CASE REPORT ARTICLE

Sudden SNHL as a sole presentation of brain stem infarction

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Abstract

Background: Although inner ear dysfunction as a prodromal sign to anterior inferior cerebellar (AICA) occlusion before appearance of other neurologic signs is not a rare situation, sudden sensorineural hearing loss (SSNHL) as a sole presentation is quite rare. The very few reports published for AICA occlusion presented with SSNHL were all in old patients with multiple systemic disorders. This report presents a case of AICA occlusion in an otherwise healthy adult young male patient that presents only with SSNHL, vertigo and vomiting. Diagnosis in such situations needs a high index of suspicion. We here recommend MRI to be done in all cases presented with SSNHL.

Key words: sudden sensorineural hearing loss, anterior inferior cerebellar syndrome, Brain stem infarction

Introduction

Very few reports had been published for brain stem infarctions presented only with sudden sensorineural hearing loss [1]. In most of these reports the patient is old with multiple systemic disorders. Our case was a young healthy patient with no systemic illnesses presented only with sudden unilateral hearing loss from anterior inferior cerebellar artery (AICA) infarction.

Inner ear dysfunction in AICA occlusion usually appears as a prodromal sign before other neurological signs appear [2]. In most reported cases, AICA (anterior inferior cerebellar artery syndrome) is usually accompanied by brainstem signs such as crossed sensory loss, lateral gaze palsy, facial palsy and Horner syndrome, in addition to the inner ear symptoms [3].

Sudden sensorineural hearing loss without associated neurological symptoms and signs is typically attributed to viral inflammation of the labyrinth. Sudden hearing loss due to AICA is usually associated with other brain stem or cerebellar manifestations [2].

Case Presentation

35 years old Saudi healthy male patient presented to the emergency department in King Fahad Hospital of the University in Al-Khobar, KSA, complaining of sudden severe dizziness & complete loss of hearing in the left ear, only 5 hours before presentation. Patient is not known to have any chronic medical illnesses.

Condition started 4 days before presentation by very mild dizziness attacks without any associated symptoms. Five hours prior to presentation he suddenly developed severe attack of vertigo, complete loss of hearing in left ear associated with severe tinnitus, mild headache, nausea and repeated vomiting (5-6 times).

No history of loss of consciousness, similar attacks, ear disease, head trauma, family history of similar illness or stroke, no palpitation, chest pain or shortness of breath . Physical examinationshows that the patient was conscious, alert, oriented to time, place and person. Patient was on wheel chair, as he was unable to stand or walk, vital signs were stable. Ears examination show intact normal tympanic membrane both sides, with no spontaneous nystagmus. Cranial nerves were all normal except left 8th nerve. Chest, CVS, abdomen, all showed no abnormality. Upper and lower limbs showed normal motor power & sensation.

Blood investigations including, complete blood count, serum electrolytes, renal and liver functions were all within normal limits. ANA & RF were negative. Audiological assessment shows, left profound SNHL, with bilateral type (A) tympanometry.

Patient was admitted as a case of left sided sudden sensorineural hearing loss of labyrinthine origin. Thin cuts CT scan was asked for the temporal bone and the brain, and proved negative for local pathology in temporal bone and normal brain study. We started our patient on steroid and antiviral treatment.

On the second day the patient condition was markedly improved as regards vertigo. Auditory brain stem responses to click stimuli at 90 dB were absent in the left ear and normal in the right side.

Due to the history of repeated attacks of the vomiting which is unusual in cases of sudden SNHL due to a peripheral lesion and the absence of spontaneous nystagmus we thought that central cause has to be ruled out. MRI and MRA were ordered and neurology consultation was asked for.

MRI revealed infarction involving the left middle cerebellar peduncle and the dorso-lateral pons (figure 1 and 2). MRA shows only absence of right anterior cerebral artery, one of the normal variations of circle of Willis, there was no visible vascular occlusion (Figure 3).

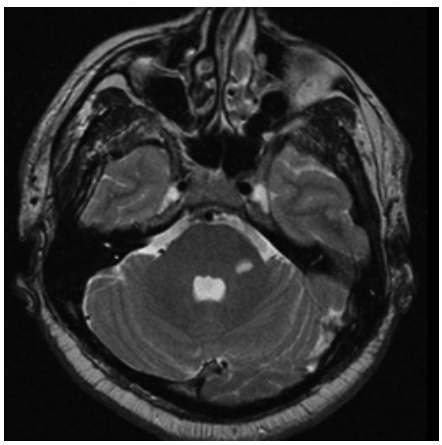

Figure (1) MRI T-2 axial cut shows hyperintense lesion of left side of brainstem.

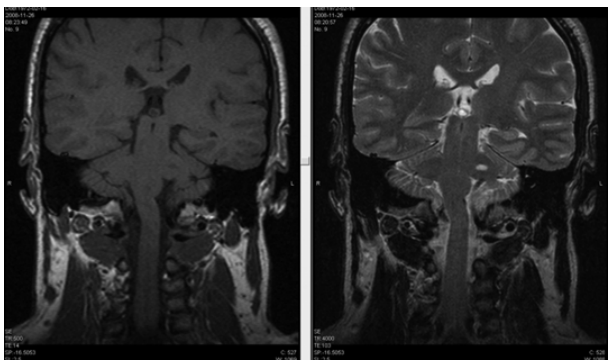

Figure (2) MRI T-1 & T-2 coronal view shows the infarction site.

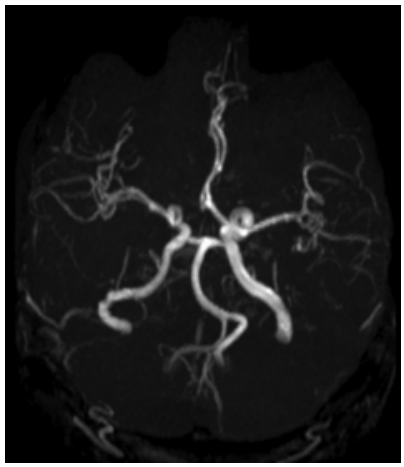

Figure (3) MRA shows circle of Willis with absence of the right anterior cerebral artery (normal variation), no visible site of occlusion.

After MRI finding patient was diagnosed as a case of brain stem infarction. The patient was transferred to the neurology department and he was started on anticoagulant therapy. Repeated audiograms did not show any hearing improvement.

Discussion

Sudden acute attack of vertigo, hearing loss and tinnitus can be caused by peripheral or central origin. Differentiation between both origins is important for diagnosis and management and to rule out possibly of life-threatening condition like stroke.

Our patient presented with sudden attack of severe vertigo, profound hearing loss in the left ear, tinnitus, mild headache, nausea and repeated vomiting. All these symptoms occurred within five hours.

Also our patient was a young, healthy adult with no history of chronic medical illnesses or precipitating factors and there was no cranial nerve involvement, no spontaneous nystagmus, no weakness or sensory deficit in the face or limbs and no other signs of stroke. Although other neurological deficits did not develop, absence of nystagmus and repeated vomiting together with headache were considered unlikely for a peripheral lesion and necessitated further evaluation with MRI.

Clinically, in localizing strokes to the brainstem the "cardinal" features are ipsilateral cranial nerve involvement and contralateral weakness or sensory deficit. MRI is frequently needed to make a specific diagnosis and to separate vascular from tumour aetiologies. Vertigo is a common early symptom of brain stem strokes. Hearing loss is a much less common symptom than vertigo [3].

The extent of anterior inferior cerebellar artery occlusion is extremely variable. Symptoms similar to Meniere's disease (fluctuating hearing, tinnitus, vertigo) can also be caused by impending transient ischaemic attack (TIA) in this distribution (4).

The site of the infarction and the absence of auditory brain stem responses on the right side may indicate affection of the cochlear nuclei, rather than the inner ear. Also this can be supported by the absence of spontaneous nystagmus and the rapid improvement of vertigo.

In patients at high risk for vascular insufficiency, close observation and follow up with MRI is recommended (1). From this case we think that MRI is very important for all patients presented with sudden sensorineural hearing loss.

The internal auditory artery supplies the facial nerve as well as the cochleovestibular nerve in the internal auditory canal, so facial nerve paralysis is commonly described in AICA infarction proximal to the internal auditory artery (2). In our case absence of facial paralysis, absence of nystagmus, the rapid improvement of vertigo and sparing of other cranial nerves nuclei may be explained by the site of infarction affecting mainly the cochlear nuclei in the brain stem.

Inner ear symptoms of sudden SNHL and vertigo as the sole manifestation of AICA is rarely reported (5). Only very few reports occurring in old, hypertensive patients with multiple chronic illnesses, this is the first case to be reported in a young healthy patient.

Conclusion

AICA infarction can present with sudden hearing loss, vertigo and tinnitus without any cranial nerves involvement, hemiplegia or other neurological signs. Any patient with acute vertigo should be evaluated carefully for the possibility of central cause. MRI is very important in the diagnosis and should be done in such cases.

References:

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