

NOTE

PURE WORD DEAFNESS DUE TO LEFT HEMISPHERE DAMAGE

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INTRODUCTION

Pure word deafness is defined as a selective deficit of auditory language comprehension, and is distinguished from cortical auditory deficits for the absence of deficits in understanding music and environmental sounds. Lichtheim (1885) suggested that this syndrome was due to the isolation of Wernicke's area from auditory linguistic input following damage to the subcortex of the left temporal lobe. Liepmann and Storch (1902) supported this view in the light of autopsy studies and more recently Geschwind (1965) has advocated a similar view. Thereafter, many cases labeled as pure word deafness have been reported with anatomical documentation of either unilateral left or bilateral cortical/subcortical damage to the temporal lobe. The number of cases that comply with the above definition of pure word deafness is, however, much smaller and in only a few has the location of the lesion been precisely delineated (Henschen, 1917; Hamanaka, Asano, Morimune et al., 1980; Kamei, Nakane, Nishimaru et al., 1981; Metz-Lutz and Dahl, 1984; Yaqub, Gascon, Alnoasha et al., 1988).

We report a case of pure word deafness, following a left thalamic hemorrhage, whose deficits were selective for auditory language comprehension and whose lesion was identified by means of computed tomography (CT), magnetic resonance imaging (MRI) and positron emission tomography (PET).

CASE REPORT

On October 17, 1982, a 55 year-old right-handed man with a 10 year history of hypertension was admitted to the Chiba Emergency Medical Center following onset of headache, disturbance of consciousness, right hemiparesis and aphasia. He had graduated from junior high school, from which time he had been employed as an automobile mechanic. CT obtained on admission showed left thalamic bleeding. The hematoma had penetrated the third ventricle and extended rostrally to the left centrum semiovale and caudally to the midbrain.

Subsequently, the deficits of consciousness and paresis gradually subsided and no abnormalities of speech remained. However, although he could hear the voices of other people, he realized that he was almost totally incapable of understanding what was being said. Since these symptoms persisted, he was transferred to the Department of Neurology of Chiba University Hospital on July 21, 1984. On examination, consciousness was normal and he was fully oriented. Visual acuity was normal, but he showed lower right quadrantopsia on confrontation. His visual reflexes to light were brisk. Vertical eye movements were limited. There was a slight diplopia when the gaze was to the right. Swallowing and articulation were normal. On the right there was a hemiparesis with increased tendon reflexes and a positive Babinski sign and sensory deficits including the right side of the face.

Neuropsychological examination did not reveal ideational apraxia, deficits of constructional behavior, hemispatial neglect or visual agnosia. WAIS verbal IQ was 89, performance IQ was 90 and total IQ was 89 (Because of severe difficulty in auditory comprehension, presentation of some problems was done visually).

CLINICAL SYMPTOMS AND EXAMINATION RESULTS OF THE PURE WORD DEAFNESS

Clinical Symptoms

Spontaneous speech was fluent with normal speed and intonation. The amount of speech and voice volume were unchanged from the premorbid state. There were no word substitutions or paraphasias. Reading aloud, reading comprehension and spontaneous writing were normal. In contrast, there were severe deficits in auditory verbal comprehension and he was almost totally incapable of understanding individual words.

Subjectively, he could discriminate a human voice from other sounds, but he stated that it sounded like a foreign language, except for his own speech which sounded normal. The hearing abnormality was particularly strong when he could not see the speaker (e.g. when listening to the telephone, radio or a tape cassette) and with unfamiliar voices. Clearly articulated words and words frequently used in daily conversation were easier to understand than poorly articulated and uncommon words.

He was however capable of identifying the number of syllables in a word and the number of words in a sentence. Moreover, if a word was pronounced slowly or was repeated, his understanding improved. Improvements also obtained with lip-reading.

In contrast, he could immediately identify environmental sounds, such as animal cries or a ringing telephone, as well as musical instruments and the names of musical melodies. All of the above symptoms remained unchanged throughout the period of testing and neuro-radiological examination.

*Examination Results (September 1984 to December 1986)**Auditory Testing (September 15, 1984)*

In pure tone audiograms, he had moderate deficits at high frequencies, but not in the range of conversational sounds, except for a slight decrease at 2000 Hz. The right ear was mildly worse than the left at all frequencies above 250 Hz. Auditory brainstem and slow vertex evoked potentials were normal.

Western Aphasia Battery (WAB) (December 10, 1986)

(Lip-reading was permitted when auditory comprehension, oral repetition and writing to dictation tests were given).

Auditory comprehension and repetition were markedly impaired, as well as writing to dictation (Figure 1). In spontaneous writing, he made only two errors (once replacing a Kana character by another and once omitting a Kana character) out of 7 sentences, but otherwise his writing showed no abnormalities. Speech, reading and object naming were normal.

Auditory Recognition Tests (September 15, 1984)

Speech sounds, environmental sounds and music recognition were tested following the methods of Shindo, Kaga and Tanaka (1981). Five separate tests of speech recognition were carried out.

(1) *Mono-syllable Identification.* The patient was required to repeat the 53 CV syllables of the Japanese writing system pronounced in random order by the examiner.

(2) *Word Recognition.* The examiner read out 20 words such as SAKE and TAKE, and the patient pointed at the corresponding card presented with three distractors and showing both a picture and the printed word. The same procedure was followed for tests (3) through (5).

(3) *Long and Short Vowel Recognition.* Twenty trials involving pairs of words differing for the length of a vowel, such as KAHDO and KADO, were given.

(4) *Accent Recognition.* Twenty trials involving pairs of words differing for accent such as HASHI and HASHĪ were given.

(5) *Prosody Recognition.* The patient was required to identify from prosody the mood expressed by 14 short sentences.

These tests were given separately to the left and the right ear.

In the environmental sound test, the patient listened to an audio tape on which a total of

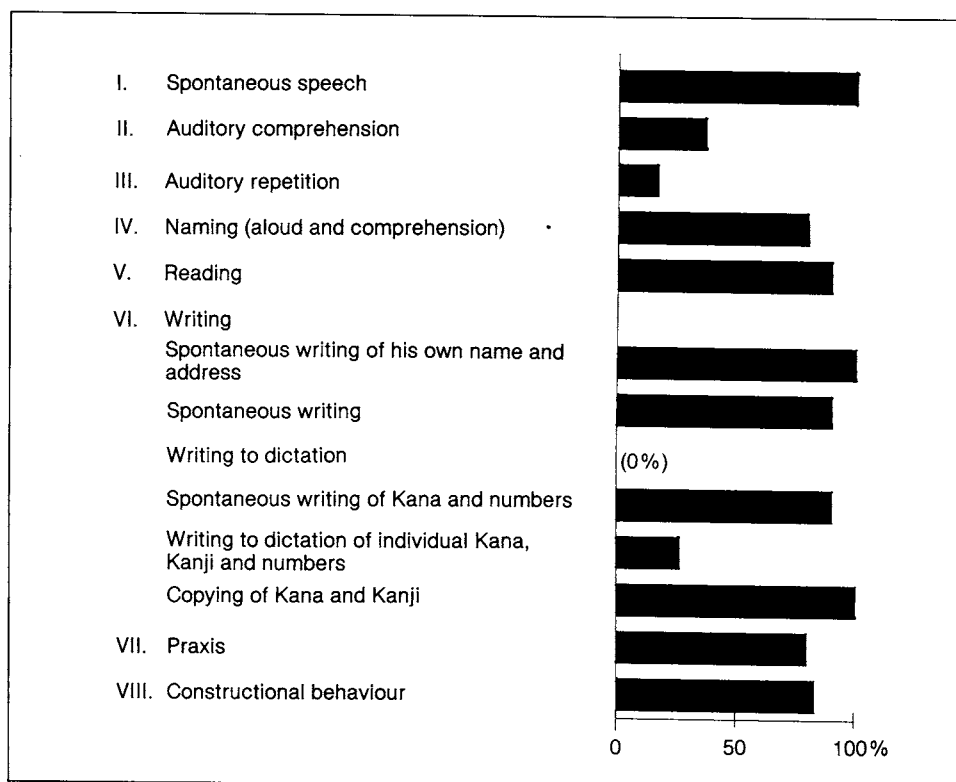


Fig. 1 - Western Aphasia Battery. Notable deficits of auditory comprehension, repetition and writing from dictation. The performance was normal in the other subtests.

24 familiar environmental sounds, such as a ringing telephone, a cat's meow, the sound of wind, etc., had been recorded. He was required to name the sound and to point to an appropriate picture corresponding to it, presented with three distractors (Kaga, Shindo and Sugishita, 1987). For human voice recognition, the spontaneous conversation of seven people (his wife, son, daughter, the patient himself, one female and two male examiners) was recorded and he was required to identify the voices by naming.

For the music recognition test, tape recordings of the following six categories were made.

(1) *Pitch Discrimination*. Two notes of different pitch were presented sequentially and the patient was required to identify the higher.

(2) *Loudness Discrimination*. Two notes of different loudness were presented sequentially and he was required to identify the louder.

(3) *Rhythm Discrimination*. Two short rhythmical sequences were presented one after the other and he was asked whether they were same or different.

(4) *Melody Discrimination and Identification*. Two short melodies were played one after the other and he was asked whether they were the same or different. He was also played melodies that were familiar to him and asked to point to one of four drawings representing the theme of the melody.

(5) *Musical Instrument Identification*. After hearing the sound of a musical instrument, he was required to point to one of four pictures identifying the instrument.

(6) *Musical Mood Identification*. After listening to one of five short melodies (two varieties of energetic music, one sad, one happy and one melodious tune), he was required to choose among three words that corresponding to the mood of the music.

TABLE I
Tests of Auditory Perception

	Correct response	
	L. ear	R. ear
Speech Recognition Tests		
1. mono-syllable identification (53 CV phonemes)	2%	2%
2. Word recognition (20 words)	50%	40%
3. Long and short vowel recognition (20 words)	70%	50%
4. Accent recognition (20 words)	55%	65%
5. Prosody recognition (14 sentences)	0%	0%
Environmental Sound Recognition Tests		
1. Environmental sound identification (pointing)	24/24 (100%)	
2. Environmental sound identification (naming)	21/24 (88%)	
3. Human voice identification	5/7	
Music Recognition Tests		
1. Pitch discrimination	5/8	
2. Loudness discrimination	4/4	
3. Rhythm discrimination	1/4	
4. Melody discrimination	3/6	
Melody identification	10/11	
5. Musical instrument identification	8/10	
6. Musical mood identification	5/5	

Table I shows that, with speech sounds, he manifested severe deficits in the monosyllable test and the prosody test, and moderate deficits in the other tests. His errors tended to be phonemic rather than semantic. In contrast, with environmental sound tests, pointing to pictures was 100% correct and naming 88% correct. With the exception of one confusion between his wife's voice and one of the female examiners' voice, he was flawless in the recognition of human voices. He showed moderate deficits in the rhythm and melody tests, but good performance in the other musical tests.

Neuroradiological Evidence

CT scan, obtained on January 29, 1987, showed a low density area (LDA) that extended from the left thalamus and posterior internal capsule to the white matter of the left temporal and parietal lobes.

In a horizontal MRI image passing through the thalamus [IR method, repeat time (Tr) of 2080 msec and inversion time (Ti) of 500 msec (March 29, 1985)], the lesion of the white matter was more clearly delineated and an area of low signal intensity (LSI) was seen in the left temporal white matter running next to the lateral wall of the left lateral ventricle (Figure 2). The left auditory radiations were damaged. A region of LSI was evident in the left thalamus and internal capsule. In a more rostral section, an area of LSI was seen in the white matter of the temporal and parietal lobes running lateral to the left lateral ventricle and extending to a portion of the centrum semiovale above. There was no notable abnormality of the left temporal cortex and in the right hemisphere. In a coronal section passing through the posterior part of the temporal lobe (Figure 3a), LSI was seen in the left temporo-parietal white matter extending from the wall of the left lateral ventricle to the centrum semiovale. In a sagittal section 30 mm to the left of the midline (Figure 4a), the left temporal and parietal white matter above the lateral ventricle showed LSI. Using the SE method, high signal intensity (HSI) regions were seen in the same areas [Tr = 2080 msec, echo time (Te) = 80 msec] (Figures 3b and 4b).

In a PET study using the ^{15}O -steady state method (March 4, 1987), cerebral metabolic rate for oxygen (CMRO_2 , Figure 5) and cerebral blood flow (CBF) were measured. The CMRO_2 of the left thalamus was markedly reduced, and that of the white matter of the left temporal

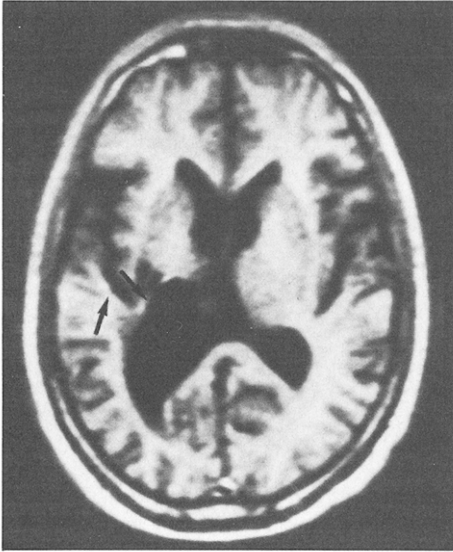


Fig. 2 - Horizontal MRI. A low signal intensity area encroaches upon the left thalamus, posterior internal capsule and white matter of the left temporal lobe. The large arrow indicates the left primary auditory area. The thin arrow indicates damage to the left auditory radiations.

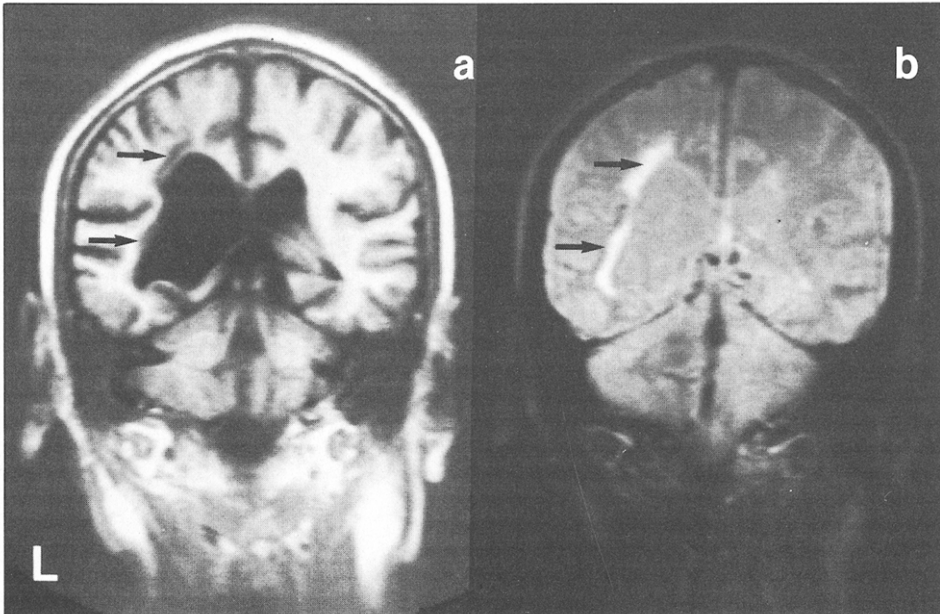


Fig. 3 - Coronal MRI using the IR method (a) and the SE method (b). The lesion running along the cerebral ventricle in the white matter of the left temporal and parietal lobes is seen as a low signal intensity area using the IR method and as a high intensity area using the SE method (arrows).

and parietal lobes was also low. Although the oxygen consumption of the left auditory cortex and the superior temporal gyrus, including Wernicke's area, was somewhat lower than that on the right (3.48 vs. 3.95 ml/100 ml/min), both were within a normal range. Similar CBF findings were obtained.

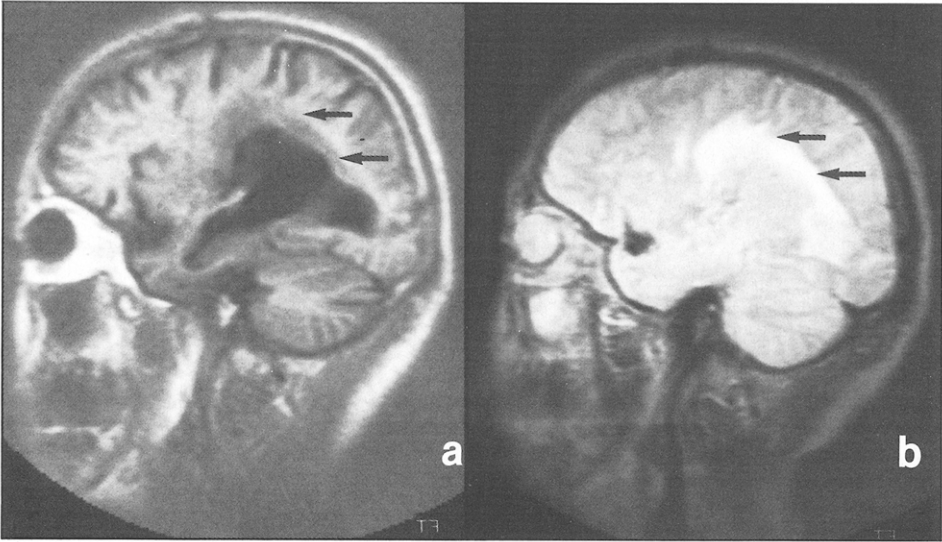


Fig. 4 - Sagittal MRI using the IR method (a) and the SE method (b). The lesion in the white matter of the left temporal and parietal lobes above the lateral ventricle is seen as a low signal intensity area in the IR method and as a high signal intensity area in the SE method (arrows).

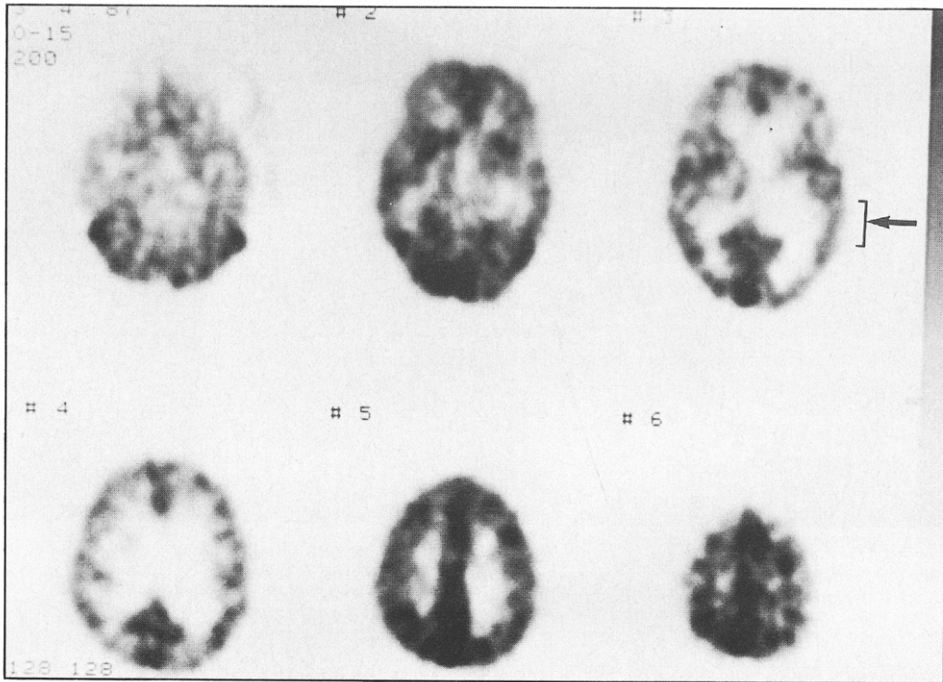


Fig. 5 - PET (CMRO₂) study shows a marked decrease in the left thalamus, and a decrease centering on the white matter of the left temporal and parietal lobes. Noteworthy, however, is the fact that oxygen consumption at the posterior half of the superior temporal gyrus (arrow) is within a normal range.

DISCUSSION

There are inconsistencies in the terminology used to discuss deficits of higher auditory functions. In the present work, we define auditory deficits of language as word deafness, those of environmental sounds (meaningful non-language sounds) as auditory sound agnosia (= auditory agnosia) and those of music as sensory amusia. When all three symptoms are present, the syndrome is called cortical auditory deficits, while deficits of auditory perception (deafness) due to cerebral lesions are referred to as cortical deafness.

The concept of pure word deafness was introduced by Kussmaul (1877) and Lichtheim (1885) to define a condition in which spontaneous speech, reading and writing were normal, but there were deficits in the auditory comprehension of language. It was regarded as a pure form of aphasia and contrasted with other forms of aphasia in which the "internal language" also was disturbed. In recent years, the selective impairment of auditory language comprehension has been contrasted with the intact recognition of environmental sounds and music (Bauer and Rubens, 1985).

Most previous case reports of word deafness present difficulties of interpretation either because they were in fact cases of cortical auditory deficits or because the examination was not sufficiently complete to discriminate pure word deafness from cortical auditory deficits. There are only seven cases which meet the criteria for the diagnosis of pure word deafness (Henschen, 1917; Hemphill and Stengel, 1940; Okada, Hanada, Hattori et al., 1963; Hamanaka et al., 1980; Kamei et al., 1981; Metz-Lutz and Dahl, 1984; Yaqub et al., 1988). In addition, there have been reports of selective deficits in the recognition of environmental sounds (Spreeen, Benton and Fincham, 1965; Fujii, Fukatsu, Watabe et al., 1990) and of sensory amusia (Edgren, 1985; Kohl and Tschabitscher, 1953).

In our case, (1) hearing was normal within the frequency range of normal conversation, except for a slight reduction at 2000 Hz, (2) writing was almost normal, except for rare paragrammas, and spontaneous speech and reading were intact, suggesting that the internal language processes were not impaired, (3) environmental sound and music recognition was normal. For this reason, our case can be considered a particularly "pure" instance of word deafness. The location of the brain lesion can be summarized as follows: (1) Both MRI and PET study showed that damage was confined to the left hemisphere, with no functional or morphological abnormality of the right hemisphere. (2) In MRI study, the main lesion was found in the left thalamus and the white matter of the left temporal and parietal lobes. The lesion of the white matter ran rostrally along the lateral wall of the posterior part of the lateral ventricle and reached the centrum semiovale. No abnormality of the temporal cortex, including primary auditory areas, Wernicke's area and the underlying white matter, was found. (3) PET study confirmed a decrease in CMRO₂ and CBF of the left temporo-parietal area, but no abnormality of the posterior part of the superior temporal gyrus (including the primary auditory area and Wernicke's area).

From autopsy studies of pure word deafness, the responsible lesion has been said to occur at either of these sites: (1) left temporal cortex and subcortex, or (2) bilateral temporal cortex and subcortex, the latter being more frequently reported. Among the seven cases of unambiguous pure word deafness, only five have had the location of the lesion clearly identified (Henschen, 1917; Hamanaka et al., 1980; Kamei et al., 1981; Metz-Lutz and Dahl, 1984; Yaqub et al., 1988). The lesion was bilateral in the case of Henschen (1917) and in that of Yaqub et al. (1988), and unilateral on the left in the three remaining cases.

More in detail, the case of Hamanaka et al. (1980) had a left-sided hematoma extending from the basal ganglia to the above-lying white matter along the posterior portion of the lateral ventricle. In the case of Kamei et al. (1981), a left-sided infarct extended from the posterior superior temporal gyrus and adjacent portions of the supramarginal and angular gyri to the posterior lateral ventricle, making a deep wedge-shaped cut into the subcortical white matter. The locus of lesion was similar in the case of Metz-Lutz and Dahl (1984): a left-sided infarct extended from the left temporo-parietal cortex to deep subcortical areas. In other words, all unilateral cases, including our own, had a subcortical lesion of the left temporal lobe (particularly of the superior temporal gyrus) and of the white matter lying deeply in the temporo-parietal lobe near the posterior part of the lateral ventricle. These findings strongly suggest that this region is crucially associated with pure word deafness.

Since the early reports of Lichtheim (1885) and Liepmann and Storch (1902), the idea has been accepted that pure word deafness represents a disconnection syndrome in which Wernicke's area is isolated from auditory linguistic information. Theoretically, three lesional patterns could produce such a syndrome: (i) interruption of the fibers connecting the left auditory cortex with Wernicke's area, and of the right auditory radiations; (ii) interruption of the association fibers linking the right auditory cortex with Wernicke's area, and of the left auditory radiations; and (iii) interruption of the association fibers linking the right auditory cortex with Wernicke's area, and of the connections between the left auditory cortex and Wernicke's area (see Figure 6). Due to the rarity of pure word deafness cases, especially of those in which the lesion has been adequately studied, it has been impossible to distinguish among these possibilities. The anatomical course of the central auditory pathways has also remained problematical. Although the course of the radiations linking the medial geniculate body with the unilateral auditory cortex has been identified (Kretschmann and Weinrich, 1984), little is known about the location of the interhemispheric fibers crossing in the corpus callosum. Damasio and Damasio (1979) and Alexander and Warren (1988) have suggested that auditory commissural fibers run backward and upward from the primary auditory cortex, proceed along the wall of the cerebral ventricles lateral to their trigonal portion, and cross in the posterior part of the corpus callosum. As discussed above, lesions of the white matter lateral to the posterior part of the lateral ventricle are common to all of the four cases with unilateral left hemisphere lesions, strongly suggesting that the commissural fibers have been damaged in this region.

In our case, MRI showed damage to the left auditory radiations, while no notable abnormalities were seen in the left auditory area or Wernicke's area in either MRI or PET studies, which makes it unlikely that the lesion laid between the left auditory cortex and Wernicke's area. In the light of the above arguments, we submit that in our patient pure word deafness was due to type (ii) lesional pattern (see Figure 6), namely, to the combined interruption of the left auditory radiations and the commissural fibers from the right auditory area.

The reason why our patient was able to name auditory sounds [that is, why there was not an auditory modality-specific anomia, as described by Denes and Semenza, (1975)], is not entirely clear. Two possibilities come to mind. The first is that there was partial damage to the commissural fibers connecting the right auditory cortex and Wernicke's area, so that only the transfer of linguistic information was compromised. Alternatively and taking into account cases in which only deficits in the perception of environmental sounds have been reported (Spreeen et al., 1965; Fujii et al., 1990), the existence of an "environmental sound" center on the right or bilaterally can be hypothesized. On this account, damage may have affected the commissural fibers from the right auditory cortex to Wernicke's area, but not the commissural fibers from the "environmental sound" center to Wernicke's area. Unfortunately, clear anatomical or physiological support for either of these hypotheses is lacking, and their distinction remains a task for the future.

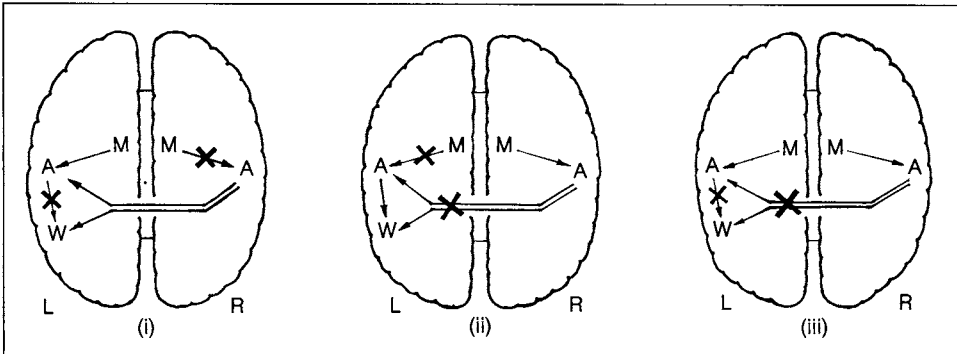


Fig. 6 - Three types of lesions which could result in pure word deafness. A type (ii) lesion is thought to be responsible for the pure word deafness in the present case. (A = auditory area; M = medial geniculate body; W = Wernicke's area; x = lesion).

ABSTRACT

We report the case of a 55 year-old right-handed man who presented with a long lasting pure word deafness following left thalamic bleeding. There was no sign of aphasia. The auditory deficit was specific for language, while recognition of music and environmental sounds was normal. CT, MRI and PET examinations showed that the lesion was anatomically and functionally confined to the left cerebral hemisphere, mainly the white matter of the temporal and parietal lobes. Wernicke's area was largely preserved. It is proposed that pure word deafness was consequent to the isolation of Wernicke's area from incoming auditory information due to the interruption both of the association fibers from the right auditory area traveling across the corpus callosum and of the left auditory radiations.

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