Cerebellar Ischemia Initially Presented as Sudden Sensorineural Hearing Loss and Gaze-Evoked Nystagmus

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Sudden sensorineural hearing loss is an important medical emergency that warrants repeated mentioning. The etiology is most often related to inner ear or vestibulo-cochlear nerve pathology [1].

When SSNHL is accompanied by gait and limb ataxia, facial paresis or paralysis, crossed sensory signs, Horner's syndrome or nystagmus lacking vestibular pathology characteristics, the clinician should consider central nervous system pathologies, such as inferior cerebellar ischemia, meningitis, neoplasms (especially those of the cerebellopontine angle) and multiple sclerosis. The characteristics of the nystagmus accompanying these conditions differ from those of peripheral unilateral vestibular pathologies. In the latter, the nystagmus has horizontal and torsional components; it is intensified by directing the gaze in the fast phase axis and by preventing visual fixation [2]. In CNS pathologies, the nystagmus has mainly one component (purely vertical or horizontal), and the intensity of the nystagmus is unaffected by visual fixation and can be gaze evoked. Vertigo arising from cerebellar ischemia may result from occlusion of the posterior or anterior inferior cerebellar arteries due to embolism or atherosclerotic disease. Considering this etiology is of foremost importance since inferior cerebellum swelling can lead to brainstem compression and death [2].

We present a rare case of inferior cerebellar ischemia in the distribution of the left AICA, manifested initially with SSNHL, vertigo and gaze-evoked nystagmus only, without any other neurological signs. This highlights the importance of a meticulous neurological examination to determine whether a patient has a central or peripheral nervous system disease.

PATIENT DESCRIPTION

A 62 year old man presented with sudden left-sided hearing loss and tinnitus accompanied by vertigo, nausea and unsteadiness. He reported previous short episodes of less severe unsteadiness and vertigo. His medical history was significant for heavy smoking, hypertension, hyperlipidemia and long-standing uncontrolled type 2 diabetes mellitus.

Physical examination revealed a spontaneous, horizontal right beating nystagmus in rest position of the eye and rightward gaze, and a left beating nystagmus on leftward gaze. Tuning fork exam suggested left-sided sensorineural hearing loss. The rest of his neurological and otological examinations were unremarkable at presentation.

Computed tomography on admission showed no signs of infarct, hemorrhage or mass affect. Audiometry performed the next day documented severe to profound sensorineural hearing loss with an SRT (speech reception threshold) of 85 decibel in the left ear and 30 decibel in the right. With the probable diagnosis of SSNHL of peripheral origin the patient was admitted and treated with prednisone (1 mg/kg/day) and acyclovir (800 mg five times/day). During the next 2 days he became aggressive, noncompliant and restless

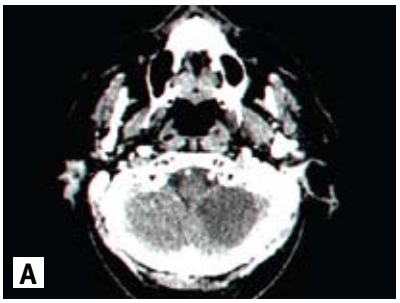
A complete left-sided facial nerve paresis was diagnosed on the third day, accompanied by blurred vision and diplopia on leftward gaze. Neurological examination at that time demonstrated postural instability, tendency to fall to the left side, gait ataxia and mild left hemiparesis. These were followed by dysarthria and left-sided hemianopsia. Repeated CT scans demonstrated a large hypodense area on the lower medial hemisphere of the cerebellum compatible with a large left cerebellar infarct in the distribution of the left AICA or PICA [Figure A]. Small filling defects were noticed in the basilar

SSNHL = sudden sensorineural hearing loss
CNS = central nervous system

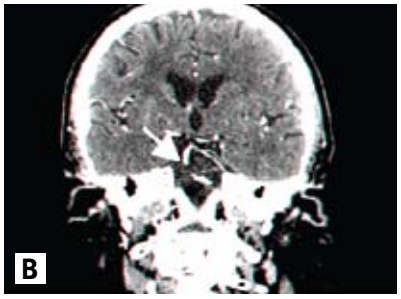
AICA = anterior inferior cerebellar artery

PICA = posterior inferior cerebellar artery

[A] Axial view of CT scan with contrast material demonstrates a large hypodense area on the left lower medial hemisphere of the cerebellum



[B] Coronal view of CT scan with contrast material demonstrates a filling defect in the basilar artery (white arrow).



artery [Figure B]. Doppler ultrasonography of the neck revealed normal flow in the right internal carotid artery and an occlusion of up to 49% in the left internal carotid artery. Anti-aggregant therapy with clopidogrel and aspirin was administered.

Thirty days later his vertiginous symptoms and ataxia had subsided, but the left hemiparesis persisted. At a follow-up visit 2 months later, he complained of persistent hearing loss on the left side. Physical examination revealed residual ataxia, the left facial nerve paresis had resolved and there was no spontaneous or gaze-evoked nystagmus. Repeated audiogram showed a slight improvement of 10 decibels on the left side.

COMMENT

The anterior inferior cerebellar artery is, with few exceptions, the feeding vessel of the internal auditory artery. The arterial supply to the facial and vestibulocochlear nerves arises from the internal auditory artery as well as the collateral arteries in the adjacent dura and petrous bone. However, the inner ear receives its arterial supply exclusively from branches of the internal auditory artery, the anterior vestibular and the common cochlear branches, which are end arteries. Therefore, occlusion of the internal auditory artery causes, in almost all cases, inner ear infarction with a variable degree of damage to the seventh and eighth cranial nerves. Clinicopathologic studies of AICA infarction have clearly shown that the lateral pons, middle cerebellar peduncle and the inner ear are the areas most commonly affected [3].

Regarding our patient, we can assume that transient ischemic attacks preceded the ischemic event in the distribution of the AICA as it bifurcates from the basilar artery. The delayed appearance of facial nerve paresis can be explained by propagation of the ischemic process to involve the recurrent penetrating arteries, several of which bifurcate from the AICA [3] and supply the more medial pons, or of a small artery that supplies the facial nerve root in the cerebellopontine angle. In AICA occlusion, it is also common to find ipsilateral facial weakness, deafness and tinnitus [4]. Therefore, from a clinical perspective our case closely resembles an AICA occlusion syndrome. Few reports describe AICA ischemia presenting with sudden deafness and vertigo only [4]. In those reports the nystagmus was described as having peripheral vestibular characteristics [4]. Our patient had a gaze-evoked nystagmus prior to the constellation of a fully expressed AICA syndrome. Gaze-evoked nystagmus is usually caused by a structural lesion in the neural integrator network in the brainstem, a neural system that converts eye velocity commands into signals that control eye position. This type of nystagmus is often caused by brainstem or cerebellar disease in particular, but it can also result from cerebral disease, weakness of one or several extra-ocular muscles or their innervations, as in myasthenia gravis or Guillain-Barré syndrome, or is due to the effect of medications such as sedatives, barbiturates or anticonvulsants [5].

In conclusion, SSNHL should always be considered a medical emergency. SSNHL and vertigo, especially when accompanied by gaze-evoked nystagmus, may be the only presenting symptoms of an acute CNS pathology, particularly cerebellar ischemia in the distribution of the AICA or PICA. These patients should be evaluated carefully – even when there are only subtle or no neurological deficiencies at initial presentation. The clinician must identify the fine characteristics of the nystagmus and differentiate between vertiginous events attributed to a peripheral vestibular pathology and CNS pathology. In the latter case, prompt intervention may be required.

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"The vanquished know war. They see through the empty jingoism of those who use the abstract words of glory, honor, and patriotism to mask the cries of the wounded, the senseless killing, war profiteering, and chest-pounding grief"

Chris Hedges (b. 1956), American journalist, author, and war correspondent, specializing in American and Middle Eastern politics and societies