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Large Vestibular Aqueduct Syndrome with High CT Density and High MR Signal Intensity

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Summary: We report a case of large vestibular aqueduct syndrome with a markedly dilated endolymphatic sac bilaterally. The density and signal intensity of the extraosseous portion of the sac were higher than those of cerebrospinal fluid on CT and MR studies. The findings may represent protein-rich and hyperosmolar fluid within the endolymphatic sac.

Index term: Ear, abnormalities and anomalies

The vestibular aqueduct is a bony canal related to the bony labyrinth of the inner ear (1). The association of congenital sensorineural hearing loss with a large or enlarged vestibular aqueduct is well known as the large vestibular aqueduct syndrome (2–4). A large vestibular aqueduct has been demonstrated by conventional tomography and computed tomography (CT) (1–4). The endolymphatic duct and sac are the nonsensory part of the membranous labyrinth (1, 5) and are contained in the vestibular aqueduct (1). Although CT scans cannot show the membranous labyrinth itself, magnetic resonance (MR) images can show the anatomy and abnormalities of the fluid spaces related to the membranous labyrinth (1, 6–13). An enlarged endolymphatic duct and sac can also be seen on MR images of patients with large vestibular aqueduct syndrome (1, 8–12). We report a case of this syndrome in which the prominently dilated endolymphatic sac contained fluid that was hyperdense and hyperintense relative to cerebrospinal fluid (CSF) on CT and MR studies.

Case Report

A 20-year-old woman had a sudden exacerbation of bilateral hearing impairment, which had been recognized

since her early childhood. An audiogram showed bilateral profound sensorineural hearing loss with a down-sloping, high-frequency component. CT scans of the temporal bone with 1.5-mm contiguous sections showed the vestibular aqueduct to be enlarged bilaterally (Fig 1A). A diagnosis of large vestibular aqueduct syndrome was made. Other inner ear structures appeared to be normal bilaterally. The extraosseous endolymphatic sacs were dilated and were isodense with the adjacent cerebellum before and after intravenous injection of contrast material (Fig 1B).

The markedly dilated endolymphatic sacs were seen clearly on MR images obtained on a 1.5-T MR unit with contiguous 3-mm sections (Fig 1C–F). T1-weighted spin-echo images were obtained with parameters of 600/15/2 (repetition time/echo time/excitations); proton density- and T2-weighted images were obtained at 3000/15, 90/1. The intensity of the dilated extraosseous portion of the endolymphatic sacs was equal to that of the cerebellum on T1-weighted images and was hyperintense relative to CSF on both proton density- and T2-weighted images.

Discussion

The vestibular aqueduct that houses the endolymphatic duct and part of the endolymphatic sac is a bony canal related to the bony labyrinth of the inner ear (1). The endolymphatic duct and sac are the nonsensory part of the membranous labyrinth (1, 5). The duct continues into a blind-ended sac, which expands under the dura mater on the posterior surface of the petrous bone (14). In some patients with congenital sensorineural hearing loss, with or without other associated inner ear anomalies, the large or enlarged vestibular aqueduct is identified with conventional tomography or CT. This condition is known as large vestibular aqueduct syndrome (1–4). Although conventional

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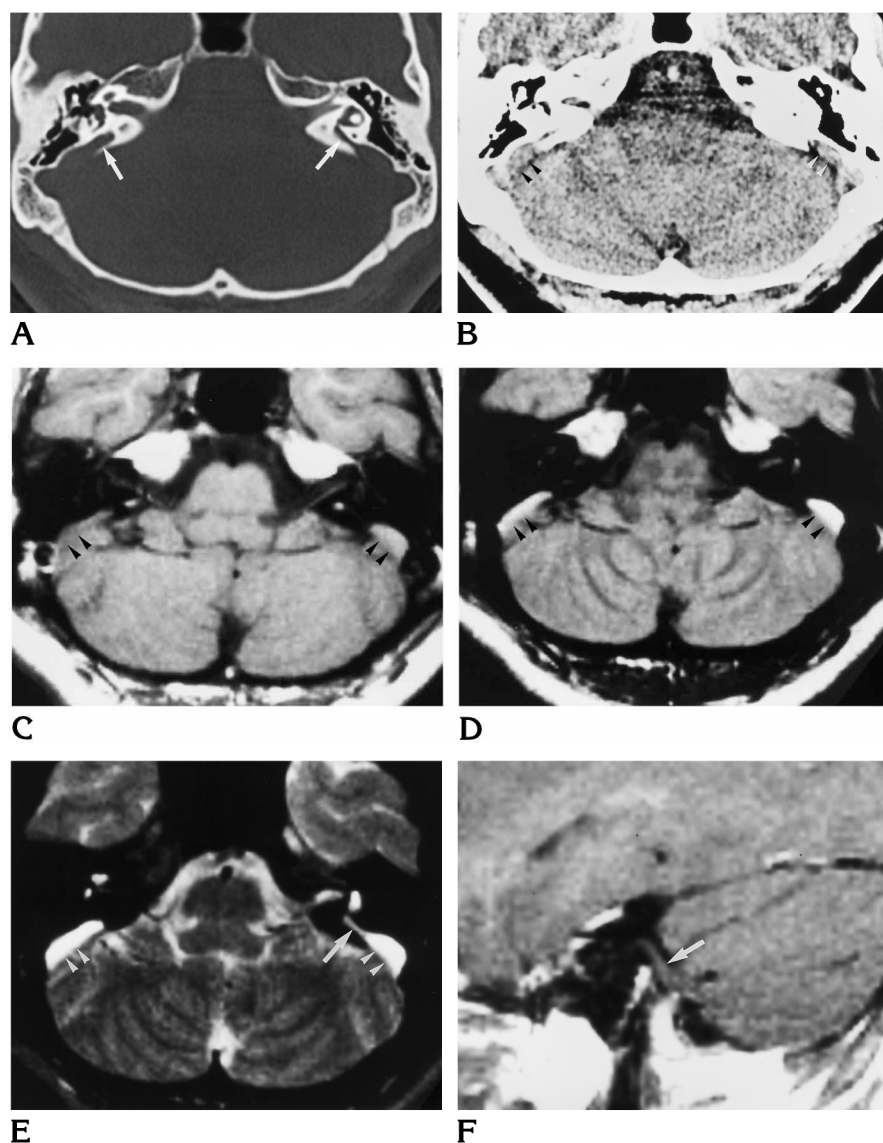


Fig 1. A 20-year-old woman with sudden exacerbation of bilateral hearing impairment.

A and B, CT scans show the large vestibular aqueduct bilaterally (arrows in A). The markedly dilated extraosseous portion of the endolymphatic sac (arrowheads in B) is isodense with the adjacent cerebellum on noncontrast and contrast-enhanced studies. The dura mater covering the dilated endolymphatic sac does not enhance so markedly that the dilated sac might be overlooked easily.

C–F, Spin-echo MR images show the prominently enlarged endolymphatic sac bilaterally. The intensity of the sac is isointense with the adjacent cerebellum on axial (C) and sagittal (F) T1-weighted images (600/15/2). The intensity is distinctly higher than that of CSF on proton density-weighted (D) and T2-weighted (E) images (3000/15, 90/1). (Arrow indicates the intraosseous portion of the endolymphatic sac; arrowheads, the extraosseous portion.)

tomography and CT cannot show the anatomic structures of the membranous labyrinth itself or the majority of their anomalies, MR imaging can give information regarding the labyrinthine fluid spaces (1, 6–13). However, precise evaluation of the signal intensity of the endolymph is usually impossible because of its small volume and the difficulty in distinguishing it from the perilymph bathing the external surface of the membranous labyrinth (1, 7, 14, 15).

Normally, the endolymphatic sac is filled with endolymph, which resembles intracellular fluid (1, 4). The endolymph within the sac has a surprisingly high protein content with a protein solute concentration of 1000 to 3000 mg/dL in humans (4). This protein solute concentration within the sac is markedly hyperosmolar in re-

lation to the osmolarity of the endolymph contained within the remainder of the membranous labyrinth (4).

The mechanism of hearing loss in large vestibular aqueduct syndrome has not been precisely elucidated. A possible theory is that the reflux of the contents of the hyperosmolar endolymphatic sac through a widely patent endolymphatic duct causes osmotic damage to the neuroepithelium (4). In our case, the dilated extraosseous portion of the endolymphatic sac was markedly hyperdense and hyperintense relative to CSF on CT and MR studies, respectively. These radiologic findings may support the theory that there is high protein or hyperosmotic liquid in the dilated sac.

In two cases previously reported (10, 12), the

density and the signal intensities of the fluid within the enlarged vestibular aqueduct were similar to those of CSF. The authors presumed an abnormal communication between the perilymphatic spaces of the labyrinth and the subarachnoid spaces through the lateral wall of the internal auditory canal. The contents of the enlarged aqueduct could represent an enlarged perilymphatic space or an enlarged endolymphatic space after fistulization into the membranous labyrinth (10). Even if the density and signal intensity of a dilated sac seem to be similar to those of CSF, high protein content in the sac is not necessarily ruled out, because CT and MR imaging may not be as sensitive to protein concentration. The enlarged extraosseous portion of the endolymphatic sac should not be mistaken for various other pathologic conditions, nor should it be overlooked when its density or signal intensity is similar to that of adjacent CSF or cerebellum.

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