

## CASE REPORT

# Distal Anterior Inferior Cerebellar Artery Aneurysm Masquerading as a Cerebellopontine Angle Tumor: Case Report and Review of Literature

Atom Sarkar, M.D., Ph.D.<sup>1</sup> and Michael J. Link, M.D.<sup>1</sup>

## ABSTRACT

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We present the case of a distal anterior inferior cerebellar artery (AICA) aneurysm masquerading as a cerebellopontine angle tumor in a 60-year-old right-handed man with previously undiagnosed polyarteritis nodosa (PAN). The patient presented with a 2-month history of progressive right-sided hearing loss, intermittent severe headache, and sudden onset of complete facial paralysis 3 weeks before admission. Magnetic resonance imaging, including postgadolinium images, showed a 1.2-cm heterogeneously enhancing mass that slightly enlarged the right internal auditory canal. A right suboccipital craniotomy was performed, and a partially thrombosed fusiform AICA aneurysm was discovered just anterior to the VII/VIII nerve complex. The aneurysm was trapped and opened, and a thrombectomy was performed. Postoperatively, the patient experienced abdominal pain; liver function tests were abnormal. Investigation revealed a small retroperitoneal hemorrhage and aneurysms of the celiac axis and gastroduodenal arteries. Further investigation revealed an increased erythrocyte sedimentation rate, and a diagnosis of PAN was made. PAN is a well-identified factor in the genesis of peripheral vascular aneurysms. Aneurysms involving the hepatic, renal, coronary, pancreatic, and tibial arteries have been described. PAN is an extremely rare cause of intracranial aneurysm. Patients who present with aneurysms in unusual locations (e.g., distal AICA) should be investigated for vasculopathy and collagen vascular disorders.

**KEYWORDS:** Anterior inferior cerebellar artery, aneurysm, peripheral vascular aneurysms, polyarteritis nodosa

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*Skull Base*, volume 14, number 2, 2004. Address for correspondence and reprint requests: Michael J. Link, M.D., Department of Neurologic Surgery, Mayo Clinic, 200 First Street SW, Rochester, MN 55905. E-mail: link.michael@mayo.edu. <sup>1</sup>Department of Neurologic Surgery, Mayo Clinic, Rochester, Minnesota. Copyright © 2004 by Thieme Medical Publishers, Inc., 333 Seventh Avenue, New York, NY 10001, USA. Tel: +1(212) 584-4662. 1531-5010,p;2004,14,02,101,107,ftx,en;sbs00391x.

Approximately 15% of intracranial aneurysms occur in the posterior circulation.<sup>1-3</sup> Most arise near the bifurcation of the basilar, vertebral, or posterior inferior cerebellar arteries. Aneurysms of the distal anterior inferior cerebellar artery (AICA) are relatively uncommon, representing about 0.1% to 0.5% of all intracranial aneurysms.<sup>2</sup>

Since the initial report by Schwartz in 1948,<sup>4</sup> only 61 additional patients underwent documented surgical/endovascular intervention for AICA aneurysms.<sup>5-8</sup> Almost all of these aneurysms were located at the basilar-AICA junction. The most common presenting feature of these lesions is subarachnoid hemorrhage. Cranial nerve dysfunction involving the trigeminal through hypoglossal nerves also has been reported. This variability depends on the anatomic course of the AICA; cranial nerves VII and VIII are most commonly involved.<sup>1,2,9-12</sup> Women are more often affected than men by a ratio of 2:1.<sup>2</sup>

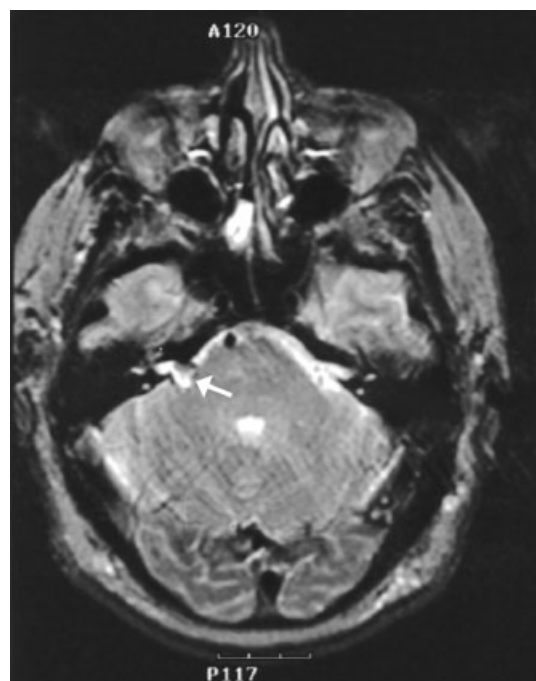
## CASE REPORT

A 60-year-old, right-handed man presented with a 1-year history of decreased hearing. An audiogram revealed right-sided high-frequency sensorineural hearing loss. One month before presentation, the patient had reported difficulty in closing his right eye. The problem had occurred abruptly, and it progressed to complete right facial paralysis in 1 week. Soon thereafter, the patient experienced significant headaches in the right retromastoid-occipital area that were poorly relieved with opioids. One week before neurosurgical evaluation, he noted a marked decrease in the hearing in his right ear. Magnetic resonance imaging (MRI) performed elsewhere (Figs. 1 to 3) revealed a small enhancing mass (10 × 8 × 10 mm) in the right cerebellopontine angle (CPA) cistern, centered on the right internal auditory canal (IAC), with some extension into the IAC. Although the location and the imaging characteristics suggested a vestibular schwannoma, other diagnostic considerations in-

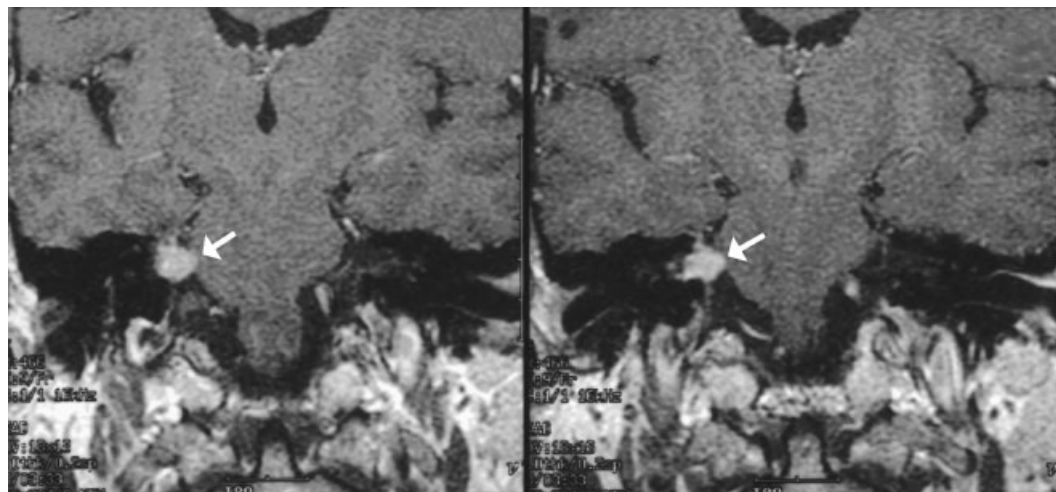
cluded a meningioma, metastasis, endolymphatic sac tumor, or vascular lesion. The patient was referred to our institution for further evaluation and treatment.



**Figure 1** Coronal T1-weighted magnetic resonance image without contrast shows a right CPA lesion (arrow). CPA, cerebellopontine angle.



**Figure 2** Axial T2-weighted magnetic resonance image shows heterogeneous signal intensity from the lesion in the right CPA (arrow). Flow voids are not evident within the lesion. CPA, cerebellopontine angle.



**Figure 3** Coronal T1-weighted magnetic resonance images with contrast show prominent contrast enhancement of the right CPA lesion (arrows). CPA, cerebellopontine angle.

The patient's neurologic examination was significant for a complete palsy of the right seventh cranial nerve (House-Brackmann grade VI),<sup>13</sup> right-ear deafness, and a few beats of nystagmus with rightward gaze. The acute-subacute onset of symptoms swayed our impression that either a tumor with hemorrhage or an aneurysmal thrombosis rapidly increased the size of the lesion, causing our patient's symptoms. The imaging was most suspicious for a tumor involving the right CPA and IAC with low signal intensity on short TR-weighted images, high signal intensity on long TR-weighted images, and rather uniform enhancement after intravenous gadolinium. Considering the patient's aggressive clinical course with pain and multiple cranial nerve palsies, we were most suspicious of a malignant tumor such as a metastasis to the petrous dura and IAC.

A right retrosigmoid craniotomy was performed the day after we examined the patient. Exploration of the right CPA revealed an aneurysm with a thin dome projecting superiorly and anteriorly from the level of the seventh and eighth cranial nerve complex. Both the facial and the vestibular nerves were stretched about the dorsal aspect of the aneurysm. The afferent and efferent aneurysm vessels were identified as the distal AICA. Aneurysm clips were applied close to the dilated

aneurysmal segment of the AICA. The aneurysm was opened, and a thick laminated clot was removed. We considered end-to-end reanastomosis of the AICA but ultimately decided against such a reconstruction because of technical constraints. The patient tolerated the procedure well, but the dysfunction of the right seventh and eighth nerve complex was unchanged. He also had increased ataxia and nystagmus. Follow-up MRI showed a small stroke in the region of the right middle cerebellar peduncle.

The patient was progressing well during inpatient rehabilitation when he experienced abdominal pain and myalgia. A serum laboratory evaluation was significant for an increased erythrocyte sedimentation rate and increased levels of creatinine and C-reactive protein. A computed tomography scan of the abdomen showed localized retroperitoneal hemorrhage. An MRI/magnetic resonance angiogram showed a splenic infarct, celiac and gastroduodenal artery aneurysms, and segmental narrowing of the splenic and hepatic arteries, all radiographically suggestive of systemic vasculitis. The constellation of symptoms, laboratory findings, and imaging were consistent with a diagnosis of polyarteritis nodosa (PAN). The patient was treated with corticosteroids and cyclophosphamide and had no further systemic or neurologic symptoms. After

16 months of follow-up, he is living independently. He has mild persistent gait ataxia but does not require ambulatory aids. The seventh cranial nerve palsy has improved (House-Brackmann grade III). He has been only intermittently compliant with treatment for the PAN, but he has had no further clinical sequelae.

## DISCUSSION

The anatomy of the AICA is highly variable.<sup>14,15</sup> Typically, the artery arises from the junction between the middle and lower thirds of the basilar artery, courses along the pons and the middle cerebellar peduncle (to which it gives a few perforating branches), then bifurcates into two major branches, called the (rostr) lateral branch and the (caudo) medial branch. The rostralateral branch courses toward the IAC close to the seventh and eighth cranial nerve complex and gives off the labyrinthine, or internal auditory, artery. It is at this point that most peripheral AICA aneurysms are located.<sup>2,11,16</sup> The caudomedial branch, in which aneurysms are seen only rarely, courses medially close to the pons, to which it sends a few perforators, and terminates in cerebellar branches. The unique variations in the specific course of the AICA likely account for the various cranial nerve palsies with which AICA aneurysms manifest.

The MRI appearance of a partially thrombosed aneurysm is complex.<sup>17</sup> The protean MRI features of such lesions make preoperative identification challenging. Flowing spins in the patent part of a partially thrombosed aneurysm can produce a hyper- or hypointense signal, depending on the flow characteristics. Typically, however, rapid flow appears as high-velocity T1- and T2-weighted signal loss. Occasionally, heterogeneous signal loss is present, depending on the specific mechanics of turbulence within the aneurysm. The signal characteristics of a thrombosed portion of an aneurysm are also complex. Its appearance on MRI depends on the particular components that constitute a

multilaminated clot—acute versus chronic thrombus, presence or absence of hemosiderin, presence or absence of calcification, and so on—and pathognomonic characterization may be precluded.<sup>18</sup>

The overall management of distal AICA aneurysms includes wrapping, proximal occlusion, trapping, intravascular embolization with coils, and direct surgical clipping.<sup>4,7,8,10,19–26</sup> In a review of the literature, no single intervention proved more successful than any other in reversing preoperative deficits or preventing postoperative deterioration in the function of cranial nerves VII and VIII. The limited available information suggests a very low incidence of ischemic complications related to trapping distal AICA aneurysms close to the IAC (particularly if seventh and eighth cranial nerve dysfunction is present preoperatively). However, the literature on distal AICA aneurysms supports the notion that location of the lesion near the IAC is a negative prognostic factor for postoperative seventh and eighth cranial nerve function.

Although cranial nerve VIII dysfunction from a vestibular schwannoma is typical, this lesion rarely manifests with facial paresis. The presence of facial weakness associated with a CPA mass should alert the clinician to an alternative diagnosis. In our patient, the VII-VIII cranial nerve dysfunction likely represented both mechanical and vascular compromise of the VII-VIII nerve complex. We believe that our patient recovered partial facial nerve function for two reasons. First, the thrombectomy performed on the trapped AICA aneurysm sac decompressed the facial nerve and relieved the mechanical insult to this structure. Second, we believe that there is an intrinsic difference in the ability of a motor nerve versus a sensory nerve to recover function after an insult. To our knowledge, our case represents the first reported reversal of a preoperative facial nerve palsy in a patient with facial nerve weakness caused by an AICA aneurysm.

This case also highlights an important aspect of clinical management. When the differential diagnosis of CPA lesions is being considered, vestibular schwannomas are by far the most common lesions. Standard treatment options for vestibular

schwannomas include microsurgical resection and stereotactic radiosurgery. Yet in this case the use of radiosurgery to a presumed tumor clearly would have been inappropriate. This case therefore emphasizes the need for surgical exploration when the radiographic or clinical diagnosis is in question.

PAN is the prototypic necrotizing vasculitis that usually affects small- to medium-size vessels in multiple systems.<sup>27</sup> Neurologic involvement ranges as widely as 15 to 65% of patients and does not usually occur until late in the course of the illness.<sup>28</sup> Peripheral neuropathy is the most frequent form of neurologic involvement. The significance of central nervous system involvement is less understood; the effects are presumably related to hypertensive or chronic vaso-occlusive changes rather than to segmental inflammation of the vessel wall.<sup>29</sup> In a review of patients with PAN observed at the Mayo Clinic, only 4 of 130 patients had central nervous system manifestations; all were strokes.<sup>30</sup> Recent reports suggest a higher incidence of central nervous system involvement, including encephalopathy, focal deficits, and seizures.<sup>28,31</sup>

The relationship between PAN and intracranial aneurysms is rarely reported.<sup>32,33</sup> Interestingly, the few reported patients with PAN and intracranial aneurysms presented either with multiple lesions or with discrete aneurysms involving the peripheral posterior circulation, as was the case with our patient. It is extremely uncommon for a patient to present with an intracranial aneurysm as the initial manifestation of PAN.

To confirm a diagnosis of PAN, other vasculitic syndromes must be ruled out.<sup>34</sup> There are no specific tests for PAN; laboratory tests merely reflect the degree of inflammation and the relative vascular impairment of specific organs. Angiographic studies and direct biopsies (skeletal muscle, sural nerve, kidney, testis, or liver) help establish the diagnosis. According to the American College of Rheumatology,<sup>35</sup> a patient must meet at least 3 of the following 10 criteria (our patient met criteria 4, 7, and 9):

1. Weight loss > 4 kg
2. Livedo reticularis

3. Testicular pain or tenderness
4. Myalgia, weakness, or leg tenderness
5. Mononeuropathy or polyneuropathy
6. Diastolic blood pressure > 90 mm Hg
7. Elevated blood urea nitrogen or creatinine
8. Hepatitis B virus
9. Arteriographic abnormality
10. Polymorphonuclear leukocytes found on biopsy of small- or medium-size arteries

## CONCLUSION

We present a case of a distal AICA aneurysm presenting as a CPA tumor in a patient with previously undiagnosed PAN. The most common treatment for distal AICA aneurysms is either clipping or trapping. Trapping is usually well tolerated; there are few reports of increased neurologic deficits. Thrombectomy or aneurysm decompression is an important aspect of the surgical approach in patients with cranial nerve palsies because it may improve postoperative cranial nerve function, as it did in our patient. The presence of a distal AICA aneurysm should raise the suspicion of underlying vasculitis.

## PAPER PRESENTED

This article was presented in poster form at the Congress of Neurosurgery meeting, San Antonio, Texas, September 2000.

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## Commentary

This article describes the rare occurrence of a distal anterior inferior cerebellar artery (AICA) aneurysm mimicking a cerebellopontine angle tumor. The authors have reviewed the subject thoroughly and have addressed several relevant issues that might arise.

As the authors state, several features of this case are atypical of an acoustic neuroma: clinical presentation with a seventh cranial nerve palsy, heterogenous signal on T1-weighted magnetic resonance images, and no enlargement of the internal auditory canal. It is easily understood why one would be swayed toward a metastatic lesion in the differential diagnosis and not consider pre-operative angiography.

Once diagnosed at surgery, it would have been interesting to know how "dominant" the AICA was in this particular patient. For example, was it an AICA-posteroinferior cerebellar artery vessel? If so, I would have hesitated before trapping the aneurysm without revascularization. It is diffi-

cult to discern whether there may have been enough redundancy in the AICA to effect the end-to-end anastomosis that the authors would have liked to have performed. The small infarct in the middle cerebellar peduncle reminds me of the desirability of revascularizing certain vascular territories (when technically feasible with a relatively low morbidity rate) when trapping is considered.

In summary, this well-documented case is yet another example of one diagnosis mimicking another and of things not always being what they seem.

Jacques J. Morcos, M.D.,  
F.R.C.S. (Eng), F.R.C.S. (Ed)<sup>1</sup>