PURE WORD DEAFNESS (ACQUIRED VERBAL AUDITORY AGNOSIA) IN AN ARABIC SPEAKING PATIENT

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SUMMARY

A 38-year-old, right-handed Arabic-speaking male developed pure word deafness three days after myocardial infarction. He could recognize Arabic music and instruments but not words of songs; a radio broadcast from the Koran, but not the individual words; a male as opposed to female voice; Arabic and non-Arabic languages; and whether sentences were questions, exclamations, or imperatives. He discerned whether the speaker was emotionally neutral, happy, angry or sad. Contextual cues and reducing the rate of speaking aided verbal comprehension. Pure tone threshold audiometry revealed mild bilateral sensorineural loss up to 2000 Hz and a moderate high frequency loss. Brainstem auditory evoked potentials were normal, cortical auditory evoked potentials abnormal. CT scan revealed bilateral infarcts subcortically just posterior to the left superior temporal gyrus and the right posterior superior and midtemporal regions. Neurolinguistic tests indicated that the deficit is prephonemic and not due to impairment of linguistic discrimination.

INTRODUCTION

Pure word deafness or acquired verbal auditory agnosia is characterized by an inability to comprehend and repeat speech, with preserved ability to identify nonverbal sounds. Reading, writing and speaking ability is also preserved. It was first described by Kussmaul (1877) and later by Lichtheim (1885). In its pure form, it is very rare. It usually evolves from or towards Wernicke's aphasia (Barrett, 1910; Ziegler, 1952; Klein and Harper, 1956; Gazzaniga *et al.*, 1973; Albert and Bear, 1974). It is described in patients with lesions deep in the left temporal lobe (Klein and Harper, 1956; Gazzaniga *et al.*, 1973; Albert and Bear, 1974). However, most recent case reports, especially those with CT scan (Auerbach *et al.*, 1982; Coslett *et al.*, 1984) or pathological findings (Brick *et al.*, 1985) show bilateral temporal lesions occurring independently in time. In this

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case study of pure word deafness in an Arabic-speaking patient we analyse the neurophysiological, neurolinguistic and neuroanatomical findings.

CASE REPORT

A 38-year-old Syrian male was admitted to the Coronary Care Unit with acute myocardial infarction confirmed by serial enzyme estimations and electrocardiography. On the second day of admission he developed multifocal premature ventricular beats treated successfully by lignocaine. After 24 h he had no arrhythmia and was haemodynamically stable; lignocaine was stopped. On the third day it was noticed that he was unable to respond appropriately to verbal commands, although he seemed to be behaving normally, speaking fluently and sensibly. He explained that he had a bad cold obstructing his hearing as the reason for not being able to understand the speech of others. With self-generated speech he seemed to make sense, but answers to questions were irrelevant, though fluent, grammatical and of appropriate vocal volume.

Examination revealed no neck bruits. All cranial nerves were intact, including normal optic fundi. No abnormality was detected in the motor system. All modalities of sensation were preserved, including cortical sensation. The tendon reflexes were symmetric and the plantar responses were flexor. His gait was normal.

Mental state and speech assessment was undertaken in Arabic, the only language the patient spoke. When he could not understand a question, it was written legibly in Arabic. He was right-handed, alert and orientated for time, place and person. Spoken speech was fluent, grammatically correct, with normal phrase length and no paraphasic errors. He listened to a tape with Arabic songs. He could recognize and appreciate music and musical instruments, but not the words of the song, although he guessed correctly that they were in the Arabic language. When he listened to a radio broadcasting a chapter from the Holy Koran, he recognized that it was from the Koran, but did not know the chapter, and could not understand the words. He was blindfolded and a series of oral tests showed the following correct scores: 4/10 for 'yes'-'no' questions; 4/10 for prepositions and 7/10 for the comprehension of proverbs. He was able to recognize all nonverbal sounds such as key jingling, telephone ringing, rustling paper, knocking on the table, hand clapping and animal sounds.

When he did not understand a specific verbal stimulus he could not tell whether it was a real word or a nonsense syllable complex. He was able to discriminate between a male and female voice and between Arabic and non-Arabic languages. He was able to tell when the same sentence was delivered as a question, imperative, or exclamation. He could also recognize when the speaker's voice was emotionally neutral, happy or angry. He could discriminate between laughter and crying.

In writing to dictation he scored 4/10 for numbers, 5/10 for single words and 0/10 for sentences; he made no mistakes in writing spontaneously, copying and answering written questions. Reading aloud and reading for comprehension were intact.

Praxis, right-left discrimination and finger identification were unimpaired. He made no mistakes in naming objects (tie, coat, shirt, glasses, newspaper), parts of the body (nose, eye, hair, finger) and geometrical shapes (triangle, rectangle, circle and diamond). He named colours correctly (red, green, gold, silver, yellow and blue). Simple addition was intact. He was able to draw a face and a house spontaneously, and he copied a three-sided cube correctly. He could draw a clock with the correct time. Immediate and remote memory was normal. He was able to remember 3 numbers and the names of 3 cities, 3 min after presentation to him on a card. He also could perform a three-part command. He could remember the names of his children, his home and work telephone numbers and the town in which he was born.

RESULTS

Neurophysiological findings

Pure tone threshold audiometry

This revealed a mild, bilateral sensorineural hearing loss at frequencies up to 2000 Hz and a moderate high frequency loss (*see* Table).

Frequency (Hz)	Threshhold (dB)		
	R	L	
250	35	35	
500	30	35	
1000	25	30	
2000	10	15	
4000	40	35	
8000	50	70	

TABLE. PURE TONE AUDIOMETRY

Brainstem auditory evoked potentials

Waves I to V were of normal latency and amplitude bilaterally indicating intact brainstem auditory pathway function.

Cortical auditory evoked potentials

These were used to assess acoustic processing above the brainstem, using a Nicolet CA 1000 with a click stimulus of 2 Hz, 60 dB above hearing threshold with contralateral masking. Filters were set at 2–100 Hz with a sweep time of 200 ms; 256 responses were averaged on each run. The potentials were recorded from monopolar montages corresponding to T3–A1, C3–A1, C4–A2 and T4–A2 of the international 10–20 system. The same procedure was repeated twice on two separate days over a weekly interval. The potentials were compared with those from two healthy controls. A cortical potential response at 20 ms was obtained from the left temporal region (T3–A1) but not from the right (T4–A2) on stimulating either ear. The potentials in the left temporal region (T3–A1) and left central region (C3–A1) were higher in amplitude when the right ear was stimulated. These findings suggest the destruction of the right primary auditory cortex and probably right ear dominance.

Electroencephalography

The EEG showed long runs of moderate amplitude of theta-delta activity in the right temporal region suggesting a structural lesion.

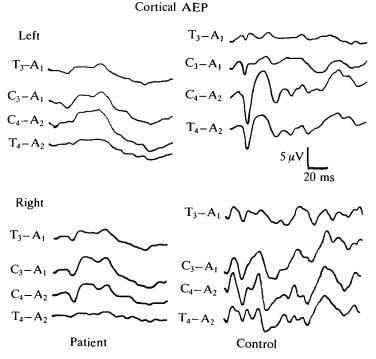


FIG. 1. Cortical auditory evoked potentials.

Click fusion and click counting

These were used to assess the ability to discriminate short duration stimuli.

Click fusion. A Medelec M592a stimulator was used to drive an earphone. The double stimuli mode was used to give an accurate delay between two equal clicks applied binaurally. A click fusion time was established by the method of ascending and descending limits. He fused clicks at intervals of 16 ms; below this he could not detect distinct clicks. This was repeated 5 times and he was consistent at this range. Five normal controls fused clicks at 2 to 3 ms.

Click counting. This was performed on Nicolet CA 1000 evoked potential apparatus. A varying number of clicks over a period of 1 s were delivered binaurally and the patient was asked to count the clicks. He was inaccurate for rates greater than 2 Hz, in the 5 times the procedure was repeated. Five normal controls were able to count accurately at 8 to 10 clicks/s.

Comment

These neurophysiological studies showed little evidence of peripheral hearing loss but an intact auditory pathway to the upper brainstem. Cortical auditory evoked potentials indicated destruction of right primary auditory cortex, but the severe deficit in temporal auditory acuity suggests that there should be another lesion on the left. Other physiological studies showed changes in an ECG series consistent with acute anterior myocardial infarction. Echocardiography, performed twice, immediately after onset of comprehension difficulty and one week later, did not reveal left ventricular thrombus. There was mild hypokinesis of the left lateral wall of the left ventricle with good left ventricular function.

Neurolinguistic findings

A number of linguistic tests in Arabic were directed towards a possible disorder in the discrimination of phonemes; all tests were performed with a native Arabic voice recorded on tape. Letters were shown on cards and he was asked to construct the heard syllables.

Vowel identification			
ي	و	i	
ΆA	Oo	Ee″	

Score=97%

FIG. 2. Three Arabic vowels used and the way they are pronounced.

Vowel identification

Arabic vowels were used; the way they are pronounced is shown in fig. 2. A total of 100 vowel stimuli were presented. The patient's score was 97%.

Phonemic discrimination/identification

All items were in a consonant vowel combination. In this case the vowel was (AA). The initial stop consonant differed according to place of articulation (labial, alveolar and velar) or voicing (voiced/voiceless).

Binary decision with a single distinctive feature. This assessed voicing or place of articulation; the nasal component was not checked. Nine tests of 100 items each were administered. The items used and the appropriate way they are pronounced in English, with the score, are shown in fig. 3. The average score was 80%.

Binary decision with two distinctive features. The phonemes differed by two features instead of one. Three tests of 100 items each were given in the same way as above (fig. 4). The average score was 92%.

Three-way decision for place of articulation. In this test a triad of stimuli was chosen which shared similar voicing but differed by place of articulation. Two tests of 100 items each are shown in fig. 5. The average score was 61%.

Ten-way decision. The patient was given a choice of 10 consonants and asked to identify each of 100 items, with equal numbers of voicing and place of articulation as one of them. They were used in consonant-vowel or vowel-consonant combinations. The vowel was (AA). There were 10 errors in voicing and 40 errors in place of articulation. (fig. 6).

Binary decision		
Single distinction		
(A) Voice/voiceles	S	
ما با	دا تا	غا خا
M_A, P_{PA}	D_A/T_{TA}	Ga/KH _{kha}
82%	84%	88%
(B) Place or articul voiced stops	ation	
دا غا	دا ما	ما غا
Da, Ga	Da, Ma	Ma Ga
78%	78%	74%
(C) Place of articu voiceless stops		
خا تا	با تا	خا با
	P_{PA}/T_{TA}	KH _{kha} P _{pa}

FIG. 3. Phonemic discrimination. Arabic consonant-vowel combination'. Binary decision-single distinction.

76%

80%

Binary decision Two distinctive features

82%

دا خا	غا با	ما تا
DA/KH _{KHA}	GA/P _{PA}	MA/T_{TA}
95%	88%	93%

Av = 92%

FIG. 4. Binary decision with two distinctive features.

Three-way decision Place of articulation

غا	دا	L	خا	تا تا	با
Ma,	Da	Ga	P _{PA}	T_{TA}	КН _{КНА}
56%			66%		

$A_{v} = 61\%$

FIG. 5. Three-way decision for place of articulation.

Ten-way decision

ب ۴ د ت ع ۲ <u>د</u> ن س ز Pp/ M/ D/ Tt/ G/ KH_{KH}/ K/ N/ S/ Z/

Score=10 errors voicing Score=40 errors place or articulation

FIG. 6. Ten-way decision used in consonant-vowel or vowel-consonant combination.

The same neurolinguistic test was administered to 3 Arabic-speaking controls; they made no mistakes (100%).

Comment

Our patient clearly showed the following. (1) No difficulty in vowel discrimination. (2) Discrimination is best when there is both a voicing contrast and a place of articulation contrast, as in the data in fig. 4. (3) Discrimination suffers most when voicing is held constant and place of articulation varies, as in the data in fig. 5. (4) It does appear that place of articulation is more important than voice although some other features such as nasality were not checked.

Neuroradiological studies

CT brain scans were performed twice, the first at 2 days and the second at 5 days after he developed pure word deafness. They showed bilateral low attentuation areas in both temporal regions, compatible with recent cerebral infarcts. The lesion on the left in the posterior superior temporal gyrus evolved over 3 days to a smaller lesion in the subcortical area just posterior to the primary auditory area. The

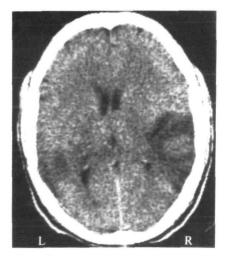


FIG. 7. CT brain scan showing bilateral temporal infarcts.

lesion on the right was more extensive and involved the posterior superior and midtemporal gyrus, in addition to the posterior parietal region. This evolved after three days to a smaller lesion involving only the cortex of posterior superior and midtemporal regions (fig. 7). The bilateral lesions were in the border zones between middle and posterior cerebral artery territories supplied by the corresponding posterior temporal artery (Damasio, 1983).

DISCUSSION

Our patient is unusual for three reasons. His inability to comprehend spoken speech was his only neurological abnormality; he exhibited 'pure' word deafness from the outset with no evolution from Wernicke's aphasia; and the pure word deafness was acquired because of bilateral, simultaneous and recent cerebral infarcts in the temporal lobes.

There was no evidence of generalized auditory agnosia, since he was able to recognize environmental (nonverbal) sounds. Our study does not support the contention in a recent review that auditory agnosia is found in all cases of pure word deafness (Buchman et al., 1986). There was no evidence of cortical deafness because he was able to recognize sounds easily, and the pure tone audiogram showed only mild sensorineural hearing loss. This finding is described in pure word deafness (Lhermitte et al., 1971). This syndrome was first ascribed to peripheral hearing loss by Freud (1891), but subsequent well-documented cases with autopsies showed a central origin (Goldstein, 1974). Our case was caused by bilateral lesions: cortical infarction destroying the right primary auditory area and geniculocortical fibres and a left subcortical white matter lesion of the posterior temporal lobe disconnecting Wernicke's area from the left primary auditory cortex, which was spared. These would support the disconnection hypotheses of Geschwind (1965) and Ulrich (1978). However, there are documented cases of pure word deafness associated with a single left temporal lobe lesion, presumably disconnecting Wernicke's area from the ipsilateral primary auditory cortex and interrupting transcallosal fibres from the contralateral auditory cortex (Gazzaniga et al., 1973; Schuster and Taterka, 1926).

To our knowledge this is the first Arabic-speaking person described with this syndrome. A modified neurolinguistic test in the Arabic language was applied, taking into consideration some differences between Arabic and English phonemes. Few previous studies (Chocholle *et al.*, 1975; Semenza, 1975; Saffran *et al.*, 1976; Blumstein *et al.*, 1977; Auerbach *et al.*, 1982) examined the role of phonemic discrimination of natural speech stimuli in pure word deafness and other types of aphasia. Auerbach *et al.* (1982) proposed that a subclassification of pure word deafness can be made: Type 1 (prephonemic) is characterized by (1) a disorder in temporal auditory acuity causing particular difficulty with rapid formant transitions leaving vowel discrimination relatively preserved; (2) increasing the number of target distinctions would improve performance; (3) place of articulation would be

the most vulnerable feature. Type 2, which is independent of temporal auditory acuity disorder, has a deficit in linguistic discrimination and is not prephonemic. The former is associated with bilateral temporal lobe lesions (M. Naeser, unpublished observations; Chocolle *et al.*, 1975), the latter with a left unilateral lesion (Chocolle *et al.*, 1975; Denes and Semenza, 1975). Our patient's neurolinguistic findings place him in type 1 and he had bilateral lesions.

Although pure word deafness has been attributed to tumour (Goldstein, 1948), infection (Goldstein *et al.*, 1975) and drugs (Donaldson *et al.*, 1981), most cases are due to stroke. In our patient the recent simultaneous infarction in both temporal regions could have been secondary to cardiogenic emboli to both posterior temporal arteries after myocardial infarction, although this seems unlikely. Therefore, because the simultaneous onset and the nearly homologous distribution would be unusual for embolism, and there were no arrhythmias on ECG monitoring and two normal echocardiograms, we believe that the most likely explanation for the bilateral simultaneous infarcts is hypotension during the myocardial infarction and subsequent hypoperfusion of these borderzone areas between the middle and posterior cerebral arteries.

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