Bilateral endolymphatic hydrops in a patient with migraine variant without vertigo

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Abstract

Objective: Endolymphatic hydrops (EH) has been well described in patients with Meniere’s disease. However, there is no study to date, to our knowledge, that examines for the presence of EH in a patient with migraine and bilateral hearing loss. Here we present the MRI findings using a sequence for detecting EH in a unique case of a patient experiencing migraine headaches and auditory symptoms without vertigo for more than 20 years.

Study design: Case report

Methods: Magnetic resonance imaging sequences included “cisternographic” three-dimensional T2, and delayed intravenous enhanced three-dimensional fluid-attenuation inversion recovery (DIVE-3D-FLAIR) sequences, performed with 2350 ms (bright perilymph) and 2050 ms (bright endolymph) inversion times. The bright endolymph images were subtracted from bright perilymph images to create a composite image with bright perilymph, dark endolymph, and intermediate bone signals.

Results: A 40-year-old female presented with a left-sided sensorineural hearing loss and severe migraine headaches that began at age 12. More recently, she experienced severe migraines with right-sided fluctuating sensorineural hearing loss, tinnitus, and aural fullness. Audiology confirmed increased right-sided hearing loss at times of symptom severity. Vestibular testing was within normal limits. MRI demonstrated the presence of severe bilateral vestibular and cochlear EH.

Conclusions: Endolymphatic hydrops of both the cochlea and vestibule can be present in patients without Meniere’s or vertigo. The relationship between migraine and Meniere’s disease may be more complex, with this patient with migraine associated bilateral hearing loss demonstrating bilateral EH. New imaging modalities allow for studies into the field of inner ear pathology, with significant implications for future research.

Introduction

Endolymphatic hydrops (EH) is an anatomic finding in which the structures bounding the endolymphatic space are distended by enlargement of the endolymphatic volume\textsuperscript{1}. This finding was first correlated with Meniere’s disease (MD) in the 1930\textsuperscript{2}-\textsuperscript{3}, leading researchers to conclude that EH is causative of Meniere’s. However, further research has shown that the connection between EH and MD is not a simple correlation. There is a subset of patients who have EH without the classical triad of symptoms that define Meniere’s disease: vertigo, hearing loss, and tinnitus\textsuperscript{4}. Additionally, MD has a significant overlap with migraine, and patients with vestibular migraine have also been found to have increased rates of EH\textsuperscript{5}. Furthermore, the comorbidity of migraine with Meniere’s disease is associated with concurrent bilateral aural symptoms and hearing loss, earlier age of onset, and a strong family history\textsuperscript{6}. The advent of new imaging techniques, allowing for the differentiation between perilymph and endolymph in the inner ear, has allowed for evaluation of EH in live patients\textsuperscript{7}. Here we present a patient with bilateral endolymphatic hydrops diagnosed by magnetic resonance imaging, who presents with auditory symptoms without vertigo, and the coexistence of severe migraine headaches.

Materials and Methods

Imaging was done on a 3-T scanner (Skyra, Siemens Healthcare, Erlangen, Germany) using a 12-channel head coil, paired with a two-piece 8-channel surface coil. 4 hours after administration of 0.2 mmol/kg gadodiamide intravenous contrast (Magnevist, Bayer HealthCare). Imaging consisted of three sequences: 1) “cisternographic” 3D turbo spin echo T2; 2) “Perilymph bright, endolymph dark” heavily T2-weighted (T2w-3D-FLAIR), obtained with an inversion time of 2350 ms; 3) “Endolymph bright, perilymph dark” T2w-3D-FLAIR, obtained with inversion time of 2050 ms. All sequences were acquired in the axial plane along the infraorbital bone, as three dimensional volumetric scans with 0.3 x 0.3 x 0.3 mm isotropic voxels. The endolymph bright T2w-3D-FLAIR images were subtracted from the perilymph bright T2w-3D-FLAIR images, in order to obtain an image with bright perilymph, dark endolymph, and intermediate signal bone. The cisternographic T2 was used to assist with anatomic reference.

Results

A 40-year-old female presents with severe migraine headaches and a stable left-sided hearing loss since age 12. Over the past year, she has developed right-sided fluctuating sensorineural hearing loss, tinnitus, and aural fullness and associated severe migraine headaches. The patient never experienced vertigo and a full battery of vestibular testing, including electronystagmography (with caloric), quantitative rotational testing, and vestibular evoked myogenic potentials were all within normal limits.

Discussion

- The presence of endolymphatic hydrops does not always correlate with Meniere’s disease. This has been previously demonstrated in studies of patients with acute low frequency sensorineural hearing loss, where a significant number of patients had EH without vertiginous symptoms\textsuperscript{8}. Temporal bone studies also confirm the presence of “asymptomatic hydrops.”\textsuperscript{9}
- Previous literature has shown a correlation between EH and vestibular migraine\textsuperscript{10}, as well as between MD and migraines\textsuperscript{11}. In this case we present a patient who has severe migraines with acute hearing loss at age 12, and presentation consistent with migraine variant without vertigo and MRI demonstrates significant bilateral endolymphatic hydrops.
- The case is consistent with findings relating the coexistence of migraine with Meniere’s disease is associated with higher incidence of bilateral hearing loss and younger age of onset.
- This suggests that endolymphatic hydrops is not the causative factor but a result of inner ear pathology.
- Migraine may be one causative factor of inner ear pathology that leads to the common final pathway of hydrops.
- New, noninvasive imaging modalities are allowing for exciting new opportunities to study EH in live patients, which will allow for further elucidations into Meniere’s disease.

References

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