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 weakness, 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. 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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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 horizontal-torsional 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-
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: 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 al19 described a patient with sudden, bilateral deafness caused by basilar artery occlusion who later developed progressive quadriplegia with mental status abnormalities. Huang et al20 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 quadripareisis (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 vertebrobasilar origin, 36 had episodic, isolated vertigo and 35 had vertebrobasilar origin, 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:
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 prograde 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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REFERENCES