Acute vestibular syndrome with down-beat nystagmus as a sole clinical presentation in AICA transient ischemic attack with an uncommon clinical course

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ABSTRACT
Isolated acute vestibular syndrome remains a diagnostic challenge in the emergency department and the initial approach should include the identification of a central or peripheral etiology. This is the case report of an elderly patient with known cardiovascular risk factors presenting with acute vertigo and unsteadiness. Neurological examination was notable only for down-beat nystagmus and diffusion-weighted MRI showed normal findings. He was treated as having an emerging anterior–inferior cerebellar artery (AICA) stroke. Even when MRI showed no signs of hemorrhage or infarction, the neurotological bedside examination was a determinant. The cochleovestibular system was not spared by the ischemic injury but a more extensive neurological damage was probably avoided by approaching this case as a stroke.

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Introduction
In the initial assessment of patients with a recent onset and acute vestibulopathy, it is crucial to differentiate between peripheral and central causes. In elderly patients and in particular those with cardiovascular risk factors, diagnosis should exclude stroke [1]. Most of the literature relies on the association of signs and symptoms of hearing impairment with acute vertigo in order to localize the site of the lesion, although this principle does not always apply since patients with acute auditory symptoms added to vestibulopathy are sometimes diagnosed with stroke in the anterior–inferior cerebellar artery (AICA) territory [2].

Even when audiovestibular and imaging tests are complementary to reach the diagnosis, the neurotological examination provides key clues which raise clinical suspicion of a possible stroke.

We present a 78-year-old male patient with known vascular risk factors and bilateral moderate sensorineural hearing loss with acute onset of vertigo, unsteadiness and down-beat nystagmus (DBN) without other neurological deficits and who despite normal findings on MRI including diffusion-weighted images was treated as having an AICA stroke. Cochleovestibular damage could not be reversed but probably a more extensive ischemic damage was prevented by making an exception to the rule.

Case report
A 78-year-old male with chronic high blood pressure, diabetes mellitus, ischemic heart disease, hypertriglyceridemia and known bilateral moderate sensorineural hearing loss, attended our emergency room with intense vertigo, gait instability and autonomic symptoms that had started 4–6 hours before. He denied tinnitus or changes in hearing level. On arrival, he showed no altered mental status, high blood pressure (172/57 mmHg) and other vital signs were within normal range. His ABCD2 score was 5. A thorough otoneurological bedside examination was performed which was remarkable for saccadic smooth pursuit in both horizontal and vertical planes, a spontaneous DBN in primary position and in rightward gaze (Figure 1(A)), unchanged with visual fixation.
suppression with Frenzel glasses. The vertical component of this DBN increased after head shaking (Figure 1(B)). No skew deviation on cover test was observed. The clinical head-impulse test was normal. On positional testing, nystagmus changed to upbeat and slightly torsional (geotropic) on hyperextension and on right Dix–Hallpike maneuver; on left Dix–Hallpike the nystagmus was down-beat. None of those

Figure 1. (A) Spontaneous down-beat nystagmus (H: horizontal; V: vertical). (B) Head shaking test. (C) vHIT for horizontal semicircular canals.
positions produced a worsening in the initial symptoms. When standing, the patient was unable to keep stance and with eyes closed falling backwards was prevented by the examiner.

The vestibulo-oculomotor reflex (VOR) was assessed with the video head impulse test (vHIT, GN Otometrics). Rightward and leftward head thrusts provoked normal responses for horizontal semicircular canals (Figure 1(C)). The bilateral superior and posterior canals were not evaluated at that time because of severe nausea and the patient being unable to keep staring at the fixation dot after the assessment of the horizontal canals.

The neurological examination was normal for mental status. Cranial nerves exploration showed no deficits. Proprioception and deep sensitivity were preserved. No paretic signs were observed. Myotatic reflexes were normal. He exhibited gait imbalance with a trend to fall backwards. Transcranial Doppler sonography showed right vertebral artery stenosis of about 50–70%. Brain MRI including diffusion-weighted images showed signs of discrete brain atrophy with cortical predominance, few signs of leukoaraiosis and no signs of ischemic or infarcted areas (Figure 2).

Immediate procedures. An acute vestibular syndrome of central origin was established and the patient was admitted in a stroke unit. Treatment was initiated immediately with anticoagulant therapy (low molecular weight heparin at 7500 IU per day) and sulpiride 150 mg daily (selective antagonist of dopamine receptors) to help reducing dizziness.

Follow-up. The symptoms gradually resolved but four days after admission, he complained of right hearing loss. While performing sensitized Romberg’s test, the patient tended to fall on his right side. Pure tone audiometry showed a complete right hearing loss and an unchanged left sensorineural hearing.

A spontaneous left beating nystagmus was observed in the primary eye position which obeyed Alexander’s
law and it also increased after head-shaking test (Figure 3(A)). No skew deviation on cover test was detected.

The VOR gain was again assessed with the vHIT (Figure 3(B)). Rightward head thrusts disclosed abnormal results for all the semicircular canals. The vertical canals (superior and posterior) showed low gains, and stimulation of the horizontal canal elicited refixation saccades (covert and overt). Leftward head turns showed reduced VOR gain for vertical canals and normal gain for the horizontal canal. Steroid treatment was initiated. A new MRI including diffusion-weighted images showed no structural changes (Figure 2).

One week after onset of symptoms and in the absence of neurological signs of ischemic progression, the patient was discharged. At follow-up, two weeks later, hearing loss was unchanged. No spontaneous nystagmus or after head-shaking test was observed. The VOR assessed with the vHIT showed low gain for all the semicircular canals after rightward head thrusts; refixation saccades appeared in an organized or isochronic fashion. A rotatory chair test was performed and normal values for gain, phase and symmetry were obtained. Vestibular myogenic potentials (VEMP) were performed using 0.5 kHz tone burst stimulation. The responses showed lower wave amplitudes on the right side and the interamplitude difference obtained was 46%.

The clinical course of the patient was satisfactory and at three-year follow-up, he showed a stable chronic left sensorineural hearing loss and a

![Figure 3](image-url)

**Figure 3.** (A) Left beating nystagmus increased after head shaking test (H: horizontal; V: vertical). (B) vHIT showed rightward abnormal results.
compensated right vestibular loss with complete right hearing loss.

Discussion

Diagnosis of an AICA transient ischemic attack or stroke remains a challenge, especially when other neurological symptoms and signs are absent or not prominent. Based on the appearance of spontaneous nystagmus with visual fixation, its direction (pure down-beating) and the normal vHIT obtained for the horizontal canals, the clinical suspicion of a central vestibulopathy was highlighted from the very beginning of the examination. Other etiologies like vestibular migraine should be considered, but the lack of migraine history and accompanying symptoms like headache, phonophobia and photophobia during the episode in an elderly patient with vascular risk factors made this diagnosis less likely. On the other hand, the presence of central type nystagmus as well as the absence of otologic symptoms at onset permits to rule out other etiologies which produce peripheral involvement, like labyrinthitis or inner-ear disease.

According to the ABCD2 score and clinical presentation, an ischemic transitory attack (ITA) or a stroke in the AICA territory was suspected. Given the clinical course and the lack of radiological findings suggesting a stroke, the second possibility was ruled out [3]. Both clinical profiles show very complex clinical presentations due to the variable amount of tissue supplied by the AICA. The AICA has two territories of arterial blood supply: the proximal stem supplies the lateral area of the pons, and the lateral branch supplies the middle cerebellar peduncle by passing laterally and curling around the upper edge of the flocculus and the lateral portion of the pons tegmentum [4]. In the cerebellar hemisphere, it anastomoses with other three main arteries including a branch of the posterior–inferior cerebellar artery (PICA) at its medial aspect. So, in complete AICA infarction, there is hearing loss, facial weakness, limb and facial sensory loss, ataxia, and cerebellar dysmetria [5]. Moreover, there are reports of lateral medullary, lateral pontine and inferior cerebellar strokes that could mimic acute peripheral vestibulopathy [2]. However, in the absence of positive radiological findings, which is not an unusual situation, in particular in the first 72 hours after onset of symptoms [6], we decided to proceed in treating the patient facing the possibility of an ischemic stroke.

The main localizing sign was the DBN. It can arise due to the loss of inhibitory action that originates in the flocculus and acts on vestibular nuclei projections [7] of anterior semicircular canals which control vertical eye movements. Therefore, without this inhibitory action, eyes are prone to upward drifts which translate into clinical DBN which is not modified by visual fixation [8]. One may speculate that the change of direction of the spontaneous nystagmus from downbeat to upbeat and torsional on the straight head-hanging and right Dix–Hallpike positions is due to dislodged otocoria from an ischemic right utricle which have migrated to the right posterior semicircular canal as any combination of inner ear damage that detaches otocoria and spares some posterior semicircular canal function could produce posterior canal benign paroxysmal positional vertigo (BPPV). Cases of sudden hearing loss with simultaneous BPPV attributed to viral labyrinthitis with a patchy pattern of labyrinthine loss have been reported [9]. Gacek [10] proposed a neural mechanism for the nystagmus of posterior canal BPPV which may result from inadequate inhibition of the posterior semicircular canal from the saccular macula, especially its superior part, caused by degeneration of otolith neurons after reactivation of latent neurotropic viral infection. Transient labyrinthine ischemia might cause loss of inner ear function if parts of the labyrinth have different sensitivity to ischemia [11].

More severe cerebellar impairment was ruled out because of the absence of other cerebellar signs, in particular gaze-evoked nystagmus [12], also PICA infarction was not considered because of the absence of skew deviation.

Similar cases are reported in the literature with cochleovestibular signs and symptoms as an initial manifestation of AICA stroke without classic brainstem or cerebellar signs as well as cases of isolated acute vestibular syndrome [13–15]. In those cases, infarction signal appeared on MRI after onset of symptoms [13,16]. In our patient, the MRI imaging did not show signs of hemorrhage or infarction; however, prompt management avoided a progression to a greater neurological damage. A recent study suggests that all TIA patients should be considered for hospitalization on the basis of a heightened risk of early ischemic events and rapid access to risk-modifying treatments such as revascularization and anticoagulation and that a triage based on the ABCD2 score alone may delay revascularization or anticoagulation in nearly 20% of patients [17]. Another study found that TIA patients had better survival at 180 days when hospitalized in stroke units than those treated in alternate wards [18]. Nevertheless, this treatment could not prevent more intense damage to the inner
ear maybe because the internal auditory artery lacks collateral blood flow.

It is known that episodic vertigo and hearing loss may be a prodrome in AICA infarction, as Lee et al. showed in a study [5,19] in which 31% of patients with AICA stroke had either recurrent or single episodes of prolonged hearing loss but there is a shortage of more studies that demonstrate this association [20]. Previous moderate SHL was not considered as a predicting factor for vascular stroke in our patient, and long standing diabetes, hypertension, hypertriglyceridemia and ischemic heart disease and right vertebral artery stenosis were the comorbidities that probably led to a complete right labyrinthine syndrome.

Conclusions

It is important to carefully assess patients with high risk factors that attend specialized consultation with acute cochleovestibular syndrome. Peripheral features at clinical presentation do not rule out a central disorder and, as shown here, a central etiology does not rule out peripheral damage. The importance of oto-neurological bedside examination should never be obviated in order to diagnose correctly a serious condition that demands prompt management.

Disclosure statement

The authors declare no conflicts of interest.

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References

[1] Saber Tehrani AS, Kattah JC, Mantokoudis G, et al. Small strokes causing severe vertigo: frequency of false-negative MRIs and nonlacunar mechanisms. Neurology. 2014;83:169–173.

[2] Kattah J, Talkad A, Wang D, et al. HINTS to diagnose stroke in the acute vestibular syndrome: three-step bedside oculomotor examination more sensitive than early MRI diffusion-weighted imaging. Stroke. 2009;40:3504–3510.

[3] Strupp M, Brandt T. Current treatment of vestibular, ocular motor disorders and nystagmus. Ther Adv Neurol Disord. 2009;2:223–239.

[4] Atkinson WJ. The anterior inferior cerebellar artery. Its variations, pontine distribution, and significance in the surgery of cerebello-pontine angle tumors. J Neurol Neurosurg Psychiat. 1949;12:137–151.

[5] Lee H, Kim HJ, Koo JW, et al. Progression of acute cochleovestibulopathy into anterior inferior cerebellar artery infarction. J Neurol Sci. 2009;278:119–122.

[6] Newman-Toker DE, Kerber KA, Hsieh YH, et al. HINTS outperforms ABCD2 to screen for stroke in acute continuous vertigo and dizziness. Acad Emerg Med. 2013;20:986–996.

[7] Highstein S, Holstein G. The anatomy of the vestibular nuclei. Prog Brain Res. 2006;151:157–203.

[8] Leigh RJ, Zee DS. The neurology of eye movements. 4th ed. USA: Oxford University Press; 2006.

[9] Boleas MS, Vasquez F, Perez N. Progressive cochleovestibular labyrinthitis. Rev Laryngol Otol Rhinol (Bord). 2007;128:63–64.

[10] Gacek RR. Pathology of benign paroxysmal positional vertigo revisited. Ann Otol Rhinol Laryngol. 2003;112:574–582.

[11] Kim JS, Lopez I, Di Patre PL, et al. Internal auditory artery infarction: clinicopathologic correlation. Neurology. 1999;52:40–44.

[12] Strupp M, Hüfner K, Sandmann R, et al. Central oculomotor disturbances and nystagmus: a window into the brainstem and cerebellum. Dtsch Arztebl Int. 2011;108:197–204.

[13] Kim HJ, Lee SH, Park JH. Isolated vestibular nuclear infarction: report of two cases and review of the literature. J Neurol. 2014;261:121–129.

[14] Lee H, Ahn BH, Baloh R. Sudden deafness with vertigo as a sole manifestation of anterior inferior cerebellar artery infarction. J Neurol Sci. 2004;222:105–107.

[15] Lee H, Baloh R. Sudden deafness in vertebrobasilar ischemia: clinical features, vascular topographical patterns and long-term outcome. J Neurol Sci. 2005;228:99–104.

[16] Son EJ, Bang JH, Kang JG. Anterior inferior cerebellar artery infarction presenting with sudden hearing loss and vertigo. Laryngoscope. 2007;117:556–558.

[17] Cutting S, Regan E, Lee V, et al. High ABCD2 scores and in-hospital interventions following transient ischemic attack. Cerebrovasc Dis Extra. 2016;6:76–83.

[18] Cadilhac DA, Kim J, Lannin NA, et al. Better outcomes for hospitalized patients with TIA when in stroke units: an observational study. Neurology. 2016;86:2042–2048.

[19] Lee H. Aud iovestibular loss in anterior inferior cerebellar artery territory infarction: a window to early detection? J Neurol Sci. 2012;313:153–159.

[20] Newman-Toker DE, Reich SG. Wrong-Way nystagmus in the AICA syndrome. Laryngoscope. 2008;118:378–379.