

Clinical and Audiological Findings in a Case of Auditory Agnosia

G. Rosati, P. De Bastiani, E. Paolino, S. Prosser¹, E. Arslan¹, and M. Artioli²

Clinica Neurologica e Clinica Otorinolaringoiatrica¹ Università di Ferrara, Divisione Neurologica² Arcispedale S. Anna di Ferrara, Ferrara, Italy

Summary. A case is reported of severe agnosia for verbal and non-verbal sounds without associated aphasic disorder. A CT scan revealed bilateral, temporal lobe lesions from two ischaemic accidents that had occurred 9 months apart. The search for subtle deficits in this patient showed normal sensitivity to changes in the intensity and frequency of simple sounds; in contrast, his ability to discriminate sound duration and musical note sequences was severely impaired. The simultaneous recording of the whole auditory-evoked response pattern revealed no abnormality in the early components, which reflect the activation of the auditory nuclei and pathways of the brain stem. However, the middle and late components were delayed and slowed. These results and others in the literature suggest that the neocortex in man, as in other mammals, plays an essential role in the temporal aspects of hearing. Also, the two main ingredients commonly recognized in auditory agnosia, i.e. word deafness and the inability to interpret non-verbal sounds, are caused by the disruption of elementary, bilaterally represented cortical functions which start the processing of every kind of auditory information.

Key words: Auditory agnosia - Word deafness - Auditory-evoked potentials - Bitemporal lesions

Introduction

The classical identification of the neocortex with the capacity for auditory sensation profoundly influenced early views of brain function in man as well as in other animals. For the human auditory system, it was predicted that bilateral, temporal lobe lesions would result in total deafness [11, 25]. By 1940, numerous cases of cortical deafness resulting from bilateral damage to the temporal lobes had been reported, which seemed to confirm this contention [2, 4, 10, 11, 16, 20, 23, 24,

Offprint requests to: Giulio Rosati, M.D., Clinica Neurologica Universidt, Corso Giovecca 203, 44100 Ferrara, Italy

27, 34]. In the last three decades, however, views on the functional role of the auditory cortex in mammals have changed dramatically, and experimental evidence points towards an essential contribution of the cortex to temporal analysis and the storage of auditory information [9, 12, 30]. The same evidence seems to indicate that the physical parameters of simple sounds can be analyzed at subcortical levels. This has led many to question the existence of true cortical deafness and to suspect that most published cases of cortical deafness were inadequately tested, that the patients were suffering from auditory inattention rather than deafness [15, 17, 33]. In the most convincing case, reported by Le Gros Clark and Russell [16], the total hearing loss was probably due to concomitant involvement of auditory subcortical structures [17]. Similarly, the temporary deafness observed in other published cases [8, 35] could have been caused by a transient inhibition of the thalamic structures underlying the lesion [17]. This criticism of the classical concept of cortical deafness is supported by the latest literature, describing extensive testing of patients with more refined audiological methods. In four exceptionally well-documented cases published in recent years [13, 14, 17], subjects with successive lesions of both temporal lobes showed only slight impairment on pure-tone audiograms. On careful auditory testing, they were able to discriminate pure sounds of different intensity and frequency, but showed deficits in the discrimination of tone duration, auditory sequences and interaural order. These perceptive disorders, which seem to indicate that the human cortex is also involved in the temporal aspects of hearing, occurred together with auditory agnosia, i.e. the inability to interpret both verbal and non-verbal sounds, even though the patient can hear them [26].

Case Report

M.C. is a 49-year-old, right-handed farmer with known hypertensive disease. In January 1979 he suddenly had difficulty in comprehending words and was admitted to the neurological division of Ferrara Hospital. On admission, he showed fluent aphasia, right-sided hemiparesis, and leftsided focal EEG changes. A left carotid arteriogram was normal and a diagnosis of left cerebral infarct was made. The symptoms cleared up within 4 weeks and the patient was discharged after a month in hospital. Neurological examination 2 months after discharge revealed no residual language deficit. He had no more complaints until October 1979, when he suffered a new stroke with a sudden left-sided hemiparesis, which cleared within 2 h. A few hours later, when seen by his family doctor, the predominant finding was a complete inability to understand anything that was said to him. Despite this, he was able to give a clear account of what had happened to him and to follow stories in the newspaper. In the subsequent months, his inability to understand spoken words remained unchanged and his wife also noted that he had difficulty in distinguishing the nature of sounds. When he was sent to us in February 1980, he appeared alert and oriented. Neurological examination revealed no abnormality except slight spasticity and increased tendon reflexes in the right limbs, without clonus or Babinski's sign. On testing higher cortical functions, a complete inability to understand spoken words and a severe lack of recognition of non-verbal sounds were detected. On repeated language evaluations (Language Test devised by the Association for the Development of Neuropsychological Research, Milan, 1974), he was unable to understand spoken language, to repeat or write from dictation. Spontaneous speech, reading, reading comprehension, writing spontaneously and copying were all preserved. Naming by vision, touch and olfaction were almost entirely correct. On the Token test [7] given by written commands, he scored 29/36. There was no deficit in articulation or speed of speech, although he spoke loudly and did not generally say more than necessary. His ability to recognize familiar, non-

Fig. 1. CT scan showing bilateral temporal low-density lesions, more extensive on the left than on the right

verbal sounds was examined by means of a test similar to that proposed by Spreen and Benton [31]. The test consisted of 26 tape-recorded environmental sounds relayed binaurally. First, the patient was asked to name the sounds. Then he was asked to match each sound with a picture selected from an array of 3 pictures for the 13 sounds in set A and an array of 2 pictures for the 13 sounds in set B. The patient was able to name only 4 of the 26 sounds correctly. In general, he said that he did not know the sound and when invited to guess the name his answer was bizarre, without auditory or semantic similarity to the test sound. His ability to match the sound with the corresponding picture was no better than chance: two and four correct choices in sets A and B, respectively. On the other hand, he was able to discriminate simple sounds of varying loudness and pitch. The patient usually gave the right answer as to how many notes were touched on an instrument, whether the second note was higher or lower, louder or softer than the first. In contrast, he had great difficulty in judging whether the second note was longer or shorter than the first. Similarly, his ability to discriminate whether two successive sequences of seven notes were the same or different [6] was severely impaired. Because of his poor musical knowledge, the possibility of an amusia could not be adequately evaluated; however, the patient was unable to recognize melodies very familiar to his generation. Of interest was the fact that be bad no difficulty in locating the source of a sound in the room or at least in giving the direction of sound. The neuropsychological examination did not reveal any additional deficit. On the WAIS his nonverbal IQ was 90. When tested by WMS Form I, the patient scored normally on information, orientation, mental control and visual representation, all given by written commands.

ACT scan showed bilateral, temporal low-absorption defects, which were felt to be most consistent with old infarctions (Fig. 1).

On the audiological evaluation, pure-tone threshold was flat and revealed a hearing loss of 40 dB for both ears. No speech-audiogram could be obtained because of the patient's failure to comprehend words even at the maximal intensity of 120 dB. Impedance values and tympanogram were normal, while ipsilateral and contralateral stapedius reflexes were present. Auditory-evoked response analysis was performed by means of the method described by Prosser et al. [22, 29]. This method gives the simultaneous recording of the whole auditory response pattern (early, middle and late auditory-evoked potentials) and therefore permitted us to follow the travel of a neural excitation along the whole auditory pathway as a composite response to unitary stimulus. The stimulus consisted of 2,000 unfiltered clicks $(100 \mu s)$ square pulses) mono-aurally delivered by earphones, with an interstimulus interval of 750 ms. The intensity of click was at 90 dB. The 0 dB reference was the average subjective threshold of 10 normal hearing subjects. The electrode position was vertex-ipsilateral mastoid, with forehead grounded. The signal was amplified and

Fig. 2. Simultaneous recording of the whole auditory response pattern (early, middle and late potentials) evoked by click stimuli within 400 ms latency, according to the method described by Prosser et al. [22, 29]. The various components of the responses are labelled according to the criteria suggested by Picton et al. [28]. Trace a is the normative template obtained by averaging three normal subject responses. Traces b and e are the patient's right and left ear responses, respectively. When compared to the normal pattern (trace a), the patient's responses (traces b and c) show a normal pattern of the early components $(0-10 \text{ ms})$, which reflect the activation of the auditory nuclei and pathways of the brain-stem. On the contrary, abnormalities can be observed in both the middle (10-50 ms) and late (50-400 ms) potentials. The patient's auditory responses show the components P_a , N_b , P_1 , N_1 , P_2 shifted in latency, with a maximum delay of 50 ms for P_1 (trace c). A remarkable increase in interpeak latency values, causing a drift of the later components, can be noted in the region between P_0 and P_a for the right ear response (trace b) and in the region between P_a and P_1 for the left ear response (trace c). The late potentials are readily identified, but their configuration seems to reflect poor synchronization of neural discharge

filtered by a band-pass 0.5-3,000 Hz (6 dB/oct). Analogical-digital conversion, averaging and data processing were performed on-line on PDP 11/03. The signal was sampled over 4,096 points with a frequency of 10 kHz. In order to display all the components of the evoked response in a single trace, the time axis was compressed non-linearly, reducing the sample number from 4,096 to 512. Figure 2 shows the patient's evoked responses to click stimuli delivered in right (trace b) and left (trace c) ears compared to the response template (trace a) obtained by averaging three responses recorded from normal hearing subjects. The various components of the responses are labelled according to the criteria suggested by Picton et al. [28]. As can be seen, no abnormality was detected in the early components (0–10 ms), which reflect the activation of the auditory nuclei and pathways of the brain-stem. Abnormalities were, however, detected in both the middle (10-50 ms) and late (50-400 ms) potentials, which would reflect the activation of the auditory thalamus and cortex [28]. When compared to the normal pattern, the patient's right and left ear responses showed the components P_a , N_b, P_t, N₁, P₂ shifted in latency, with a maximum delay of 50 ms for P_1 . A remarkable increase in interpeak latency values, causing a drift of the later components, was noted in the region between P_0 and P_a for the right ear response and in the region between P_a and P_1 for the left ear response. Moreover, some components (P_a, P_2) of the left ear response appeared to be poorly defined because of their low voltage and consequent decrease of signal to noise ratio. The late potentials were readily identified, but their configuration seemed to reflect poor synchronization of neural discharge.

In the following months, the patient's visits to the neurological clinic were frequent and regular. It was consistently observed that no change occurred in his neuropsychological and audiological abnormalities. His most recent neuropsychological evaluation in May, 1981 showed no modification of auditory agnosia. Sometimes an effort to understand spoken words by lip-reading was noted but, as reported by his wife, this ability was very limited and this strategy not often used.

Discussion

In the case reported here, there was clinical evidence of a condition of auditory agnosia without associated aphasic disorders. The patient showed a severe lack of recognition of non-verbal sounds and a complete inability to understand spoken language, to repeat or write from dictation. Spontaneous speech, reading, reading comprehension, writing spontaneously and copying were all preserved. A CT scan revealed bilateral damage to the temporal lobes from two successive ischaemic accidents that had occurred 9 months apart. The first accident affected the left hemisphere and caused a receptive aphasia that cleared up within a month; the second, right-sided stroke caused the reported findings, which have remained essentially unchanged until the present time.

In this case, as in other published cases of auditory agnosia [1, 17, 19, 26], the first, left-sided stroke caused only a temporary impairment in the comprehension of spoken language, whereas the second led to complete loss of this ability. In Lhermitte's opinion [17], a clinical evolution of this sort can hardly be explained on the basis of a strict lateralization of language to the dominant left hemisphere. On the contrary, it suggests that cortical brain mechanisms involved in speech perception are represented more bilaterally than those underlying expressive speech. However, speech perception requires psychological processes distinct from all other forms of human perception [18] and physiological mechanisms not available to non-human mammals [21]. This apparent uniqueness of speech code is no doubt related to the phenomenon of cerebral dominance and strongly suggests that only the dominant left hemisphere is capable of identifying speech sounds [32]. So, in our own case, it is likely that the recovery of the aphasic disorder after the first stroke was due to a reorganization of function within the left hemisphere. Inability to understand spoken language caused by the second, right-sided stroke was not aphasic in nature, but a component of a more generalized disorder resulting from disruption of elementary, bilaterally represented mechanisms involved in the perception and analysis of critical acoustic cues. With regard to the fact that aphasia cleared up in spite of the extensive damage to the left hemisphere, it must be emphasized that areas concerned with language seem to be less precisely localized within the dominant left hemisphere than the motor cortex and the primary visual areas [3]. This, if true, would suggest that neural basis of language may vary from subject to subject and that individual variations exist in the capacity for reorganization and recovery of function after damage to the language areas [26]. This view would account for the fact that our own patient, like the case recently reported by Oppenheimer and Newcombe [26], could still talk, read and write, although the left hemisphere was dominant for speech, as suggested from the details of the first episode of aphasia with associated right-sided hemiparesis.

Our patient was able to discriminate simple sounds of varying loudness and pitch. In contrast, he was unable to discriminate simple sounds of varying duration and musical note sequences separated in time. Identical findings have been reported by Lhermitte et al. [17] in three cases of auditory agnosia, including two examined by post-mortem. Taken together, these findings support the idea that the neocortex in man, as in other mammals [30], plays an essential role in the temporal aspects of hearing. In contrast, the physical parameters of sound could be analyzed at subcortical levels. From this, it would seem reasonable to suppose that many of the temporal functions which the human auditory cortex shares with non-human mammalian cortex are represented bilaterally and that such functions are especially disrupted in auditory agnosia. In other words, if one agrees with the view that speech perception is also partly dependent on many of the same temporal analyses known to be performed by the auditory cortex in non-human mammals [30], the two main ingredients commonly recognized in auditory agnosia, i.e. word deafness and the inability to interpret non-verbal sounds, would be caused by disruption of the phylogenetically older, bilaterally represented cortical functions that start the processing of every kind of sound.

On the audiological evaluation, this patient had a nearly flat pure-tone hearing loss of 40 dB. Similar deficits have been found in most published cases of auditory agnosia $[1, 5, 14, 15, 17]$ and no doubt are not adequate to account for the impairment observed in these patients. In our own case, moreover, the simultaneous recording of the whole auditory-evoked response pattern did not reveal any abnormality in the early components and, therefore, seemed to exclude any involvement of the auditory nuclei and pathways of the brain-stem. In contrast, the middle and late components were delayed and slowed.

References

- 1. Adams AE, Rosenberger K, Winter H, Z611ner Ch (1977) A case of cortical deafness. Arch Psychiatr Nervenkr 224:213-20
- 2. Anton G (1899) Über die Selbstwahrnehmung der Herderkrankungen des Gehirns der Kranken bei Rindenblindheit und Rindentaubheit. Arch Psychiatr Nervenkr 2:86-127
- 3. Bogen JE, Bogen GM (1976) Wernicke's region: Where is it? Ann NY Acad Sci 280:834-843 4. Bramwell E (1927) A case of cortical deafness. Brain 50:579-580
- 5. Chocolle R, Loth D, Chedru F, Botte MC, Chain F (1975) Observations audiologiques sur
- un malade atteint d'agnosie auditive. Acta Oto-Rhino-Laryngologica Belgica 29:768-772
- 6. Denes G, Semenza C (1975) Auditory modality-specific anomia: evidence from a case of pure word deafness. Cortex 11 : 401-411
- 7. De Renzi E, Faglioni P (1975) L'esame dei disturbi afasici di comprensione orale mediante una versione abbreviata del test dei gettoni. Riv Pat Nerv Ment 96:252-269
- 8. Earnest MP, Monroe MA, Yarnell PR (1977) Cortical deafness: demonstration of the pathologic anatomy be CT scan. Neurology (Minneap) 27:1172-1175
- 9. Efron R (1963) The effect of handedness on the perception of simultaneity and temporal order. Brain 86:261-284
- 10. Heilbronner K (1897) Ober Asymbolie. Psychiatrische Abhandlungen. Breslau, Poland, 1 Fasc, Heit 3/4
- 11. Henschen SE (1918) Über die Hörsphäre. J Psychol Neurol 22:319-474
- 12. Hirsh IJ, Sherrick CE (1961) Perceived order in different sense modalities. J Exp Psychol 62 : 423-432
- 13. Jerger J, Weikers NJ, Sharbrough RW, Jerger S (1969) Bilateral lesions of the temporal lobe. Acta Otolaryngol (Stockh) Suppl 258
- 14. Jerger J, Lovering L, Wertz M (1973) Auditory disorder following bilateral temporal lobe insult: Report of a case. J Speech Hear Dis 37:523-553
- 15. Kanshepolsky J, Kelley JJ, Waggener JD (1973) A cortical auditory disorder. Neurology (Minneap) 23 : 699-705
- 16. Le Gros Clark WE, Russell WR (1938) Cortical deafness without aphasia. Brain 61:375-383
- 17. Lhermitte F, Chain F, Escourolle R, Ducarne B, Pillon B, Chedru F (1971) Etude des troubles perceptifs auditifs dans les lésions temporales bilatérales. A propos de trois observations dont deux anatomo-cliniques. Rev Neurol 124:329-351
- 18. Liberman AM, Cooper FS, Shankweiler DP, Studdert-Kennedy M (1967) Perception of the speech code. Psychol Rev 74:431-461
- 19. Mahodeau D, Lemoyne J, Foncin JF, Dubrisay J (1958) Considérations sur l'agnosie auditive. Rev Neurol 99:454-471
- 20. Marchand L (1904) Surdimutité par lésion symetrique des lobes temporaux. Bull Mem Soc Anat 6: 473-475
- 21. Mattingly IG, Liberman AM (1969) The speech code in the physiology of language. In: Leibovic KN (ed) Information processing and the nervous system. Springer, Berlin
- 22. Michelini S, Arslan E, Prosser S, Pedrielli F (1981) Logarithmic display of auditory evoked potentials. J Biomed Engn (in press)
- 23. Mills CK (1891) On the localization of the auditory centre. Brain 14:465-472
- 24. Mott FW (1907) Bilateral lesions of the auditory cortical centre: complete deafness and aphasia. Br Med J 2:310-315
- 25. Munk H (1881) Uber die Funktionen der GrofShirnrinde: Gesammelte Mitteilungen aus den Jahren 1877-1880. Hirschwald, Berlin
- 26. Oppenheimer DR, Newcombe F (1978) Clinical and anatomic findings in a case of auditory agnosia. Arch Neurol 35:712-719
- 27. Pick A (1896) Neue Beiträge zur Pathologie der Sprache. Arch Psychiatr Nervenkr 28:152
- 28. Picton TW, Hillyard SA, Krausz HI, Galambos R(1974) Human auditory evoked potentials. I: Evaluation of components. Electroencephalogr Clin Neurophysiol 36:179-190
- 29. Prosser S, Arslan E, Conti G, Michelini S (1981) Registrazione simultanea dei potenziali uditivi del tronco e della risposta corticale. Boll Ital Audiol Fon 4:50-57
- 30. Ravizza RJ, Belmore M (1978) Auditory forebrain: Evidence from anatomical and behavioral experiments involving human and animal subjects. In: Masterton RB (ed) Handbook of behavioral neurobiology, vol 1. Plenum Press, New York, pp 459-501
- 31. Spreen O, Benton AL (1963) Sound recognition test for clinical use. Neurosurgery Center, University of Iowa (unpublished memorandum)
- 32. Studdert-Kennedy M, Shankweiler D (1970) Hemispheric specialization for speech perception. J Acoust Soc Am 48:579-594
- 33. Vignolo LA (1969) Auditory agnosia: A review and report of recent evidence. In: Benton AL (ed) Contributions to clinical neuropsychology. Aldine Publishing Co, Chicago, pp 172-231
- 34. Wernicke C, Friedlander C (1883) Ein Fall von Taubheit in Folge von doppelseitiger Läsion des Schläfenlappens. Fortschr Med 1:177-185
- 35. Ziegler DK (1952) Word deafness and Wernicke's aphasia. Arch Neurol Psychiatry 67: 323-331