Careful inspection of the course of the AICA in the IAC, of the IAC bony anatomy, and for the “blurry dot sign” (for example, with MR angiography and CT angiography) is crucial, and diagnostic cerebral angiography is the definitive test if a vascular lesion is suspected. Because patients with intracanalicular AICA aneurysms may present with SAH due to rupture, early diagnosis and definitive treatment can prevent morbidity and mortality.

Embolization and open surgery with intraoperative angiography are both good options to treat these lesions; however, embolization may be impossible when there is risk of injury to the lateral brainstem. Intraoperative angiography in a hybrid operating room allowed confirmation of AVM obliteration and no flow to the flow-related AICA aneurysm dome, preventing the need for a larger surgical exploration of the IAC.

## References

1. Kshetry VR, Hsieh JK, Ostrom QT, Kruchko C, Barnholtz-Sloan JS. Incidence of vestibular schwannomas in the United States. *J Neurooncol*. 2015;124(2):223–228.
2. Carlson ML, Link MJ. Vestibular schwannomas. *N Engl J Med*. 2021;384(14):1335–1348.
3. Zotta DC, Stati G, Paulis DD, Galzio RJ. Intrameatal aneurysm of the anterior inferior cerebellar artery. *J Clin Neurosci*. 2011;18(4):561–563.
4. Pritz MB. Aneurysms of the anterior inferior cerebellar artery. *Acta Neurochir (Wien)*. 1993;120(1–2):12–19.
5. Gross BA, Du R. Natural history of cerebral arteriovenous malformations: a meta-analysis. *J Neurosurg*. 2013;118(2):437–443.
6. Jayaraman MV, Mayo-Smith WW, Doberstein CE. Intracanalicular aneurysm of the anterior inferior cerebellar artery revealed by multi-detector CT angiography. *AJNR Am J Neuroradiol*. 2003;24(7):1338–1340.
7. Dalley RW, Robertson WD, Nugent RA, Durity FA. Computed tomography of anterior inferior cerebellar artery aneurysm mimicking an acoustic neuroma. *J Comput Assist Tomogr*. 1986;10(5):881–884.
8. Marchini AK, Mosimann PJ, Guichard JP, Boukobza M, Houdart E. Anterior inferior cerebellar artery aneurysms mimicking vestibular schwannomas. *J Neuroimaging*. 2014;24(4):404–406.
9. Goto S, Ohshima T, Yamamoto T, et al. A case of ruptured intrameatal aneurysm successfully treated with coil embolization. *Nagoya J Med Sci*. 2015;77(1–2):307–313.
10. Suzuki K, Meguro K, Wada M, Fujita K, Nose T. Embolization of a ruptured aneurysm of the distal anterior inferior cerebellar artery: case report and review of the literature. *Surg Neurol*. 1999;51(5):509–512.
11. Gleeson MJ, Cox TCS, Strong AJ. Aneurysm of the anterior inferior cerebellar artery mimicking an intracanalicular acoustic neuroma. *J Laryngol Otol*. 1989;103(1):107–110.

## Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

## Author Contributions

Conception and design: Liu, Kurland, Ali, Golfinos. Acquisition of data: Riina, Liu, Kurland, Ali, Golfinos. Analysis and interpretation of data: Riina, Liu, Kurland, Golfinos. Drafting the article: Riina, Liu, Kurland, Ali. Critically revising the article: Riina, Liu, Kurland, Ali, Nossek. Reviewed submitted version of manuscript: Liu, Kurland, Golfinos, Nossek. Administrative/technical/material support: Golfinos.

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