

Case Report

Generalized Auditory Agnosia following a Subarachnoid Hemorrhage; A case report

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A 65-year-old male developed a subarachnoid hemorrhage(SAH) caused by a ruptured right internal carotid artery-posterior communicating artery aneurysm. Clipping of the aneurysm, removal of the hematoma, and external decompression were performed. Thereafter, he developed a bacterial meningitis after a spinal lumbar drainage that was done for the treatment of hydrocephalus. He was admitted to our hospital for rehabilitation 4 months after the onset of the SAH. He was still lethargic and delirious at the time of admission. He could not recognize spoken words, environmental sounds or music one month later, but was able to speak and understand the written words. He was diagnosed to have generalized auditory agnosia, based on almost normal pure tone audiometry, otoacoustic emission test, and ABR. Brain CT disclosed a right temporal and frontal lesions, but not in the left side. The eZIS and vbSEE analysis of the SPECT images disclosed a lesion in the left Heschl's transverse gyrus that could not be detected on CT. We emphasized that the detailed analysis of the SPECT images is useful to demonstrate the lesions that can not be detected by CT.

ACTA MEDICA NAGASAKIENSIA 57: 99 - 103, 2012

Keywords: Generalized auditory agnosia, auditory agnosia, eZIS, vbSEE, Subarachnoid hemorrhage

Introduction

Cortical auditory disorders in adults were first described by Kussmaul¹ and Lichtheim². Cortical auditory disorders encompass cortical deafness, auditory agnosia (amusia, environmental agnosia), and pure word deafness, all of which cause difficulty integrating and understanding different types of auditory input (e.g., speech, music, and environmental sounds)³. Most authors agree that specific syndromes do exist with significant variability in clinical presentation. There has long been interest in cortical auditory disorders. Patients with pure word deafness^{4,5} cannot discriminate among spoken words or even distinguish among speech sounds but can label non-verbal sounds and respond appropriately to

intonations of speech. Cortical or central deafness is another variant³. Patients cannot recognize any auditory sounds, although the sensation of having heard sounds is intact (i.e., patients respond to sounds, but then deny having heard them)⁶. Auditory agnosia is another common syndrome within the general category of cortical auditory disorder. A patient with generalized auditory agnosia can neither differentiate spoken words, nor state what produces a specific noise (eg, a blowing whistle), nor recognize music. It can be characterized as an "apperceptive disorder" because the problem involves the inability to perceive patterns rather than the meaning of sounds^{3,7}. Alternatively, a patient can have either isolated receptive amusia, which is the inability to recognize music, or environmental agnosia, which is the

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Received May 15, 2012; Accepted June 8, 2012

inability to recognize nonspeech sounds (eg, dog barking)³. One thing that all of these different syndromes have in common is an injury to the cortical auditory system. This report present a case of generalized auditory agnosia.

Case report

History: A 65-year-old, right-handed male suddenly developed severe headache and vomiting and was admitted to a hospital with a subarachnoid hemorrhage (SAH) that had been caused by a ruptured right internal carotid artery-posterior communicating artery aneurysm. A brain CT scan revealed a hyperintense area in the basal cistern, left Sylvian fissure, and a hematoma in the right Sylvian fissure. Clipping of the aneurysm, removal of the hematoma, and external decompression were thus performed. Thereafter, he developed a bacterial meningitis after a spinal lumbar drainage that was done for the treatment of hydrocephalus. He was admitted for rehabilitation 4 months after the onset of SAH. He was still lethargic and delirious at the time of admission. A general physical examination revealed a normal heart rate and rhythm. His blood pressure was 131/81mmHg. He could not recognize spoken words, environmental sounds or music one month later, but was able to speak and understand written words. Although he was lethargic, he sometimes suddenly spoke loudly or shouted, and thereafter began to

yell at anyone who was around him. He looked to have very mild left hemiparesis, but the deep tendon reflexes were normal, and Babinski reflexes were not elicited. He frequently showed frontal lobe symptoms such as difficulty in inhibiting emotions, anger, and excitement, and a stereotyped behaviors such as continuous wiping of a stain on the desk with a towel. The MMSE score was 22, particular decrease in the scores of time and place orientation. The Kohs Block Design Test could not be administered because of his mental instability. However, he could read newspapers and play shogi (Japanese chess) when he was mentally calm.

Neuropsychological assessment: The following assessments were performed while he was seemed to be in a composed and cooperative mental state.

Standard Language Test of Aphasia (SLTA); This test was performed and the patient completed the test questions as well as he could. The scores for the items associated with hearing such as a word, sentence, and command comprehension could not be determined, however, the items of reading such as word naming, reading of kanji (Chinese characters), the reading of kana (Japanese syllabray), the reading of short sentences, the comprehension of kanji words, and the comprehension of kana words were ;19/20, 5/5, 5/5, 5/5, 10/10, and 10/10, respectively. He was unable to recognize spoken words, but he was able to communicate in writing, and no paraphasia was observed. He could calcu-

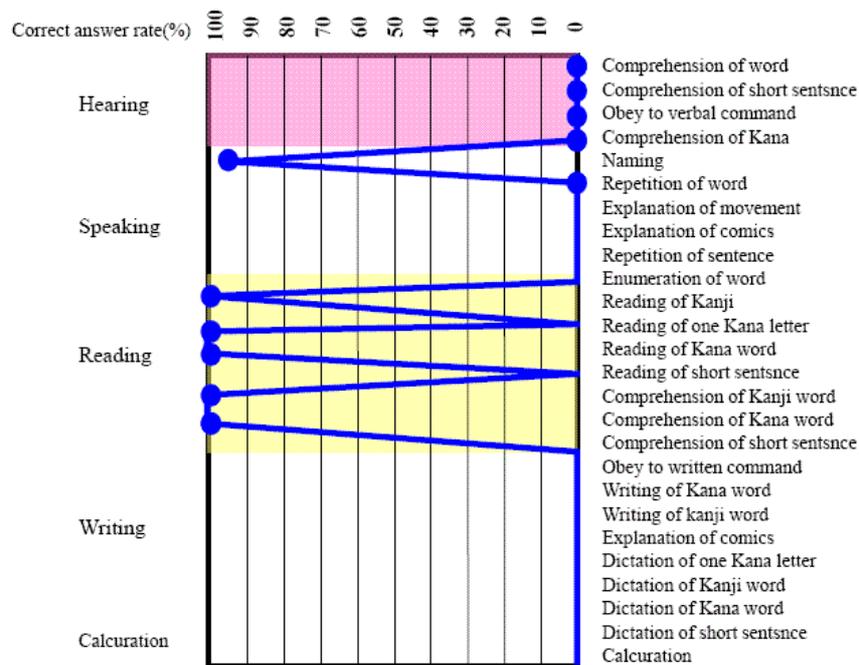


Figure 1. Results of SLTA. The hearing items could not be evaluated, but reading items could be successfully completed and scored. The scores for the hearing items were lost, however, the scores for the reading items were well preserved.

late without errors (Fig.1).

A pure tone audiometry showed 36dB at 1000Hz in the right ear and 57.5dB in the left ear. An examination of the brainstem auditory evoked potential showed normal wave forms and latencies from wave I to wave V (Fig.2). His otoacoustic emission test revealed a very mild hearing impairment in the left ear, and normal hearing in the right ear. The stapedial reflex test was normal. His auditory disorder was therefore considered to be in the cortical auditory systems based on these results.

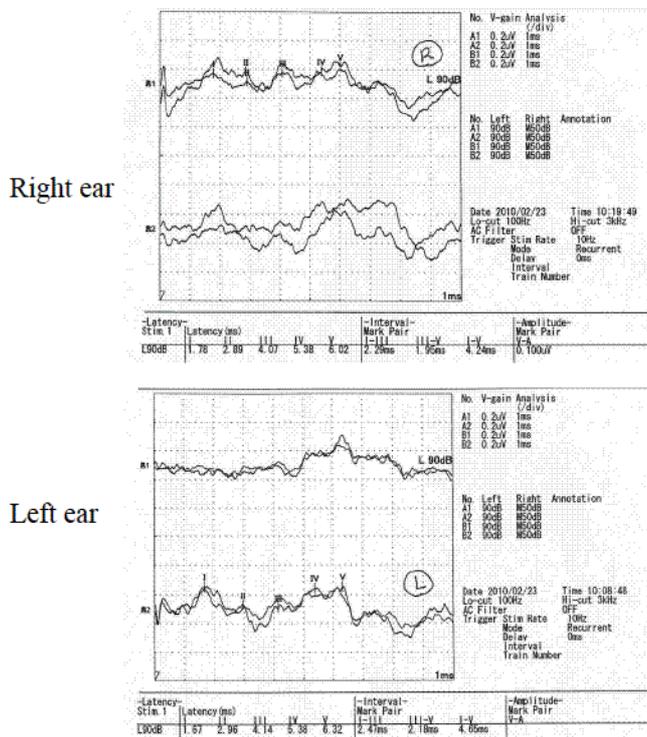


Figure 2. Brainstem auditory evoked potentials. The forms and latencies from wave I to wave V were normal.

A sound recognition test was conducted using a tape presenting familiar, meaningful non-verbal sounds including the sounds of human voices (crying of babies, laughing voice), animal sounds (dog, cat, cow, horse, chicken, crow), instrument sounds (trumpet, drum, telephone), and natural sounds (wind, rain, stream), artificial sounds (street car, car), and music (hatopoppo, momotarou (familiar songs for school children)), while requiring the patient to name these sounds. He could neither comprehend the verbal sounds nor nonverbal sounds, but he could correctly point out the picture card among the 6 cards that was required by written name (Fig.3).

Radiological examinations: A CT scan on admission revealed two lesions, a hypointense lesion in the right



Figure 3. The patient could neither comprehend the verbal sounds nor nonverbal sounds, but he could correctly point out the picture card among the 6 cards that was required by written name. This picture shows that he could identify the picture of a drum that was written on the white board.

anterior temporal region, and a hypointense lesion in the right frontal cortex spreading to the subcortical region. No lesion was found in the left temporal cortex (Fig.4).

123-IMP-SPECT images were analyzed using the easy-Z-score Imaging System (eZIS)⁸ and Voxel-Based Stereotactic Extraction Estimation (vbSEE)⁹

The level 3 images in vbSEE disclosed a decrease in the upper, middle and lower temporal gyri, Heschl's transverse gyrus, inferior parietal lobule, superior parietal lobule, angular gyrus, inferior, middle, and frontal gyri, medial frontal gyrus, superior and middle occipital gyri, spindle gyrus, insular gyrus, parahippocampal gyrus, and uncus in the right side, and left Heschl's gyrus (Fig.5).

Electroencephalogram showed slow waves (δ wave) in the right frontal lobe.

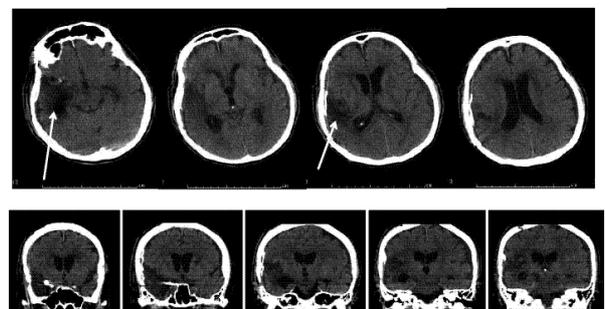


Figure 4. Computed tomography of the brain. A hypointense lesion that spread from the cortex to subcortical region in the Right temporal lobe, and in the frontal lobe.

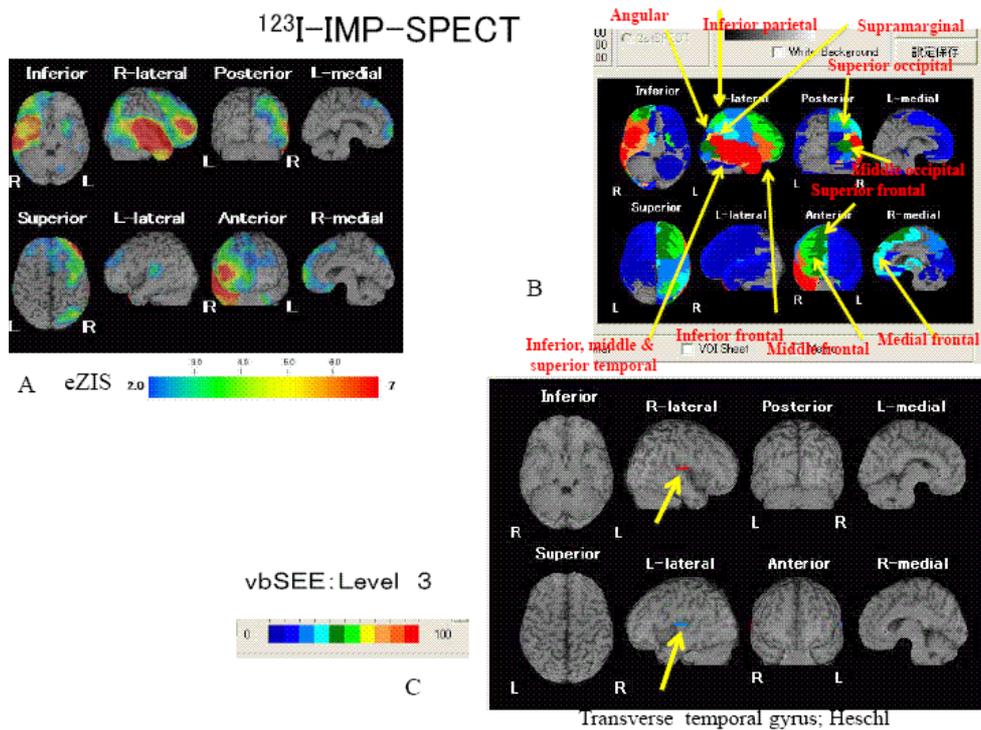


Figure 5. eZIS and vbSEE analysis of SPECT images.

A. eZIS images. A marked decrease in the right temporal and frontal lobes.

B. The level 3 images of vbSEE. Decreased regions are shown in the picture.

C. Bilateral decrease in the superior transverse gyri (Heschl).

Discussion

The patient was lethargic and delirious when he was admitted to the hospital. He could not recognize spoken words or any environmental sounds one month later, but he was able to speak and understand written words. Cortical deafness and auditory agnosia are usually related to each other and are frequently associated with aphasia¹⁰. Auditory signals cannot be perceived in the cortex in cortical deafness, because of damage to both the temporal or temporoparietal lobes including the primary auditory cortex on both transverse gyri (Heschl) and audiometry reveals severe bilateral hearing loss. Cortical deafness was excluded in this patient because there was a mild hearing impairment in a pure tone audiometry. Patients with pure word deafness cannot distinguish spoken words or even differentiate various speech sounds, but they can label nonverbal sounds and respond appropriately to intonations of speech^{4,11}. The issue in this patient was not only limited to the spoken words, but also was associated with the labeling of nonverbal sounds. Sensory aphasia was also distinguished because the patient could communicate through writing, and could comprehend written words. Finally, generalized auditory agnosia was diagnosed because he could neither differentiate spoken words,

nor state what produces a specific noise, nor music, and normal brainstem auditory evoked potential and otoacoustic emission test.

There are two main etiologies of cortical auditory disorders: namely acquired and developmental. Acquired causes, usually in adulthood, are most commonly associated with stroke (bilateral temporal lobe). The main etiology of generalized auditory agnosia in adult is stroke such as cerebral hematoma, cerebral infarction, moyamoya disease, and subarachnoid hemorrhage. Other less common etiologies are brain injury, tumor, encephalitis¹², mitochondrial encephalomyopathy¹³, adrenoleucodystrophy¹⁴, primary progressive aphasia¹⁵, Alzheimer's disease¹⁶, moyamoya disease¹⁷, and carbamazepine medication¹⁸. In principle, any pathology that can affect the cortical function is capable of causing difficulty in the perception of relevant sound stimuli.

Shindou M³ reported 10 cases of generalized auditory agnosia; 3 cases of cerebral infarction, 3 cerebral hematoma, 2 moyamoya disease, 1 SAH, and 1 injury. There are several reports of generalized auditory agnosia that occurred after SAH¹⁹⁻²². SAH dose usually not induce focal lesions. The main mechanism of auditory agnosia in the cases of SAH is cerebral infarction induced by vasospasm or focal hematoma in the regions including the auditory cortex¹⁹⁻²¹.

The underlying anatomical substrates for cortical auditory disorders are thought to be the bilateral temporoparietal areas, especially the superior temporal gyrus of the dominant hemisphere (Heschl gyrus). The middle temporal gyrus, the insula, and surrounding association areas have also been implicated in cortical auditory disorders. The left temporal lobe has commonly been thought to process the perception and production of speech sounds whereas the right temporal lobe appears to control its prosodic and emotional components²³. Recently, researchers have found that the right temporal lobe processes slower acoustic sounds (nonspeech), whereas faster acoustic sounds (including speech) are represented bilaterally.^{24,25} Further, damage to the right hemisphere alone has not been found to cause either speech perception or auditory nonspeech deficits²⁶.

The hypointense lesions on CT in the right temporal and frontal lobe in the current patient may have been an infarction induced by vasospasm. While the left Heschl's gyrus lesion which was not demonstrable on CT, but was revealed by a vbSEE analysis of SPECT may be due to blood in the Sylvian fissure or a complicated inflammatory lesion after the shunt operation.

Therefore, the lesions in the right auditory cortex and its association areas, and the left auditory cortex were assumed to have caused the generalized auditory agnosia observed in this case.

Finally this case demonstrates that both the eZIS and vbSEE analysis of SPECT images is useful to demonstrate the presence of lesions that are not apparent on the CT findings.

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