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Persistent Cortical Deafness: A Case Report.

Jessica Rigon[a,*], Marianna Cavinato[a], Irene Battel[b], Margherita Pivi[c], Francesca Meneghello[a], Carlo Semenza[d]

[a] *IRCCS S. Camillo, Lido di Venezia*
[b] *Macquarie University, Sidney*
[c] *Department of Linguistics, University of Venice*
[d] *Department of Neuroscience, University of Padova*

Introduction

Auditory disorders resulting from bilateral lesions of the temporal cortex and its radiations include cortical deafness, auditory agnosia, pure word deafness and phonagnosia. In cortical deafness any auditory signal cannot be perceived in the cortex in spite of normal peripheral hearing. Cortical deafness rarely persists for longer than a few weeks, generally evolving into other cortical auditory syndromes. We report behavioural and electrophysiological findings in a case of cortical deafness persisting for at least two years. Similarities and differences with previously reported cases of persistent cortical deafness, including its peculiar pattern of speech production, will be discussed.

Case report

CDB is a 55 year old, right-handed woman with normal hearing before two ischemic brain lesions. An MRI showed bilateral lesions to the superior temporal and transverse temporal (Heschl's) gyrus, temporal and polar planum, insula, angular gyrus, supramarginal gyrus, and frontal-parietal operculum. The right hemisphere was further damaged in the inferior frontal gyri and in the antero-lateral occipital areas. No damage to subcortical areas was found.

CDB did not show motor and sensory dysfunctions with the exception of complete deafness for sounds and a hardly intelligible speech production. Pure tone audiometry showed no response binaurally up to 110 dB hearing level (HL). The startling reflex to sudden sounds was absent.

CDB's comprehension of written material was excellent. Her written production was good but affected by a peculiar pattern of agrammatism, perhaps analogous to that affecting congenitally deaf people.

Electrophysiological data.

BAERs resulted in a normal range, indicating the integrity of the cochleocortical pathway to the inferior colliculi. MLAEPs appeared within normal limits in the right hemisphere, but were inconsistent on the left side. Finally, for LLAEPs, the N1, P2 and N2 were obtained with a substantial increase of latency.

Discussion.

CDB showed an unusually persistent case of cortical deafness. A comparison with similar cases reveals

* Corresponding author.
E-mail address: jessica.rigon@ospedalesancamillo.net .

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interesting similarities and differences.

1. Unlike in all other persistent cases reported in literature, no lesion was found in subcortical structures.
2. Unlike in all other cases, LLAPs were present, although with increased latency. MLAEPs were disturbed only on the left side.
3. Similarly to other patients, the patient did not show a startling reflex.
4. Unlike previously reported cases (whereby sometimes the phenomenon was specifically studied), CDB showed no sign of detection or localization of sounds she was not aware of ("deaf hearing").
5. CDB's agrammatism in writing, likely due to the extension of her lesion to the inferior frontal cortex, may be influenced by her hearing disorder.

Cortical deafness is still a poorly understood condition: factors leading to its persistence need to be identified by careful comparison among cases.