Bilateral Sudden Deafness as a Prodrome of Anterior Inferior Cerebellar Artery Infarction

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Background: Acute ischemic stroke in the distribution of the anterior inferior cerebellar artery is known to be associated with hearing loss, facial weakness, ataxia, nystagmus, and hypalgesia. There have been few reports on bilateral deafness and vertebrobasilar occlusive disease. Furthermore, previous reports have not emphasized the inner ear as a localization of bilateral deafness.

Objective: To describe the presentation of acute ischemic stroke in the distribution of the anterior inferior cerebellar artery as sudden bilateral hearing loss with minimal associated signs.

Design and Setting: Case report and tertiary care hospital.

Patient: A 66-year-old man with diabetes mellitus developed sudden bilateral deafness, unilateral tinnitus, and vertigo 7 days before the onset of dysarthria, facial weak-

ness, and ataxia. T2-weighted magnetic resonance imaging scans showed hyperintensities in the right lateral pons and right middle cerebral peduncle and a possible abnormality of the left middle cerebellar peduncle. A magnetic resonance angiogram showed moderately severe stenosis of the distal vertebral artery and middle third of the basilar artery. The patient's right limb coordination and gait improved steadily over several weeks, but there was no improvement in hearing in his right ear.

Conclusions: The relatively isolated onset of deafness as well as the severity and persistence of the hearing loss led us to conclude that the hearing loss in this case was likely due to prominent hypoperfusion of the internal auditory artery, with labyrinthine infarction as the earliest event. Vertebrobasilar occlusive disease should be considered in the differential diagnosis of sudden bilateral deafness.

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UDDEN DEAFNESS may be defined as more than 20 dB of sensorineural hearing loss occurring over minutes to hours.1 Sudden deafness occurs unilaterally in most cases, and bilateral, simultaneous, sudden deafness is a rare condition, accounting for 0.44% to 3.4%^{2,3} of cases of sudden deafness. Unilateral, sudden deafness in a young patient is usually idiopathic and is often thought to be viral^{4,5}or autoimmune⁶ in origin. On the other hand, sudden deafness in an older patient with known cerebrovascular occlusive disease suggests the likelihood of ischemia in the distribution of the internal auditory artery, ordinarily a branch of the anterior inferior cerebellar artery (AICA).7-10 Characteristically, sudden bilateral deafness due to AICA ischemia is associated with multiple brainstem signs.^{11,12} The AICA supplies the dorsolateral pons, middle cerebellar peduncle, inner ear, and anterior inferior cerebellum, including the flocculus.^{13,14} Infrequently, infarction of the AICA territory is preceded by isolated episodes of vertigo or unilateral hearing loss.⁷⁻¹⁰ There have been few reports on bilateral deafness and vertebrobasilar occlusive disease.11,12,15,16 Furthermore, previous reports have not emphasized the inner ear as a localization of

bilateral deafness.^{11,12,15,16} We describe a patient with AICA infarction who presented with isolated sudden bilateral deafness as the initial symptom.

REPORT OF A CASE

A 66-year-old man with type 2 diabetes mellitus suddenly developed bilateral hearing loss, tinnitus in his right ear, and vertigo, all of which he noticed when he got up in the morning. He did not have dysarthria, weakness, ataxia, diplopia, dysphagia, Horner syndrome, or sensory loss. The hearing loss persisted, but the vertigo subsided within 1 day, and subsequent examination showed only mild unsteadiness of gait. Seven days after the initial onset of the bilateral hearing loss and vertigo, the patient presented with exacerbation of the hearing loss in his right ear, combined with right-sided tinnitus, vertigo, nausea, and incoordination. On examination, he had a spontaneous left-beating nystagmus with a horizontaltorsional component in primary position and with gaze to the right or left. There was diminished right-sided facial sensation, a right-sided facial palsy with peripheral features, and dysmetria of the right limbs. Audiometry showed moderate sensorineural hearing loss of 55 dB with 100% speech dis-

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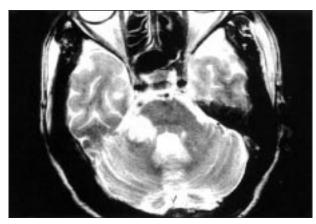


Figure 1. Axial T2-weighted magnetic resonance image shows hyperintensities involving the right middle cerebellar peduncle, right lateral pons, and possibly the left middle cerebellar peducle.

crimination on the right and 45 dB with 100% speech discrimination on the left. Electronystagmography showed no response to caloric stimulation of the right ear; and oculomotor testing showed asymmetrical abnormalities of both smooth pursuit and optokinetic nystagmus.

T2-weighted magnetic resonance imaging scans revealed hyperintense foci involving the right dorsolateral pons, right middle cerebellar peduncle, and possibly the left middle cerebellar peduncle (**Figure 1**). A magnetic resonance angiogram showed moderately severe stenosis of the distal right vertebral artery and the middle third of the basilar artery (**Figure 2**). A transthoracic echocardiogram and an electrocardiogram revealed no abnormalities.

During several days of anticoagulation in the hospital, the patient's vertigo and nausea improved. A follow-up audiogram showed a profound hearing loss in the right ear, and the hearing loss in the left ear had improved to 30 dB (**Figure 3**). His right limb coordination and gait improved steadily over several weeks, but there was no improvement in hearing in his right ear. At last follow-up, he remained on a daily regimen of warfarin and has had no further neurological symptoms.

COMMENT

Sudden deafness with vertigo usually suggests the diagnosis of an inner ear disorder, eg, Meniere disease, acute labyrinthitis, autoimmune inner ear disease, or perilymphatic fistula. Previous reports of sudden, bilateral deafness caused by vertebrobasilar occlusive disease have been rare. In 1943, Adams¹⁵ was the first to completely describe the syndrome associated with AICA occlusion. In his patient, tinnitus was an early symptom, appearing soon after vertigo, and the patient had acute bilateral, asymmetrical hearing loss, suggesting bilateral ischemia. Adams alluded to the difficulty of assigning a time of onset to the symptom of hearing loss, noting that "deafness ... may not at all attract the attention of an unobservant patient." Details of the timing of hearing loss in patients with stroke have seldom been reported, suggesting that the issue of transient ischemic attacks consisting of hearing loss could have been frequently overlooked. In 1990, Amarenco et al¹⁷ described a series of 20 patients with AICA distribution strokes:

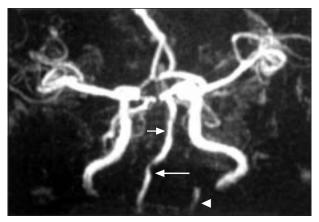


Figure 2. Magnetic resonance angiogram shows moderately severe stenosis of the distal right vertebral artery (long arrow) and middle third of the basilar artery (short arrow). The left distal vertebral artery is also visualized (arrowhead).

5 of the 20 patients had hearing loss as part of the stroke syndrome. However, none had hearing loss at stroke onset, although 2 had tinnitus at onset. All patients with hearing loss had vertigo or unsteadiness at stroke onset. In 1998, Roquer et al¹⁸ described 15 patients with AICA strokes. All 15 patients had vertigo or ataxia, and 6 of the 15 had hearing loss. However, the timing of hearing loss in relation to vertigo and ataxia was not reported. Stephan et al¹¹ described a patient with sudden, bilateral deafness caused by basilar artery occlusion who later developed progressive quadriplegia with mental status abnormalities. Huang et al¹² described 7 patients with bilateral, sudden deafness in vertebrobasilar occlusive disease. All but 1 of the 7 patients had bilateral hearing impairment as the initial symptom. However, 3 of the 7 patients also had quadriparesis (n=1), hemianesthesia (n=1), or dysarthria (n=1) at onset, and 4 had vertigo at onset. The report did not include audiometric data, limiting localization. Five of the patients in this series had poor outcomes, including the locked-in syndrome and severe ataxia with abasia. One patient had permanent bilateral deafness and diplopia, and only 1 patient made a good recovery. Presumably, these 2 previous reports reflect severe ischemic damage to multiple structures in the posterior fossa. Therefore, the clinical course in our patient may have been remarkable in that there were few residual deficits.

In our patient, bilateral deafness and tinnitus occurred several days before the sudden onset of AICA stroke with typical brainstem signs. The analogous presentation of isolated vertigo has become well recognized, following its description by Grad and Baloh.7 These authors reported that of 84 patients with vertigo of presumed vascular origin, 36 had episodic, isolated vertigo and 35 had definite caloric paresis or weakness. It was suggested that vertebrobasilar ischemia may selectively damage the inner ear because of this structure's high energy requirements and the lack of adequate collateral blood supply.^{7,8} With few exceptions, the internal auditory artery receives its blood supply from the AICA, and the inner ear receives its sole blood supply from the internal auditory artery. Previous reports have suggested that the inner ear is one of the most commonly affected areas in AICA strokes.7-10 Hearing loss for pure tones is unusual with central lesions, even in the late stages.¹⁹ Our patient had persistent, unilateral,

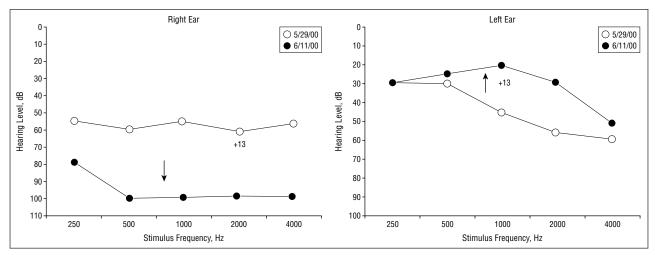


Figure 3. Pure-tone air conduction audiograms obtained on 2 different dates, 13 days apart (+13). The patient's hearing levels are plotted against stimulus frequency on a logarithmic scale. The decibels were measured according to the American National Standards Institute, 1989.

profound deafness on pure tone audiometry, but the other deficits essentially resolved. The combination of these audiometric results and the marked improvement of central signs is consistent with the hypothesis that the major locus of injury in our patient was the inner ear. Audiometry in our patient showed good speech discrimination, suggesting a cochlear localization. Moreover, the root entry zone of the eighth cranial nerve is believed to have abundant collateral blood supply,^{20,21} making injury to cranial nerve VIII an unlikely possibility. Given these facts, we speculate that the prodrome of hearing loss in our patient resulted from a fairly restricted insult to the inner ear. Despite bilateral hearing symptoms, definite magnetic resonance imaging abnormalities of the brainstem were found only on the right. We speculate that the prominence and bilaterality of hearing symptoms and the absence of the definite left-sided brain involvement are best explained by the selective vulnerability of the inner ear to ischemia.

Territory strokes of the AICA have been shown to be associated with basilar artery branch occlusive disease.^{17,22,23} Because our patient had evidence of a focal segment of reduced flow in the basilar artery close to the origins of the AICA, it is possible that an atheromatous plaque within the basilar artery extended into the AICA ostia to different degrees. By this mechanism, decreased blood flow in the right and left AICAs might cause either selective damage to the inner ear or more widespread damage to the middle cerebellar peduncle, lateral pons, and cerebellum. In conclusion, an oligosymptomatic presentation in which sudden hearing loss predominates may be a prodrome of brainstem infarction. The mechanism of this presentation is likely ischemia of the labyrinth. In some such patients, basilar artery stenosis may be associated with low flow through narrowed ostia of the AICA, although the evidence for this speculation is indirect. Vertebrobasilar occlusive disease should be routinely considered in the differential diagnosis of sudden deafness.

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