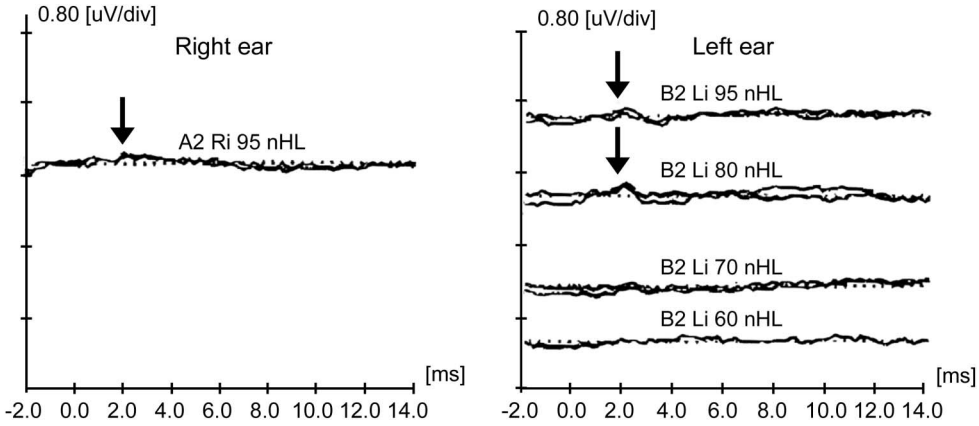
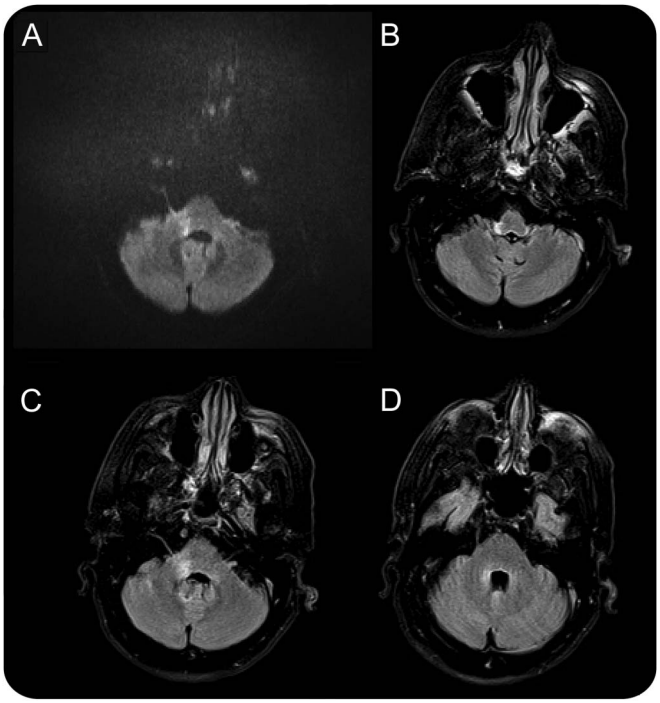
Bilateral hearing loss as a manifestation of neuromyelitis optica

Figure 1 Brainstem auditory evoked potentials at presentation



Brainstem auditory evoked potentials performed in both ears at the time of presentation show the bilateral presence of wave I (arrows) and the absence of all subsequent waveforms. These findings are consistent with lesions between the proximal cochlear nerves and the superior olivary nuclei. This is worse on the right, correlating with the MRI findings in figure 2.

Figure 2 MRI scans at presentation



Axial diffusion-weighted imaging (A) and fluid-attenuated inversion recovery (B–D) MRI at the time of presentation show restricted diffusion and T2 hyperintensity in the bilateral inferior cerebellar peduncles as well as in the right middle cerebellar peduncle. These changes are in the region of the cochlear nuclei, which are located near the fourth ventricle and inferior cerebellar peduncles. Dysfunction at this level of the auditory pathway is the likely explanation of this patient's hearing loss.

A 54-year-old woman with definite neuromyelitis optica (NMO) presented with bilateral hearing loss that progressed to complete deafness over 2 days. Otoscopy, tympanometry, otoacoustic emission testing, and brainstem auditory evoked potentials (figure 1) were consistent with central lesions bilaterally. MRI showed T2 hyperintensities and restricted diffusion near the cochlear nuclei, more prominent on the right (figure 2). Hearing loss resolved after administration of IV methylprednisolone followed by plasmapheresis. The cochlear nuclei are located in the dorsal medulla, adjacent to the fourth ventricle. Such periventricular regions highly express aquaporin-4 and have been implicated as sites of brain involvement in NMO.[1,2]

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