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Audiologic findings in unilateral deafness resulting from contralateral pontine infarct

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Focal brain stem infarction has been uncommonly associated with unilateral sensorineural hearing loss (SNHL). Kumar et al. (1986)[1] found a single case of brain stem infarction among 200 cases of unilateral SNHL. Previous case reports of unilateral SNHL after brain stem infarct have been the result of lesions of the ipsilateral cochlear nuclei in the dorsolateral medulla.[2,3] Wada and Starr (1983)[4] determined that an experimentally produced lesion of the lateral lemniscus produced a unilateral contralateral abnormality in the N_3 portion of wave III of the auditory brain stem response (ABR) in guinea pigs. We report a case of unilateral SNHL after a contralateral focal pontine infarct verified by magnetic resonance imaging (MRI) and ABR testing.

CASE REPORT

A 63-year-old man with a history of hypertension was admitted to the Veterans Administration Medical Center–Long Beach in June 1994 with a urinary tract infection. During his admitting physical examination, a neurologic examination was performed that revealed a right-sided hearing loss. The neurologic examination was otherwise normal. He reported that he had had a stroke in April 1994 and had spent several days in another hospital. The records and MRI scans from that hospitalization were obtained. The admission physical examination from April revealed a blood pressure of 170/86, a central right facial palsy, a right-sided hearing loss, nystagmus (direction not given), and unsteady gait. Laboratory examination revealed normal cerebrospinal fluid chemistries and cell counts and negative VDRL. The MRI from April 1994 demonstratd a dorsally located infarct involving the medial left portion of the upper pons, as well as ischemic changes of the periventricular deep white matter (Figs. 1 and 2).

He was referred 3 weeks after the stroke to the Audiology Department for evaluation of his right-sided hearing loss. Audiogram revealed a flat, severe SNHL in the right ear and a moderate high-frequency SNHL above 2000 Hz in the left ear (Fig. 3). Speech discrimination was 0% in the right ear and 92% in the left ear. Acoustic reflexes were absent in the right ear. ABR was performed with condensation and rarefaction clicks presented 10/second at 95 dB nSL (Fig. 4). In the left ear, all components were identified, and their intercomponent times were within normal limits. In the right ear, wave I was present for condensation clicks at 1.98 msec, and no other waves were identified. Transient and distortion-product otoacoustic emissions (OAEs) were performed. In the right ear, transient emissions were present with energy present through 3000 Hz with 81% reproducibility, similar to those obtained from the left ear (Fig. 5). Distortion-product OAEs (2F2 − F1 = 1.221; F1 = 70 dB SPL; F2 = 70 dB SPL) were present across frequencies in the right ear and through F2 = 3000 Hz in the left ear. Repeat MRI with gadolinium of the internal auditory canals was performed, and they were normal.

DISCUSSION

Although we used the combined term *sensorineural hearing loss,* the majority of patients with SNHL have lesions of the sensory apparatus, whereas "neural" lesions, including spiral ganglion degeneration, eighth nerve lesions, and central auditory pathway lesions from the cochlear nuclei to the temporal cortex, are found less frequently.[5] Among the causes of hearing loss, cerebrovascular infarction of the auditory pathways has been described infrequently. A 1986 report of 200 cases of unilateral SNHL found 42 individuals with confirmed retrocochlear lesions, of which there was one lesion adjacent to the cochlear nucleus and a second patient with a pontine lesion.[1] The most commonly found central lesions in that study were cerebellopontine angle tumors, central nervous system lues, and multiple sclerosis. Lownie and Parnes (1991)[2] found two cases of ipsilateral SNHL caused by infarcts of the cerebellum

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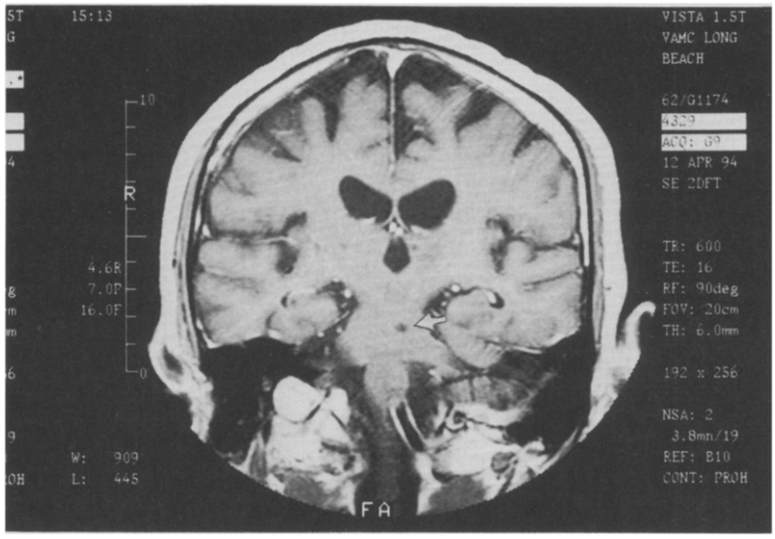

Fig. 1. Coronal T_1-weighted MRI of the brain demonstrating left pontine infarct *(arrow)*.

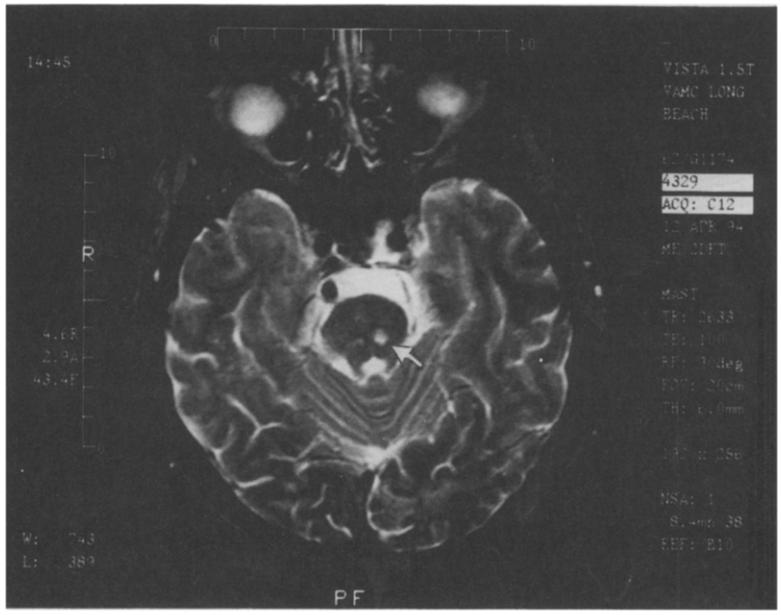

Fig. 2. Axial T_2-weighted MRI of the brain demonstrating left pontine infarct *(arrow)*.

and cochlear nucleus. In both patients there was significant improvement of the hearing loss in the weeks after infarction. Bales (1989)[3] reported a patient with a hearing loss ipsilateral to a pontine infarct who had a moderate-to-severe hearing loss, poor speech discrimination, and absent waves IV and V, with normal waves I through III. The hearing improved to normal in 1 month. Armington et al. (1988)[6] performed MRI in 176 patients with SNHL and found an abnormality believed to be responsible for the SNHL in 50 patients, 3 of whom had brain stem infarcts involving the auditory pathways (one in the cochlear nucleus; the locations of the other two were not given). There have been no previous reports of lesions of the contralateral upper pons producing hearing loss in human beings.

The brain stem auditory pathways are shown in Fig. 6. The cochlear nerve projects to the dorsal and ventral cochlear nuclei in the medulla adjacent to the inferior cerebellar peduncle. Second-order neu-

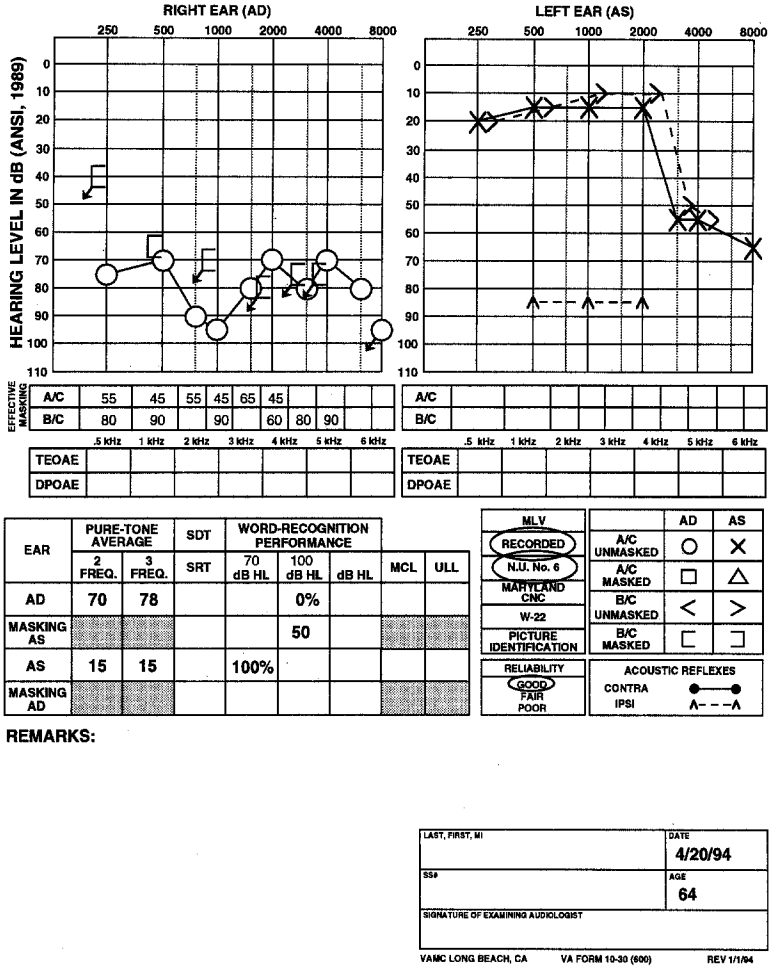

Fig. 3. Audiogram of patient demonstrating severe right SNHL and poor speech discrimination. Left ear has normal hearing to 2000 Hz and moderately severe high-frequency SNHL.

rons decussate to the contralateral medulla through the trapezoid body. Fibers then ascend in the lateral lemniscus in the pons to the inferior colliculus of the midbrain. Wada and Starr (1983)[4] produced lesions of the trapezoid body, lateral lemniscus, or inferior colliculus in guinea pigs to determine the effects on the ABR. Lesions of the trapezoid body produced attenuation of ipsilateral waves III, IV, and V. Lesions of the inferior colliculus had no effect on the ABR. When unilateral lesions of the lateral lemniscus were created in 11 animals, ABR to ipsilateral stimulation was normal, but a loss of the N_3 portion of wave III evoked by contralateral stimulation was found, and no changes were noted in waves II or P_3. Their previous work in cats had localized P_3 to the pons, with components of N_2, P_3, and N_3 generated in the brain stem contralateral to the ear stimulated. In contrast, Waves I and P_2 originate in the ipsilateral cochlear nerve and cochlear nuclei.[7] We posit that the patient in this article, who had bilateral high-frequency cochlear hearing loss, had a right-sided neural hearing loss from a brain stem infarction in the area of the contralateral pontine lateral lemniscus. The timing of the hearing loss at the onset of the stroke, and audiogram, ABR, OAE, and MRI findings all support this theory. The presumed mechanism of the right-sided hearing loss is interruption of the auditory pathway at the contralateral lateral lemniscus. Because the cochlea and cochlear nerve are normal on the right side, OAEs and ABR wave I are normal to ipsilateral stimulation in the right ear. It is unclear why waves II and III are missing to ipsilateral stimulation because they originate distal to the site of the lesion. Perhaps retrograde neuronal loss occurred.

The patient in this article had normal low-frequency transient (click) and distortion-product evoked OAEs. Evoked OAEs are sounds produced

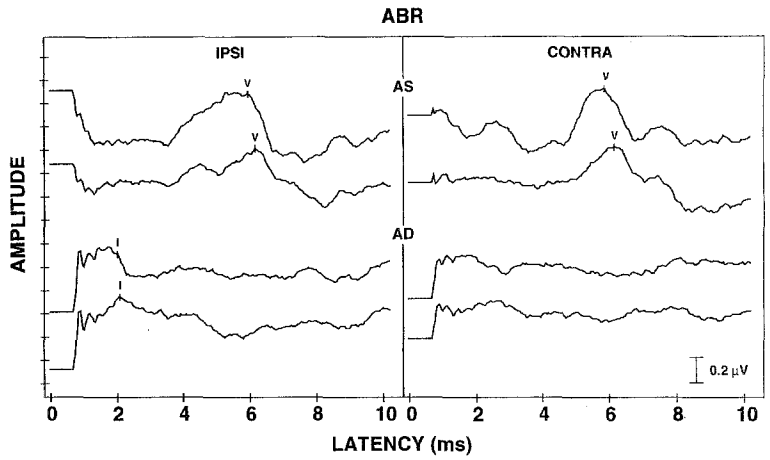

Fig. 4. ABR of patient demonstrating normal responses in the left ear and absent waves II through V in the right ear. The left ear responses have wave V at normal latency to both ipsilateral *(IPSI)* and contralateral *(CONTRA)* stimuli *(four upper tracings)*. In the right ear *(two lower left tracings)*, wave I was present for condensation clicks at normal latency, and no waves were identified for contralateral stimulation *(two lower right tracings)*. All tracings were obtained with condensation clicks.

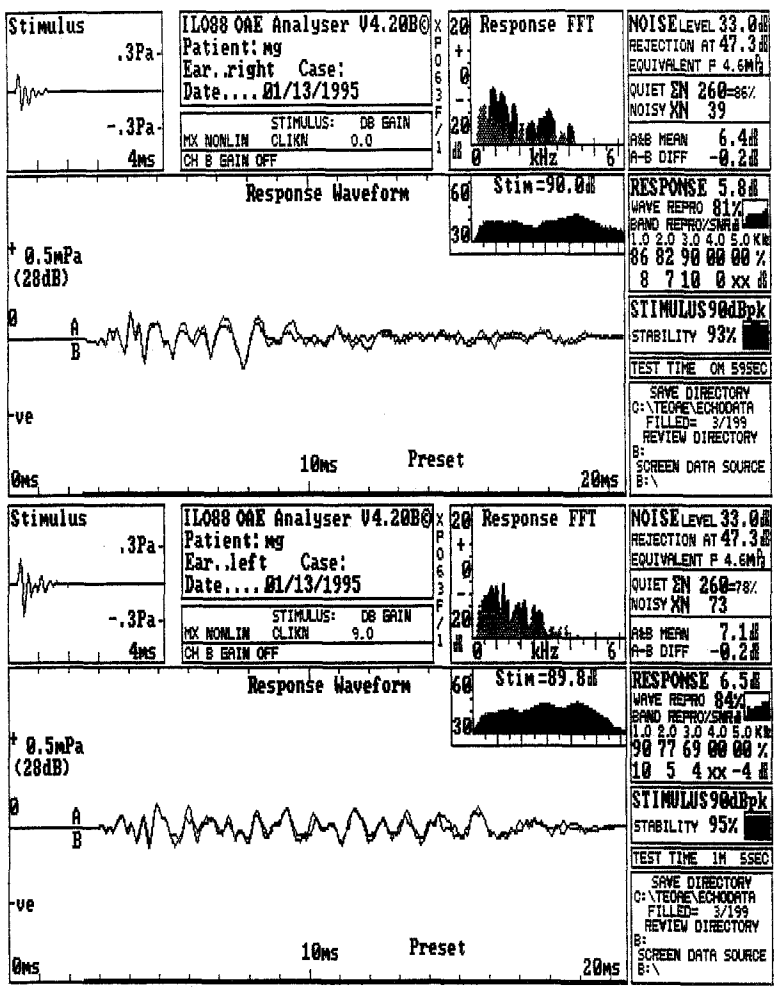

Fig. 5. Normal transient OAEs bilaterally through 3000 Hz.

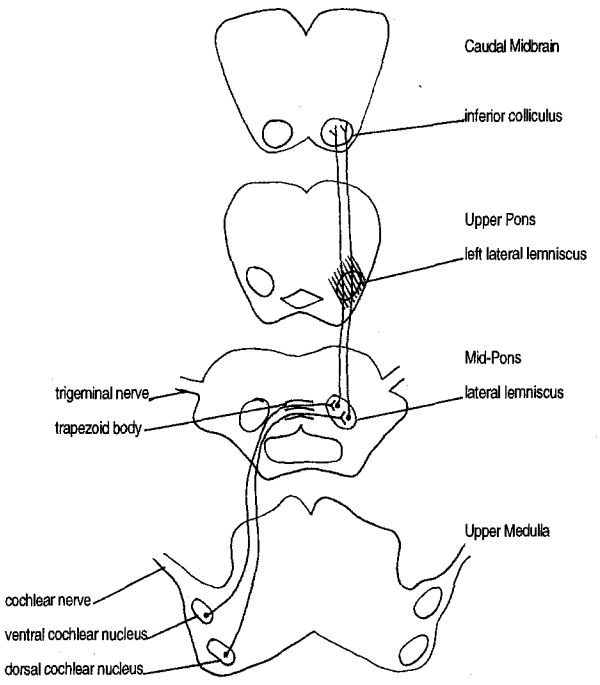

Fig. 6. Diagram of the brain stem auditory pathways with proposed location of infarct *(cross-hatched area).*

by the outer hair cells in response to acoustic transients presented to the ear.[8] Lonsbury-Martin et al. (1991)[9] and Sininger (1993)[10] emphasize the value of evoked OAEs in the differentiation of sensory from neural hearing loss. Several authors have used evoked OAEs to confirm neural hearing loss. Bonfils and Uziel (1988)[11] found click-evoked OAEs in several patients with acoustic tumors and hearing loss worse than 30 dB (cochlear hearing losses greater than 30 to 40 dB eliminate click-evoked OAEs), and Cane et al. (1994)[12] found transient evoked OAEs in 47% of 45 patients with cerebellopontine angle tumors. Robinette (1992)[13] reported the case of normal click-evoked OAEs present in a patient with profound hearing loss and multiple sclerosis who had a lesion of the auditory nerve as it entered the brain stem. Musiek et al. (1994)[14] recorded click-evoked OAEs in a "centrally deaf" patient. Starr et al. (1993)[15] reported the case of a child with moderate hearing loss by pure-tone audiogram who had no ABR to stimuli up to 90 dB nHL but who had a clear cochlear microphonic and robust OAEs. Berlin et al. (1994)[16] presented five cases of adults with bilateral SNHL, absent ABR in four of the patients (wave I only was present in the fifth patient), and normal click-evoked OAEs. Three of these patients had Charcot-Marie-Tooth disease, and the cause of hearing loss in the other two remained undiagnosed.

All patients were lacking the normally found suppression of click-evoked OAEs by continuous contralateral white noise that has been attributed to the efferent olivocochlear system. They believed that these patients had primary auditory nerve pathway disease with normal outer hair cell function. Our case represents the first report of a patient with unilateral severe hearing loss caused by brain stem infarction, with bilaterally symmetric distortion-product and transient evoked OAEs. This case demonstrates the value of OAEs in confirming the diagnosis of neural hearing loss.

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