Sensorineural Hearing Loss Due to Vertebrobasilar Artery Ischemia – Illustrative Case and Literature Review

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Abstract

Acute sensorineural hearing loss is commonly caused by peripheral vestibulocochlear disorders such as sudden deafness, Meniere’s disease, and Ramsay Hunt syndrome, but is rarely due to infarction of the vertebrobasilar artery. In this report, a case of right anterior inferior cerebellar artery syndrome presenting with sudden deafness and vertigo is described in order to feature acute sensorineural hearing loss due to vertebrobasilar artery ischemia, and sensorineural hearing loss due to vertebrobasilar artery ischemia is reviewed and discussed. A 79-year-old man presented with right acute sensorineural hearing loss preceded by occasional, minute-long periods of dizziness without cranial neural symptoms other than vestibulocochlear symptoms. Magnetic resonance imaging (MRI) revealed infarction of the right anterior inferior cerebellar artery territory. The vertebrobasilar artery supplies the vestibulocochlear organ, brainstem, and cerebellum, whose abnormalities are related to vestibulocochlear symptoms. Vertigo is a major symptom associated with vertebrobasilar artery ischemia. Further, acute sensorineural hearing loss is caused by hypoperfusion of the vertebrobasilar artery. Vertigo and/or acute sensorineural hearing loss could be a prodrome of subsequent infarction of the vertebrobasilar artery territory. The artery most often responsible for acute sensorineural hearing loss is the anterior inferior cerebellar artery, whereas ischemia of the basilar artery, the posterior inferior cerebellar artery, and the superior cerebellar artery rarely cause acute sensorineural hearing loss. Patients with acute sensorineural hearing loss who are at a high risk of cerebrovascular disease must be examined with imaging tools such as MRI.

Keywords: Basilar artery; Hearing loss; Vestibular; AICA; Cerebellar; PICA

Introduction

The majority of patients with acute sensorineural hearing loss have peripheral vestibulocochlear disorders such as sudden deafness or Meniere’s disease. However, ischemia of the vertebrobasilar artery can also cause vestibulocochlear symptoms such as sensorineural hearing loss and vertigo [1]. The vertebrobasilar artery supplies the inner ear, brainstem, and cerebellum. Ischemia of the vertebrobasilar artery may lead to infarctions of cerebellar arteries such as anterior inferior cerebellar artery (AICA), posterior inferior cerebellar artery (PICA) syndrome, superior cerebellar artery (SCA), or brain stem infarction. Further, the distributions and origins of the vertebrobasilar artery are various and complex. Symptoms depend on whether neural tracts are included within the ischemic lesion in the brain. Typically, a central lesion results in cerebellar symptoms or cranial nerve deficits as well as vestibulocochlear nerve and sensorimotor symptoms. Unless neuronal deficits other than sensorineural hearing loss and vertigo are present, hearing loss due to ischemia of the vertebrobasilar artery is very similar to peripheral vestibulocochlear disorders such as sudden deafness or Meniere’s disease [1]. Acute sensorineural hearing loss occurs at an estimated incidence of approximately 5 to 20 per 100,000 persons per year [2, 3]. The incidence of vertebrobasilar artery infarction in which patients initially present with acute sensorineural hearing loss is 1.2–1.4%, and 1.0–1.2% of these patients show only vestibulocochlear symptoms without other neurologic deficits [4, 5].

The aim of the article is to reveal the characteristics of acute sensorineural hearing loss caused by vertebrobasilar artery ischemia. Therefore, a case of right acute sensorineural hearing loss with vertigo due to AICA syndrome, which has symptoms similar to peripheral vestibulocochlear disorders such as sudden deafness with vertigo, is presented, and the vertebrobasilar artery anatomy and various causes of hearing loss associated with vertebrobasilar artery ischemia are reviewed and discussed.

Illustrative Case

A 79-year-old man had experienced occasional, minute-long periods of dizziness for 8 months. He presented with sudden onset right hearing loss, vertigo, nausea, and vomiting. Pure-tone audiometry showed profound sensorineural hearing loss on the right (Figure 1A). He exhibited left-beating gaze and positional nystagmus with a counterclockwise component, and the stepping test revealed a deviation reaction toward the right side. The finger to nose, knee to shin, and hand pronation supination tests did not show cerebellar disturbance. He did not exhibit any cranial nerve symptoms other than vestibulocochlear symptoms. A routine brain computerized tomography scan, which was performed to rule out cerebral hemorrhage and extensive cerebral infarction, was normal. He was first diagnosed with right-sided sudden deafness. However, he demonstrated right-beating nystagmus when gazing rightward, left-beating nystagmus when gazing straight forward and leftward, and direction-changing positional nystagmus 7 days later. Magnetic resonance imaging (MRI) of the brain was performed and a T2-weighted MRI showed high signal intensity in the right anterior inferior cerebellum, including part of the right tonsil (Figure 2). A T1-weighted MRI showed low signal intensity in the same area. An electronystagmogram revealed right-sided canal paresis of 83% by the caloric response and loss of visual suppression by left-sided ice water stimulation. He was diagnosed with right acute sensorineural hearing loss due to vertebrobasilar artery ischemia.
loss due to right AICA syndrome. An electrocardiogram revealed atrial fibrillation. He was treated with heparin sodium followed by ticlopidine hydrochloride and warfarin potassium. Pure-tone audiometry showed that his hearing levels on the right side had recovered to the same level as that on the opposite side 1 month later (Figure 1B). An MRI showed improvement of the ischemic area 2 months later.

Review of the literature

Previous cases presenting with sensorineural hearing loss without cranial nerve deficits in vertebrobasilar artery ischemia are shown in (Table 1) [4–29]. Sensorineural hearing loss due to AICA syndrome is usually ipsilateral (88%) [6–19], whereas sensorineural hearing loss due to ischemia in the territory of the PICA is also ipsilateral but very rare [20,30]. Although SCA syndrome does not usually lead to hearing loss, contralateral hearing loss has been rarely reported [23,31]. Basilar artery occlusion rarely results in hearing loss, and hearing loss is usually bilateral in these cases [4,5,24–29,32]. Hearing loss has been demonstrated in 0–31% of patients with vertebrobasilar artery insufficiency [33–40].

Discussion

Vertebrobasilar artery anatomy

The internal auditory artery, which supplies the cochlea, usually originates from the AICA and occasionally from the PICA ramifying from the rostral vertebral artery (VA) or the caudal basilar artery (BA) [41]. A major anastomosis between the AICA and the PICA is a common variant [42]. The AICA can arise from either side of the caudal basilar artery (BA) [33]. Many variations exist regarding from where the AICA and PICA arise from the vertebrobasilar artery, and the branches of the AICA have various multiple anastomoses from the lateral medullary artery. However, the inner ear artery is the final artery. The pontes consist of anterior, lateral, and posterior parts, and the BA, SCA, and the AICA irrigate these territories [43]. The anteromedial and anterolateral pontine arteries, which arise from the BA, supply the anterior part of the pons, whereas the lateral pontine artery, which arises from the BA and AICA branches, supplies the lateral pons. The lateral pontine artery territory contains the lateral lemniscus, the superior olive, and the lateral sides of the corticospinal tract. The AICA branches supply areas containing the superior vestibular nucleus and the lateral lemniscus. The posterior pons is irrigated by the SCA branch [43]. Typically, the AICA branches from the first or middle third of the BA and first passes the lateral pons, and then the rostrolateral artery and the caudomedial artery. The internal auditory artery, which is a terminal artery, originates from the superior lateral branch of the AICA and perfuses the inner ear. The superior lateral branch sends branches out to the adjacent pons and extends to the flocculus and anterior inferior cerebellum. The inferior medial branch of the AICA perfuses the lateral pons and extends to the flocculus. The superior lateral branch and inferior medial branch often anastomose the PICA [44].

The internal auditory artery bifurcates the common cochlear artery and the anterior vestibular artery; subsequently, the common cochlear artery bifurcates the main cochlear artery and the vestibulocochlear artery, which is divided into the posterior vestibular artery and the cochlear ramus [8,41,45,46]. The common cochlear artery perfuses the apical three-fourths of the cochlea and the cochlear ramus perfuses the basal area on the final fourth of the cochlea. The anterior vestibular artery perfuses the utricle, which is the superior part of the saccule, and the anterior and horizontal semicircular canals. The inferior part of the saccule and the posterior semicircular canal are perfused by the posterior vestibular artery [8,45,46].

Hearing loss originating from infarction of the AICA territory

The AICA supplies the caudolateral pons, which includes the vestibular nucleus, cochlear nucleus, superior and inferior vestibular nerves, and the cochlear nerve. Infarction in the territory of the AICA is called AICA syndrome [47–53]. The major symptoms are vertigo,
nystagmus, gait and limb ataxia, Horner’s syndrome, crossed sensory signs, ipsilateral hearing loss, tinnitus, ipsiversive facial hemianesthesia, and facial palsy [47–53]. A neurotological dysfunction with vertigo and/or hearing loss suggests infarction in the territory of the AICA [9–19,54,55]. However, cases in which all of these symptoms are rare, and the symptoms of each case vary and depend on the ischemic area. Infarction in the territory of the AICA often causes canal paresis and sensorineural hearing loss [9–19, 49, 54,55]. Lee et al. reported that 90% of patients with AICA syndrome have acute sensorineural hearing loss and vertigo [10]. Transient ischemic attack due to thrombosis or vessel spasm sometimes occurs as a prodrome [49,53]. However, in the prodromal stage, cochlear symptoms such as hearing loss are minor, whereas vertigo is a major symptom [53]. Canal paresis and hearing loss are commonly caused by ischemia in the inner ear [54]. After a long follow-up period, 81% of these patients show improved hearing [34] and 67% of these patients show improvement of canal paresis [56].

**Hearing loss originating from infarction of the PICA territory**

Ischemia in the territory of the PICA causes lateral medullary syndrome and infarction of the inferior cerebellum [57,58]. Cerebellar infarction in the territory of the PICA leads to dizziness, gait unsteadiness, and nystagmus. Ischemia of the PICA does not typically result in auditory symptoms because the auditory pathway is not generally involved in the territory of the PICA [59]. However, sensorineural hearing has been reported to occur in the case of infarction in the territory of the PICA (PICA syndrome) in rare cases [20,30,31,60]. Lee found that only 1% of patients reported sudden deafness because of a non-AICA territory vertebrobasilar ischemic stroke [30]. In his study, sudden deafness due to non-AICA territory vertebrobasilar ischemic stroke was mostly associated with infarction in the territory of the PICA, especially the medial branch [31,58,61]. These rare cases of sensorineural hearing loss are caused by variations in the origin vessels of the internal auditory artery. The internal auditory artery originates most often from the AICA (80–83%), second most often from the BA (17%), and least often from the PICA (2–3%) [41,62,63]. These rare cases of sensorineural hearing loss most likely occur when the internal auditory artery furcates from the PICA.

**Hearing loss originating in infarction of the SCA territory**

SCA syndrome generally does not cause hearing loss [64] because the internal auditory artery does not originate from the SCA. However, although ischemia in the territory of the SCA does not cause peripheral sensorineural hearing loss, it very rarely results in retrocochlear hearing loss [23,31]. The impaired auditory pathway is presumably the lateral lemniscus. The auditory pathway projects from the cochlea to the auditory cortex via the cochlear nerve, cochlear nucleus, superior olivary nucleus, lateral lemniscus of the pons, and inferior colliculus. The main pathway crosses the brainstem at the level of the superior olivary nucleus, and then ascends toward the auditory cortex via the lateral lemniscus of the pons and inferior colliculus. Therefore, impairment of the lateral lemniscus leads to contralateral sensorineural hearing loss [23,31,65].

**Hearing loss due to basilar artery occlusion**

Occlusion of the basilar artery is a lethal condition, with a mortality rate ranging from 40% to 86% [22]. Vertigo, nausea, headache, and motor and oculomotor deficits are the common symptoms [66]. Basilar artery occlusion rarely results in hearing loss [4,5,25–29,32,66], and the reported cases of hearing loss due to basilar artery occlusion are generally bilateral [4,5,24–29]. Unilateral hearing loss due to basilar artery occlusion is very rare [29,32], and these patients generally have ischemic lesions within the cerebellum or the cerebellar artery territories, such as the AICA or PICA, as revealed by MRI [5,24–28]. However, 1 case with unilateral vestibulocochlear symptoms, including unilateral sensorineural hearing loss due to basilar artery occlusion, did not show ischemic lesions in the AICA territory, PICA territory, or cerebellum by T1-, T2-, and diffusion-weighted MRI [25]. In this case, MRI revealed an ischemic lesion only within the pontine artery territory [25]. In contrast to vertebrobasilar insufficiency (VBI), BA occlusion usually leads to more extensive ischemic lesions and causes bilateral hearing loss [4,5,24–26, 28] or unilateral hearing loss [29]. Temporal bone histopathology in patients with occlusion of the VA or BA showed degenerative changes in the unilateral labyrinth and vestibulocochlear nerve [32]. Therefore, hearing loss associated with BA occlusion may be due to degenerative changes in the unilateral labyrinth and vestibulocochlear nerve. The brainstem area or the cerebellum is usually impaired because of BA occlusion. The auditory pathway projects from the cochlea to the auditory cortex via the cochlear nerve, and continues to the cochlear nucleus, superior olivary nucleus, and lateral lemniscus of the pons and inferior colliculus. Hypoperfusion in the territory of the BA may impair the central pathway in addition to causing peripheral sensorineural hearing loss.

**Hearing loss due to vertebrobasilar artery insufficiency**

Because the internal auditory artery is the final artery, the cochlea is considered susceptible to ischemia due to VBI and occlusive disease [8,32,67–70]. VBI is sometimes associated with hearing loss and/or tinnitus [33–40,62,71,72]. Subjective hearing loss has been reported in 0–31% of patients with VBI [33–40], whereas unilateral hearing loss occurs in 7% of patients and bilateral hearing loss is rare (0.5%) [34, 40,71]. Yamashita et al. reported that the symptoms associated with VBI include headaches, tinnitus, visual dysfunction, hearing loss, extremity weakness, unsteadiness, extremity numbness, drop attacks, dysarthria, loss of consciousness, facial weakness, and hoarseness. Cochlear symptoms such as hearing loss and/or tinnitus were found in 36% of VBI patients, with tinnitus and hearing loss reported in 30% and 21% of these patients, respectively [33]. Ischemic lesions associated with hearing loss involve, in order of descending frequency, the AICA territory, the PICA territory, and the brainstem territory [71]. Audiological examination indicated that the major involved site was the cochlea and that retrocochlear hearing loss was minor [33,34]. In the case of cochlear hearing loss, the auditory brainstem response was generally normal or the absolute latencies of
all waves were delayed, but the interpeak latencies of waves I-III-V were normal [33,34,62]. The interpeak latencies of waves I-III-V are prolonged in patients with retrocochlear hearing loss [33,34,62].

Retrocochlear hearing loss is associated with ischemic lesions that involve the central auditory pathway [34,79]. Hypoperfusion to the vestibular labyrinth probably disturbs the vestibular function. The following cerebellar lobules have oculomotor functions: the vermis lobules VI and VII, the Crus I and II of the ansiform lobule, the hemisphere of the simplex lobule, dorsal paraflocculus, ventral paraflocculus, flocculus, uvula, nodulus, caudal dentate nucleus, lateral posterior interposed nucleus, and caudal pole of the fastigial nucleus [80–88]. The flocculus, uvula, and nodule are connected to the vestibular nuclei [82, 87–91], and the purkinje cells of the cerebellum project inhibitory fibers to the vestibular nuclei and cerebellar nuclei [82,87,91]. Disturbances in these areas sometimes cause vertigo and dizziness, which are similar symptoms to those of acute labyrinthine dysfunction.

Features of prodromal vestibulocochlear symptoms

Infarction in the territory of the AICA rarely presents with symptoms that are similar to those of pure peripheral vestibulocochlear disorders such as sudden deafness. The case presented in this study is similar to pure peripheral vestibulocochlear disorders because clear cranial nerve symptoms other than vestibulocochlear symptoms were
absent. However, acute sensorineural hearing loss in this case of AICA syndrome was preceded by episodes of recurrent minute-long dizziness. The feature of prodromal recurrent minute-long dizziness was different from the characteristics of typical peripheral vestibulocochlear disorders such as Meniere’s disease, vestibular neuritis, and sudden deafness with vertigo because vertigo is characterized by hour- or day-long for Meniere’s disease and day- or week-long for vestibular neuritis and sudden deafness. The prodromal recurrent minute-long vertigo of vertebrobasilar artery ischemia is concomitant to the feature of TIA. Recurrent attacks of vertigo and/or dizziness and/or tinnitus preceded acute sensorineural hearing loss in 31% of previous reports, and vestibular symptoms such as vertigo or dizziness accompanied or preceded acute sensorineural hearing loss in 87% of previous reports (Table 1). Only 5 cases presented with auditory disturbance without vestibular symptoms. Further, previous reports showed that patients with vertebrobasilar artery ischemia sometimes demonstrate only inner ear dysfunction. The inner ear is susceptible to ischemia because the internal auditory artery, which is the artery of the inner ear, is the terminal artery. Therefore, hypoperfusion of the vertebrobasilar artery is one of the factors responsible for acute sensorineural hearing loss. Acute sensorineural hearing loss with vertigo and/or dizziness could be a prodrome of subsequent infarction of the vertebrobasilar artery territory.

Imaging such as T1-, T2-, or diffusion-weighted MRI or magnetic resonance angiography is suitable for detecting ischemic lesions. However, MRI is not a cost-effective method for examining all patients with acute sensorineural hearing loss because it has a low positive rate (2.4%) and is expensive [92]. Because recurrent vertigo is a major symptom in the prodromal stage of vertebrobasilar artery ischemia [53], patients with recurrent vertigo preceded by acute sensorineural hearing loss should be carefully monitored. Especially, patients with acute sensorineural hearing loss and recurrent vertigo who have risk factors of cerebrovascular disease such as hypertension, exposure to cigarette smoke, diabetes, athero-fibrosis, coronary artery disease, dyslipidemia, carotid artery stenosis, sickle cell disease, postmenopausal hormone therapy, poor diet, physical inactivity, and obesity [93] must be examined with MRI.

Conclusion

Acute sensorineural hearing loss could be a prodrome of subsequent critical infarction due to vertebrobasilar artery ischemia, such as AICA syndrome, PCA syndrome, or basilar artery occlusion. Acute sensorineural hearing loss associated with vertebrobasilar artery ischemia is usually accompanied by vertigo or is preceded by recurrent episodes of vertigo.

References


