Pure word deafness

J.Sales-Llopis

Neurosurgery Department, University General Hospital of Alicante, Foundation for the Promotion of Health and Biomedical Research in the Valencian Region (FISABIO), Alicante, Spain

The selective inability to comprehend the spoken word, in the absence of aphasia or defective hearing, is defined as pure word deafness (auditory verbal agnosia). Reported cases of this rare disorder have suggested the site of involvement to be strategically placed, interrupting fibers from left and right primary auditory receptive areas which project to Wernicke's area in the dominant hemisphere.

This involved area is supplied by cortical middle and posterior temporal end-artery branches of the middle cerebral artery.

Case series

Among approximately 100 extracranial-intracranial bypass procedures performed, three patients developed the syndrome of pure word deafness. A cortical or posterior temporal branch of the middle cerebral artery was utilized as the recipient artery in each instance. In all cases, the deficit was transient, delayed (i.e. the patient did not awaken from anesthesia with the deficit), and unrelated to the preoperative deficit. The syndrome of pure word deafness may be more common after bypass than is currently recognized. Localized arterial spasm, dysautoregulation secondary to operative manipulation, or focal brain swelling may be operative mechanisms in the development of this and other delayed, reversible deficits related to bypass surgery [1)].

Case reports

2015

Joswig et al. report a clinical case of a 19-year-old male patient who developed pure word deafness due to the local compressive effect of a pineal germinoma on the inferior colliculus of the quadrigeminal plate. After percutaneous radiation therapy the size of the tumor decreased significantly, while audiometry demonstrated a complete regression of the auditory deficit. Since pure word deafness is commonly attributed to temporal lesions, the inferior colliculus represent an exceptional site for these symptoms. The pathophysiological background and the literature on pure word deafness, especially the one related to neoplasms of the tectal region is scarce [2)].

2014

After resection of a Vein of Galen aneurysm and arteriovenous malformation [3)].

2000

A 73-year-old female case with tentorial meningioma suffering from pure word deafness is reported. The patient initially presented with hydrocephalus, and was treated with a ventriculo-peritoneal(V-P) shunt. A year after the V-P shunt, she suffered from a symptom of deafness. On admission, her repetition and auditory comprehension were severely impaired, while reading and visual comprehension were almost normal. Auditory brain stem response (ABR) revealed normal latency between wave I and V, while wave VI and VII was disappeared. Middle latency response(MLR) showed no wave peak. On MRI, tentorial meningioma compressed bilateral medial geniculate bodies, but not auditory radiation or temporal lobe. 99mTc-HMPAO single photon emission computed tomography(SPECT) showed hypoperfusion in the left temporal lobe, considered as a diaschisis resulting from the isolation of left temporal lobe from auditory input via bilateral medial geniculate bodies [4)].

1993

A 56 years old right-handed female, a housewife, had suffered from right putaminal hemorrhage which had been treated surgically in July, 1985. Since it was difficult to approach the trans-sylvian fissure, evacuation of the hematoma was performed transcortically through the superior temporal gyrus. She was neurologically free and lived normally until she suddenly became completely deaf in April, 1991. On admission, she was alert, well oriented and spontaneously stated that she was deaf. She showed no response to verbal commands or loud noises but followed complex written commands. No other neurological deficit was observed. In addition to an irregular low-density area in the right temporal lobe, CT scan revealed a left putaminal hemorrhage. Audiological testing disclosed pure tone thresholds to be below 90db on both sides. Auditory brain stem response (ABR) showed waves I to VI in normal latency ranges, but wave VII was not observed. She was treated conservatively. Although her hearing loss had been gradually improving for 1 month, she suffered from word deafness, an inability to comprehend speech, for the following 2 months. Clinical features, CT findings as well as results of ABR suggest that hearing loss observed in this case, cortical deafness, was presumably due to bilateral damage of acoustic radiation of the temporal lobe. It was also suggested that, although she was asymptomatic, cortical and subcortical damage to the right temporal lobe following evacuation of the putaminal hematoma may have been extensive. During surgical procedures for evacuation of the putaminal hematoma, it has to be always kept in mind not to damage the auditory cortex and its radiations [5)].

1977

A 44-year-old male who suffered from an uncertain illness complicated by fever, jaundice and generalized seizures seven years previously. Following an apparent convulsion, the patient was noted to be unable to understand spoken language without loss of ability to recognize and respond to sounds or marked impairment of speech or reading. The evidence suggested bilateral cerebral hemisphere disease more marked on the right. The abrupt onset without progression is consistent with a vascular or ischemic etiology. Conclusions about the nature of the lesion and areas involved must await further studies and ultimately tissue examination [6)].

1)

Kanter SL, Day AL, Heilman KM, Gonzalez-Rothi LJ. Pure word deafness: a possible explanation of transient deteriorations after extracranial-intracranial bypass grafting. Neurosurgery. 1986

Feb;18(2):186-9. PubMed PMID:
2)
Joswig H, Schönenberger U, Brügge D, Richter H, Surbeck W. Reversible pure word deafness due to inferior colliculi compression by a pineal germinoma in a young adult. Clin Neurol Neurosurg. 2015 Sep 9;139:62-65. doi: 10.1016/j.clineuro.2015.08.034. [Epub ahead of print] PubMed PMID: 26372938.
3)
Bervini D, Oxenham V, Morgan MK. Pure word deafness after resection of a Vein of Galen aneurysm and arteriovenous malformation: report of a case. Clin Neurol Neurosurg. 2014 Aug;123:146-9. doi: 10.1016/j.clineuro.2014.06.004. Epub 2014 Jun 11. PubMed PMID: 24956543.
4)
Karibe H, Yonemori T, Matsuno F, Honmou O, Minamida Y, Uede T, Tanabe S, Hashi K. [A case of tentorial meningioma presented with pure word deafness]. No To Shinkei. 2000 Nov;52(11):997-1001. Japanese. PubMed PMID: 11215275.
5)
Nishioka H, Takeda Y, Koba T, Yano J, Ohiwa Y, Haraoka J, Ito H. [A case of cortical deafness with bilateral putaminal hemorrhage]. No Shinkei Geka. 1993 Mar;21(3):269-72. Japanese. PubMed PMID: 8487932.
6)
Shoumaker RD, Ajax ET, Schenkenberg T. Pure word deafness. (Auditory verbal agnosia). Dis Nerv Syst. 1977 Apr;38(4):293-9. PubMed PMID: 849705.