

## Anterior Inferior cerebellar artery infarction misdiagnosed as inner ear disease

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**Key-words.** Anterior inferior cerebellar artery infarction; vertigo; sensorineural hearing loss; diffusion magnetic resonance imaging

**Abstract.** *Anterior Inferior cerebellar artery infarction misdiagnosed as inner ear disease.* **Objective:** The clinical presentation of anterior inferior cerebellar artery (AICA) infarction may mimic that of inner ear disease.

**Methodology:** This report presents two patients with cerebellar artery infarction initially misdiagnosed with inner ear disease.

**Case Report:** Both the patients presented with sudden hearing loss and vertigo. The patient in case 1 was initially diagnosed with idiopathic sudden sensorineural hearing loss. The patient in case 2 presented with 17 days of vertigo and hearing loss. Both were correctly diagnosed with AICA infarction after performing magnetic resonance imaging.

**Results and Conclusions:** We differentiated AICA from inner ear disease based on the variability in degree and frequency range of hearing loss, the duration of vertigo, and the manifestation of nystagmus. Because cases of AICA infarction and inner ear disease may present with similar symptoms, a detailed examination including clinical course assessments, laboratory findings, and neurological imaging is essential for appropriate diagnosis and treatment.

### Introduction

Anterior inferior cerebellar artery (AICA) infarction was first described by Goodhart *et al.*<sup>1</sup> in 1936 and later defined by Adams<sup>2</sup> in 1943. Over half a century later, there is still much confusion regarding its diagnosis.

The AICA supplies blood to the pons and cerebellum; its labyrinthine branch also supplies the inner ear with blood. Because the vasculature of the inner ear receives blood flow from the AICA, ischemia or infarction of this artery may result in symptoms commonly seen in inner ear disease, such as vertigo, tinnitus, and hearing loss. These symptoms of AICA infarction are not unique to that diagnosis. Vertigo, in particular, is a common presenting symptom in the emergency room, suggestive of a range of conditions.

In patients with no or few overt symptoms, AICA infarction may be extremely difficult to diagnose. A 2009 study by Kim *et al.*<sup>3</sup> found that, out

54 patients with AICA infarction, four presented with symptoms limited to hearing loss and vertigo. Diagnosis may be further obscured by early misdiagnosis, i.e., therapy for inner ear damage may mask some key indicators of AICA infarction.

In otolaryngology, hearing loss with vertigo is a common condition with many possible explanations. In addition to AICA infarction, the symptoms are consistent with sudden deafness with vertigo, which is associated with hearing loss that ranges from severe loss to complete deafness. AICA may also be confused with a diagnosis of Meniere's disease, typically presenting with mild hearing loss accompanied by dizziness and vertigo lasting half a day. The pattern of nystagmus is limited to affected side during the time of an attack, reversing directions after a few hours.

In a 2008 issue of *Laryngoscope*, Newman-Toker *et al.*<sup>4</sup> argued that differentiating central causes from peripheral causes of acute vestibulopathy is a challenging but important task. While the

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MS conceived the study and helped draft the manuscript. All authors have read and approved the final manuscript.

presentation may seem similar, there are distinct differences that can help clinicians distinguish the two. For instance, in the acute phase of static, destructive peripheral vestibular lesions, the fast phase of nystagmus is away from the side of the lesion, not towards it. In central vestibular dysfunction, the direction of nystagmus is not constant.

In the present report, we describe two cases of AICA infarction where the only presenting symptoms were vertigo and hearing loss.

## Case report

### Case 1

Case 1 was a 64-year-old man who experienced sudden onset vertigo on waking. Immediately after the onset of vertigo, he noticed a feeling of fullness in his right ear, tinnitus, and hearing loss. The man consulted an ear, nose, and throat (ENT) clinic and received intravenous drip infusion of a medicine and vitamin B12 the same day, after which blood circulation improved. However, as there was no improvement in hearing, he was referred to our hospital after two days. The patient was diagnosed with idiopathic sudden sensorineural hearing loss (ISSNHL) with vertigo on the right side and hospitalized. No additional neurologic symptoms were noted. His past medical history included diabetes, hypertension, chronic myocardial infarction, left cerebellar infarction, chronic renal failure, hyperlipidemia, hyperuricemia, hypothyroidism, and sleep apnea.

An audiogram (Figure 1A) revealed sensorineural hearing loss of approximately 75 dB in the right ear, and gaze-, positioning-, and positional-nystagmus were evident in the rightward direction (Figure 1B). Six hours after hospitalization, his gaze nystagmus disappeared and non-gaze nystagmus decreased (Figure 1C), but nystagmus direction continued on the affected side. We conducted an electronystagmogram (ENG) immediately after hospitalization and performed the Eye Tracking Test (ETT), optokinetic nystagmus test (OKN), caloric test, and visual suppression test (VS). The caloric test revealed a good reaction on both sides, and the ETT, OKN, and VS had normal results. Taken together, these findings suggested the absence of brainstem-cerebellar disorders.

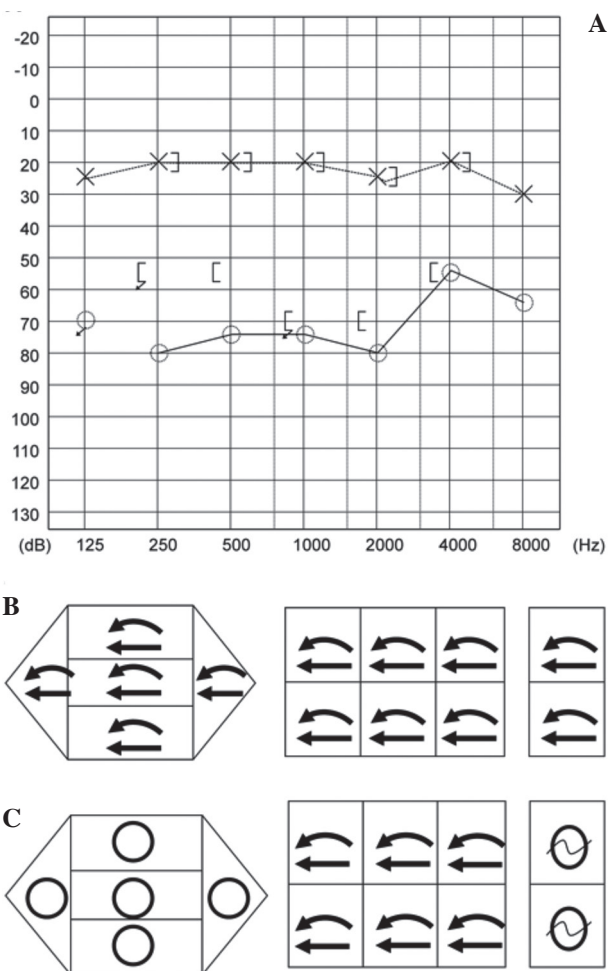


Figure 1

Test results for Case 1 on hospital admission.  
 A: Standard audiometry test: sensorineural hearing loss of about 75 dB was evident in the right ear.  
 B: Gaze and non-gaze nystagmus were both detected in the rightward direction.  
 C: Gaze and non-gaze nystagmus 6 h after hospitalization. Gaze nystagmus was not seen but non-gaze nystagmus remained in the same direction (i.e., towards the right).

On the second day of hospitalization, the patient's vertigo worsened, and gaze nystagmus reappeared. As nystagmus is not a characteristic symptom of inner ear disease, magnetic resonance imaging (MRI) was performed and revealed an AICA infarction in the right cerebellum (Figure 2).

### Case 2

Case 2 describes a 57-year-old man who experienced vertigo, cold sweats, and vomiting on waking. From the time of symptom onset, the man experienced dysacusia and hearing loss in his right

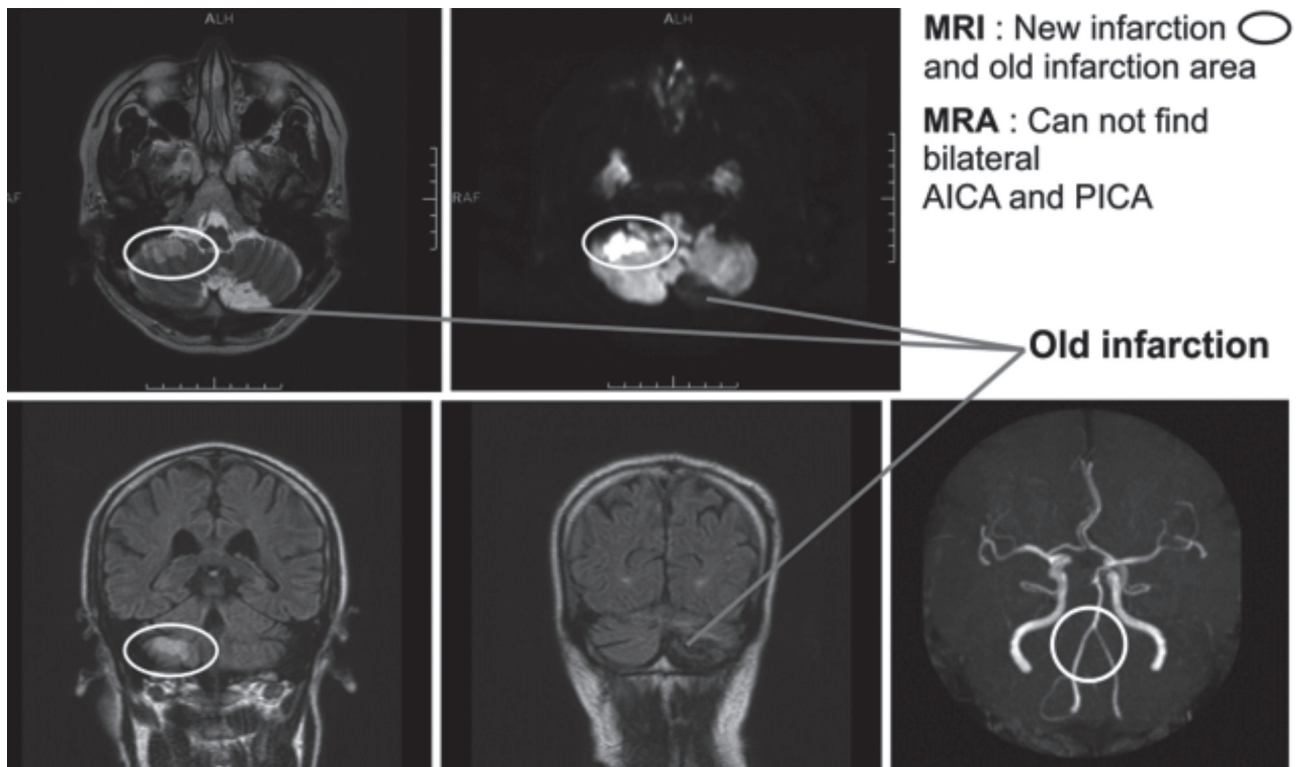


Figure 2

MRI and MRA findings of Case 1.

MRI showed a new infarction. MRA could not detect bilateral AICA (anterior inferior cerebellar artery) and PICA (posterior inferior cerebellar artery).

ear. In addition to his medical history of hypertension, diabetes, hyperlipidemia, and hyperuricemia, he was a heavy smoker for at least a decade and reported drinking alcohol twice weekly. The patient stayed in bed for three days due to his symptoms; when they persisted, he visited a hospital emergency room and underwent brain computed tomography. No abnormal results were detected.

His symptoms continued, along with such severe episodes of vomiting that he was unable to leave his bed.

Seven days after symptom onset, the man consulted the ENT clinic, where an audiometer revealed high-tone hearing loss in his right ear. We suspected that the patient was being treated for Ménière's disease or sudden deafness with vertigo. When the man's vertigo and hearing loss persisted for 10 days after the ENT visit, he presented to our hospital 17 days after the initial onset of symptoms. At the time of consultation, the patient had difficulty standing on one leg and was unable to hear a voice on the other end of a telephone. During hospitali-

zation, no facial tactile blunting, peripheral facial nerve paralysis, dysesthesia, or headache was observed. The patient's right hearing level (Figure 3A) decreased predominantly in the low-middle tone range (125-4000 Hz). The patient also exhibited non-gaze nystagmus to the left (Figure 3B). Owing to the persistent symptoms and presence of atypical nystagmus, we performed MRI at his initial hospital visit, and the patient was diagnosed with a right AICA infarction (Figure 4).

## Discussion

When patients present only with the comparatively common symptoms of vertigo and hearing loss, differentiating between central nervous system disorders and peripheral inner ear disease can be challenging.<sup>5</sup> Distinguishing between the two is critically important in patient care.<sup>6</sup>

Owing to the overlap with symptoms of inner ear disease, cases of AICA infarction with advanced hearing loss may be misdiagnosed as sudden deafness with vertigo<sup>7,8</sup> or Ménière's disease.

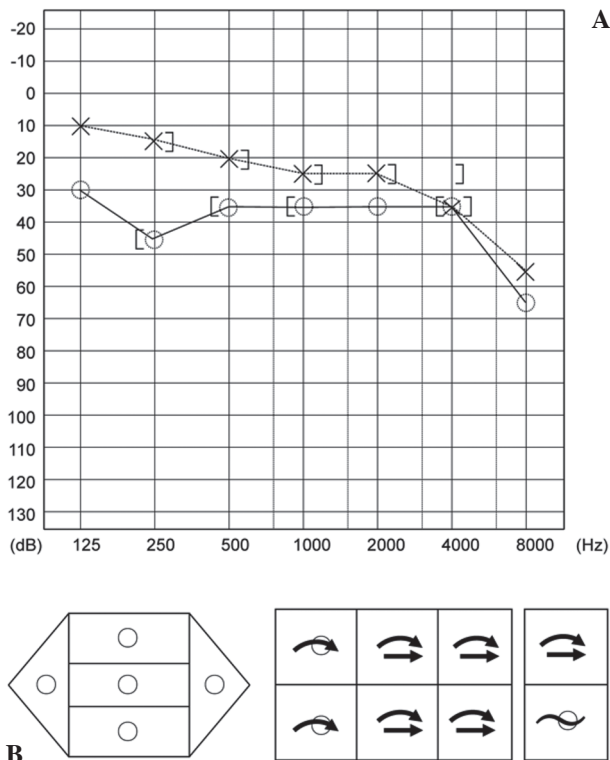


Figure 3

Test results for Case 2 at the initial hospital visit. A: Standard audiometry test: the right side indicates sensorineural hearing loss of the low tone type. B: Gaze nystagmus was not seen; however, non-gaze nystagmus was detected in the leftward direction. The strength of nystagmus changed according to the head position.

Previous reports have discussed cases of AICA syndrome where initial symptoms included hearing loss and vertigo, with additional neurological symptoms appearing later in the clinical course.<sup>10,11</sup> Although many patients will ultimately receive the correct diagnosis of an AICA infarction, the delay in diagnosis and treatment can have grave consequences.

In addition to AICA infarction, Nowé *et al.* showed that MRI also revealed neurovascular conflicts in the cerebellopontine corners in many cases.<sup>12</sup>

In our first case, the patient was initially diagnosed with ISSNHL with vertigo. Hearing loss was at a moderate to high level, and the patient exhibited gaze nystagmus of the affected side two days after symptom onset. Although ISSNHL with vertigo is quite common, the side of hearing loss and the direction of nystagmus are typically opposite in direction. Additionally, the relapsing-remitting nature of the patient's nystagmus further supported the idea that his diagnosis was not consistent with ISSNHL. In light of the clinical presentation, it was unsurprising that this patient's MRI scan revealed the correct diagnosis of AICA syndrome.

In contrast, the second patient presented with slight hearing loss in addition to several days of vertigo and severe truncal ataxia. The slight hearing loss and dizziness led clinicians to an initial diagnosis of Ménière's disease. However, this case was also characterized by strong sustained nausea

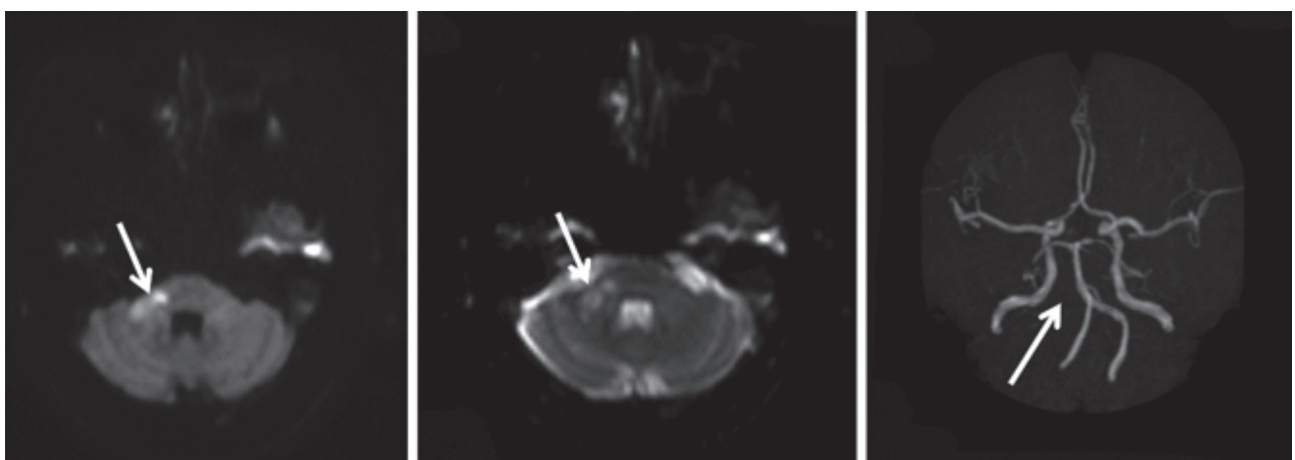


Figure 4

MRI and MRA findings of Case 2. Diffusion-weighted MRI indicated an infarction, and T2-MRA could not detect the right AICA.

and vomiting, inconsistent with inner ear disease. An MRI was performed and confirmed an AICA infarction.

These cases demonstrate the importance of considering the complete clinical presentation when evaluating a patient presenting with hearing loss. Individual symptoms of AICA infarction may mimic inner ear disease, but the additional symptoms indicate the true nature of the condition. This includes not only the present complaints, but also any additional risk factors associated with cerebrovascular disease, such as hypertension, hyperlipidemia, and diabetes – all of which were found in both the cases described above. The clinical course, medical history, laboratory findings, and neurological imaging findings are essential for accurate diagnosis and appropriate treatment of AICA infarction.

### Conclusion

For an otolaryngologist, it is essential to be able to distinguish vertigo and hearing loss due to inner ear damage from those caused by cerebrovascular disorders. Familiarity with the features of inner ear disease and AICA infarction is important for accurate diagnosis and appropriate treatment.

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