

Sudden Sensorineural Hearing Loss as Prodromal Symptom of Anterior Inferior Cerebellar Artery Infarction

Francesco Martines^a Francesco Dispenza^a Cesare Gagliardo^b Enrico Martines^c
 Daniela Bentivegna^c

^aSezione di Otorinolaringoiatria, Dipartimento di Biomedicina Sperimentale e Neuroscienze Cliniche, e Sezioni di ^bScienze Radiologiche, e ^cAudiologia, Dipartimento di Biopatologia e Biotecnologie Mediche e Forensi, Università degli Studi di Palermo, Palermo, Italia

Key Words

Sudden deafness · Anterior inferior cerebellar artery infarction · Internal auditory artery · Inner ear

Abstract

Sudden sensorineural hearing loss is a clinical condition characterized by a sudden onset of unilateral or bilateral hearing loss. In recent years sudden deafness has been frequently described in association with anterior inferior cerebellar artery (AICA) infarction generally presenting along with other brainstem and cerebellar signs such as ataxia, dysmetria and peripheral facial palsy. The authors report a rare clinical case of a 53-year-old man who suddenly developed hearing loss and tinnitus without any brainstem or cerebellar signs. Computed tomography of his brain was normal, and the audiological results localized the lesion causing deafness to the inner ear. Surprisingly, magnetic resonance imaging showed an ischemic infarct in the right AICA territory. This case represents the fifth in the literature to date but it confirms that AICA occlusion can cause sudden deafness even without brainstem or cerebellar signs. Therefore, we recommend submitting the patient for neuroimaging, as an emergency, in order to exclude infarction of the AICA territory. By doing this, it may be possible to limit the extent of the lesion by commencing early therapy.

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Introduction

Sudden sensorineural hearing loss is defined as more than 20 dB hearing loss occurring in 3 contiguous frequencies over minutes to hours, frequently accompanied by pressure in the ears, tinnitus, vertigo and disequilibrium [1]. Even if numerous etiologies have been postulated as an explanation for this syndrome, including viral infection (varicella-zoster virus, cytomegalovirus, Epstein-Barr virus, rubella virus, parvovirus), ototoxicity, autoimmune disease, drugs (salicylates, aminoglycosides) and vestibular schwannoma, often sudden sensorineural hearing loss is considered an idiopathic disease as no etiology can be identified in the majority of cases.

Recently, sudden deafness has been described in cases of cerebellar infarction of the anterior inferior cerebellar artery (AICA) area because the blood supply to the inner ear is from the internal auditory artery (IAA), which in turn usually originates from the AICA. In these cases, sudden deafness is characteristically associated with other brainstem and cerebellar signs such as ataxia, dysmetria, hypalgesia and peripheral facial nerve palsy [2].

We report the fifth case in the literature of a patient with AICA infarction causing unilateral sudden deafness without any cerebellar signs at presentation [1–4]. We diagnosed this patient using neuro-otological and neuroaudiological examination.

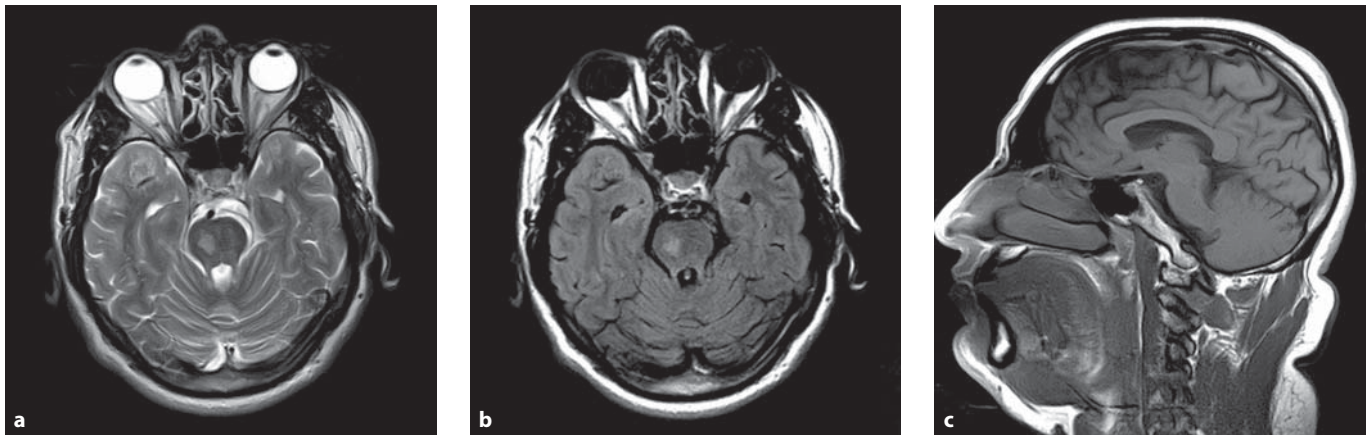


Fig. 1. Brain MRI. **a, b** Axial planes at the level of the superior cerebellar peduncles in fast spin echo T₂-weighted and fluid-attenuated inversion recovery T₂-weighted images, respectively, showing a high-signal lesion. **c** Sagittal plane in fast spin echo T₁-weighted imaging evidencing a normal signal intensity.

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Case Report

A 53-year-old man developed right-sided sudden hearing loss accompanied by tinnitus, unsteadiness of gait and severe headache upon awakening in the morning. On admission, he was conscious and had normal blood pressure (135/87 mm Hg), a pulse rate of 80 beats/min and a body temperature of 37°C. He did not come up with any history of temporal bone fracture, meningitis, autoimmune disease or exposure to ototoxic drugs. He was a light smoker.

On examination he had spontaneous, left beating nystagmus with horizontal and torsional components, in the primary position. No nausea and vomiting were reported. The nystagmus was inhibited by visual fixation, indicating a peripheral vestibular weakness. The external auditory canals, and the tympanic membranes were normal bilaterally. There were no neurological deficits like dysarthria, facial or limb sensory loss, facial or limb weakness, ophthalmoparesis, diplopia or Horner's syndrome. Also, no dysmetria could be demonstrated on finger-to-nose and heel-to-shin testing.

His hemogram, blood coagulation and clotting profile, blood sugar levels, lipid profile, and renal and liver functions were normal. Autoimmune markers, sickling test and urine homocysteine tests were negative. Chest X-ray, ECG and transesophageal echocardiography were normal as well.

Audiometry showed right sensorineural hearing loss with pure-tone audiometry thresholds of 75 dB suggesting cochlear involvement. The patient had a speech discrimination score of 70%. Weber's test was lateralizing to the left. Tympanometry proved to be normal bilaterally. The stapedial reflex was elicitable on the affected side as sign of cochlear hearing loss (recruitment).

The study of auditory brainstem responses was normal on the left side while it showed a delay in absolute latencies of all waves on the right side. However, interpeak latencies of waves I, III and V were normal on the affected side.

Vestibular evoked myogenic potentials were absent on the right side and normal on the left side.

A computed tomography (CT) scan of the brain performed at presentation in the emergency department did not reveal any ischemic and/or hemorrhagic brain lesions. Although CT is not routinely done for sudden sensorineural hearing loss, this patient had a CT scan because of gait difficulty and persistent headache. The patient was subjected to a magnetic resonance imaging (MRI) scan after a day because of persisting symptoms, which demonstrated an acute ischemic infarct in the right AICA territory with involvement of the pons, especially on conventional/morphological T₂-weighted sequences (fast spin echo; fluid-attenuated inversion recovery) and confirmed on functional sequences (diffusion-weighted images; apparent diffusion coefficient; fig. 1–3).

He was treated with 400 ml/day glycerol, 60 mg/day edaravone, furosemide 20 mg/day and sodium heparin 5,000 IU b.i.d. s.c.; the treatment was administered for 10 days, then the patient was discharged.

The patient underwent follow-up audiography 1 and 3 months after discharge which showed persistent hearing loss on the right side.

Discussion

Sudden onset of hearing loss or vertigo is generally attributed to injury of the inner ear or the eighth cranial nerve [5]. These symptoms are most commonly due to as yet idiopathic causes or sometimes caused by viral infections. AICA infarction can cause similar symptoms as the blood supply to the inner ear is from the IAA, which in turn originates from the AICA [3]. The AICA typically arises from the junction of the middle and lower thirds of the basilar artery, courses along the pons and the middle cerebellar peduncle and then bifurcates into 2 major branches: the rostralateral and caudomedial

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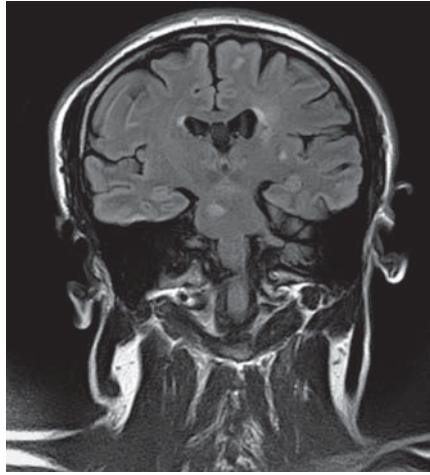


Fig. 2. Coronal plane in fluid-attenuated inversion recovery T₂-weighted imaging showing moderately increased signal intensity of the pons.

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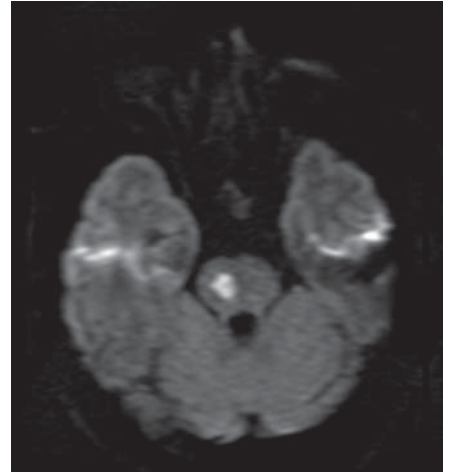


Fig. 3. Diffusion-weighted imaging showing a high signal lesion expression of cytotoxic edema typical of acute ischemic stroke.

branches. The rostralateral branch courses towards the internal acoustic canal close to the seventh and eighth cranial nerve complex and gives off the IAA. This again divides into 2 main branches, the common cochlear artery and the anterior vestibular artery and thus supplies the inner ear. The caudomedial branch courses medially close to the pons, to which it sends a few perforators and terminates as the cerebellar branches [6]. Because of its usually small size, the AICA supplies only a small area of the anterior and medial cerebellum. Proximal branches of the AICA usually supply the lateral portion of the pons, including the facial, trigeminal, vestibular and cochlear nuclei, the root of the seventh and eighth cranial nerves and the spinothalamic tract [4].

The most common mechanism of AICA infarction is either by thrombotic narrowing of the AICA itself or by extension of the basilar artery plaque into the AICA, blocking the AICA orifice [7]. Clinical features of AICA infarction depend on factors such as size and location of obstruction. However, the typical presentation includes sudden deafness associated with other brainstem or cerebellar signs like ataxia, hypalgesia, peripheral facial palsy, vertigo and nystagmus. To date, the literature reports only 4 cases in which this pathological entity is associated with a labyrinthine-type syndrome without other brainstem or cerebellar signs [1–4].

Our patient presented with a sudden onset of right-sided deafness, tinnitus, vertigo and nystagmus without any cerebellar signs, and represents the fifth case in the literature. The labyrinthine-type syndrome that follows infarction in the AICA territory could be explained by several different mechanisms. The first is by occluding the IAA artery which supplies the inner ear and arises

from the AICA in 83% of cases. Other mechanisms include ischemic injury of the vestibular and cochlear nuclei in the pons, involvement of fibers of the eighth nerve in the lateral pontine area or involvement of the floccular lobe or its connections [4]. In addition to a variety of audiovestibular symptoms like sudden deafness, recurrent vertigo, continuous disequilibrium or tinnitus, acute ischemic stroke in the distribution of the AICA is characteristically associated with brainstem and cerebellar signs manifesting as crossed sensory loss, lateral gaze palsy, facial palsy, Horner's syndrome, ataxia and dysmetria. This is because infarction might occur in the pons with involvement of facial and abducens nuclei, pyramidal tracts and sensory tracts which are anatomically close to the vestibular and cochlear nuclei as well as in the cerebellum [1]. Therefore, when vertigo or hearing loss is associated with other brainstem or cerebellar signs, the diagnosis of AICA infarction is straightforward.

However, since the IAA supplying the inner ear is a branch of the AICA and is an end artery with minimal collaterals from other major arterial branches, partial ischemia of the AICA may rarely lead to isolated deafness or vertigo, mimicking more common vestibular disorders including Ménière's syndrome, vestibular neuritis or labyrinthitis, especially when the pontine territory is not involved [8].

In view of different therapeutic strategies and prognoses of strokes involving the AICA, it is of great importance to differentiate labyrinthine infarction from more benign disorders involving the inner ear. As this case amply shows, in the presence of acute vestibulocochlear syndrome, the possibility of AICA infarction should be considered, particularly in elderly patients with vascular risk

factors and no prior history of hearing or vestibular dysfunction, even when the classic brainstem or cerebellar signs are absent.

Therefore, we strongly suggest that these patients should be admitted and evaluated with an MRI within few days, for possible progression of devastating AICA and/or vertebrobasilar ischemia that evolved on a delayed basis (2–7 days). In fact, MRI is universally accepted in the protocols of diagnosis of sudden hearing loss but more often is performed after 10–15 days.

It is extremely useful in detecting small asymptomatic vascular lesions within the AICA territory or alteration of the perfusion, which could have repercussions on the circulation of the inner ear.

In the case described, neurological symptoms were absent and the findings of hearing threshold, stapedial reflex testing and auditory brainstem responses supported the initial hypothesis of cochlear damage. All these findings were further authenticated by a negative brain CT scan ruling out any acute supratentorial and cerebellar ischemic or hemorrhagic lesions. However, the presence of unsteadiness of gait and severe headache on admission, associated with spontaneous left beating nystagmus and the absence of vestibular evoked myogenic potentials on the affected side, provoked us to perform an MRI that

easily showed an infarct in the right AICA territory with involvement of the pons. The early diagnosis leading to early medical intervention was able to limit the extent of the lesion solely to the labyrinth.

Conclusion

AICA infarction can present as sudden sensorineural hearing loss with or without vertigo. Clinicians should be aware of the possibility of AICA infarction particularly in older patients with sudden deafness and vascular risk factors, even when classic brainstem or cerebellar signs are absent. It is necessary to perform a neurological examination and to consider a prompt neuroimaging with MRI to achieve the true diagnosis in case of sudden deafness secondary to infarction of the vertebral-basilar artery territory which is difficult to be picked up in a brain CT, especially if the lesion is localized in the brainstem.

Disclosure Statement

The authors report no conflict of interest.

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