

Review Article

Ictal Pure Word Deafness with Auditory Hallucination

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Abstract

Pure word deafness is a disorder of auditory verbal comprehension without dysfunction of speaking, reading, or writing. This rare symptom arises from bilateral lesions in the temporal lobes or disconnection between the primary auditory cortex and Wernicke's area. I present a right-handed 33-year-old woman who presented with pure word deafness and complex auditory hallucination with intermittent amnesia. She had no generalized convulsion. There was no intracranial lesion on plain or gadolinium-enhanced magnetic resonance imaging. Electroencephalogram revealed theta waves in the left temporal-occipital region followed by high-amplitude slow wave burst during hyperventilation. Carbamazepine completely ameliorated the pure word deafness and auditory hallucination. This is the first report of pure word deafness resulting from primary complex partial seizure. The epileptic focus might lie along the auditory word processing pathway between the left auditory cortex and Wernicke's area.

INTRODUCTION

Pure word deafness is a disorder of auditory verbal comprehension without dysfunction of speaking, reading, or writing. This rare symptom arises from bilateral lesions in the temporal lobes or disconnection between the primary auditory cortex and Wernicke's area [1]. I report a patient with epilepsy whose predominant symptoms were pure word deafness and auditory hallucination lasting for at least five days. Electroencephalogram showed paroxysmal generalized spike and wave complex during hyperventilation. Anti-epileptic drug, carbamazepine, completely resolved her language problem.

CASE PRESENTATION

A right-handed 33-year-old woman presented with abrupt occipital headache with nausea and sparkling flash. There was no past history of brain injury or neurological disease. Also, there was no family history of neurological disease or sinistrality. She had graduated from university and worked in an office. The neurological examination was unremarkable. Brain computed tomography was normal. A physician in the emergency room gave her non-steroid anti-inflammatory drugs. A week later, she came again complaining that she could not understand what people said. She could hear sounds and read sentences shown to her. She could respond to written questions by speaking. She was referred to the neurology department after two days.

She came into the outpatient clinic walking. She was alert, and there was no meningeal sign. There was no abnormal finding on physical and neurological examinations. Her spontaneous speech was relatively scant but fluent, and involved no phonological or verbal paraphasia. She could not understand what I said, but she

could understand what I wrote. She could read these sentences aloud without any hesitation. She could write several words on my instructions. There was no mistake in her writing. During the interview, there was a moment when she could not read or understand written command. Her mother also had noticed that she had not been able to read words on several opportunities. The patient said that she had amnesic intervals and auditory hallucinations. She was aware that they were hallucinations. The hallucinations were known and unknown melodies, and sometimes the speech of a man. She could recognize the man's verbalizations as language, and not mere noise. She partially remembered what he said, in Japanese. The same melody sounded repeatedly from the left side. Brain magnetic resonance (MR) imaging and intracranial MR angiography were normal.

On the electroencephalogram (EEG) performed the same day, an 11-12 c/s alpha wave was seen in the background. Although photic driving was seen on 12 and 15 c/s stimulation, there was no photosensitivity. During hyperventilation, high-amplitude slow wave burst was evident after 7-8 c/s theta waves appeared in the left temporal-occipital region (Figure 1). Laboratory tests showed normal blood cell counts, liver and renal functions, glucose level, and levels of electrolytes, and there was no inflammatory sign. Immediately, she was treated with 200 mg of carbamazepine. After three days of the treatment, her auditory verbal comprehension was completely improved. However, her auditory hallucinations continued. She could explain precisely how people's speech had sounded: To her, people's speech sounded like a foreign language, and she could not understand what they said at all. She could hear and distinguish the sounds of a telephone ringing and an ambulance. She could hear the melodic

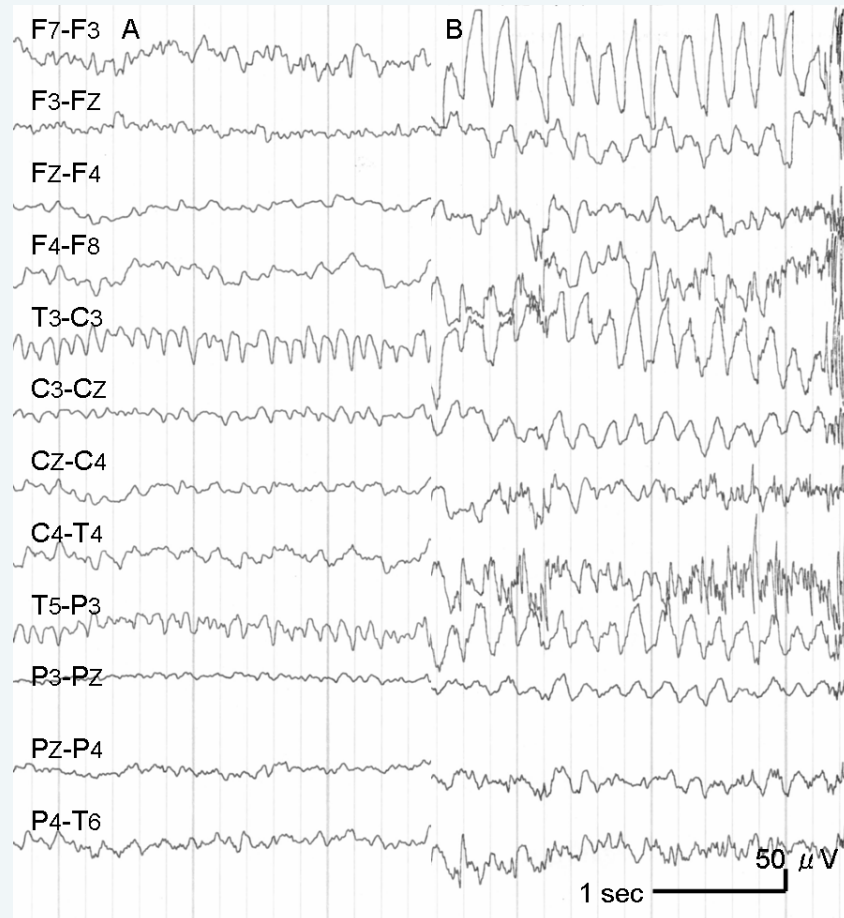


Figure 1 Electroencephalogram of the present patient performed during word deafness and auditory hallucination. After the theta waves appeared in the left temporal-occipital region (A), high-amplitude slow wave burst developed (B).

sound of a microwave oven alerting her to the completion of cooking and understand what it meant. But it sounded like a mere sound. The symptom of word deafness lasted at least for five days. She could have been having an epileptia partialis continua.

Auditory brainstem response and gadolinium-enhanced brain MR imaging were normal. Carbamazepine (400 mg/day) completely suppressed her auditory hallucinations, word deafness and EEG abnormalities.

DISCUSSION

Although her auditory function and neurolinguistic function during the attack was not precisely assessed, this patient's symptom can be summarized as pure word deafness associated with epilepsy. The intermittent difficulty in reading or amnesia might have been secondary generalization of the partial epilepsy. Since she could recognize non-verbal sounds, auditory agnosia and cortical deafness could be excluded. Ictal deafness reported by Ghosh et al. [2] was also ruled out because she could understand non-verbal sounds. Although the defective comprehension was not limited to speech, but involved musical melody, some cases of pure word deafness accompanied disturbance in musical discrimination [3]. Our patient also complained of complex auditory hallucination. This auditory hallucination might have

been epileptic phenomenon or alternatively release phenomenon of the auditory cortex.

This rare symptom of word deafness arises from bilateral temporal lobe lesions or left temporal lobe lesion. Word deafness resulting from seizure had been reported in only one case of bilateral temporal lobe lesions [4]. The case reported by Fung et al. [4] had a recent hemorrhagic infarction in the left midtemporal cortex. The patient also complained of auditory hallucination ("muffled voice") before an episode of intermittent word deafness or "incomprehensible distant voices" at the onset of EEG seizure. Recent functional neuroimaging study revealed the activation of bilateral superior temporal sulci, extending more posteriorly on the left, during listening to sinewave speech [5]. Taken together, there is a possibility that the focus of epilepsy in the presented case and Fung's case could lie along the auditory word processing pathway. The EEG findings support this hypothesis. Since the presented patient could understand written commands, Wernicke's area could be preserved intact. The dissociation between disturbed auditory verbal perception and preservation of the comprehension of hallucinated language suggests that the word deafness in the present case resulted from disconnection of the primary auditory area and Wernicke's area. This is analogous to visual command hallucination in a patient with pure alexia [6]. While the primary activation of so-called

auditory word form area (AWFA) might have produced complex auditory hallucination, the functional disconnection of afferent fibers selective from AWFA to Wernicke's area might have caused the pure word deafness.

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