

CASE REPORT

Vertigo revealing unilateral enlarged vestibular aqueduct in two adults

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INTRODUCTION

Vestibular aqueduct enlargement is the most common imaging abnormality in patients with congenital inner ear defects [1-2]. The mechanism is an early arrest in development of the endolymphatic canal and sac around the fifth to eighth weeks in utero, when the initial vesicle normally elongates and narrows to form the vestibular aqueduct [1].

Gene mutations have been found in some patients; transmission was autosomal recessive, and variability occurred in penetrance and disease expression [1]. Autosomal recessive hearing loss due to vestibular aqueduct enlargement occurs either as part of complex syndromes, most notably Pendred syndrome, or as DFNB4 nonsyndromic deafness. The hearing loss is sensorineural or mixed [3], usually starts in childhood, and more rarely is accompanied with attacks of vertigo.

We report two sporadic cases of childhood-onset hearing loss in females who started experiencing vertigo in adulthood. Imaging studies showed unilateral vestibular aqueduct enlargement in both patients.

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CASE REPORTS

Case n°1

This 55-year-old woman presented with vertigo. She reported a long-standing history of deafness in the right ear, with no cases of deafness in family members. She first experienced two attacks of vertigo lasting several hours and several days, respectively, at 53 and 54 years of age. After the second attack, she experienced multiple episodes of vertigo lasting a few seconds and often triggered by position changes, most notably lying down or turning around in bed, usually toward the left. Forced flexion or extension of the neck during the day caused no discomfort; neither did coughing, sneezing, or exposure to loud noises induce vestibular symptoms. She reported intermittent aural fullness on the right independently from the vertigo.

The otologic and neurological examination was normal. Positional maneuvers in the plane of the posterior or lateral canal showed no evidence of benign paroxysmal positional vertigo. Findings were normal from a thorough physical examination; in particular, no goiter was present. Pure-tone audiometry showed predominantly sensorineural hearing loss that was bilateral but predominated markedly on the right (Figure 1). Speech audiometry results were consistent with these findings. Early auditory evoked potentials were normal. Videonystagmography showed normal findings, with symmetric responses to caloric testing. Magnetic resonance imaging (IRM) disclosed vestibular aqueduct enlargement in the right ear, with no other inner ear abnormalities (Figure 2). Thyroid function tests and other laboratory findings were normal. Betahistine therapy was given. More than 1 year later, the patient reported no further vertigo attacks, although she continued to experience mild positional vertigo with minimal incapacitation.

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Figure 1: Pure-tone audiometry in Case 1: predominant sensorineural hearing loss most marked in the high-frequency range; the hearing loss is bilateral but is more severe on the right.

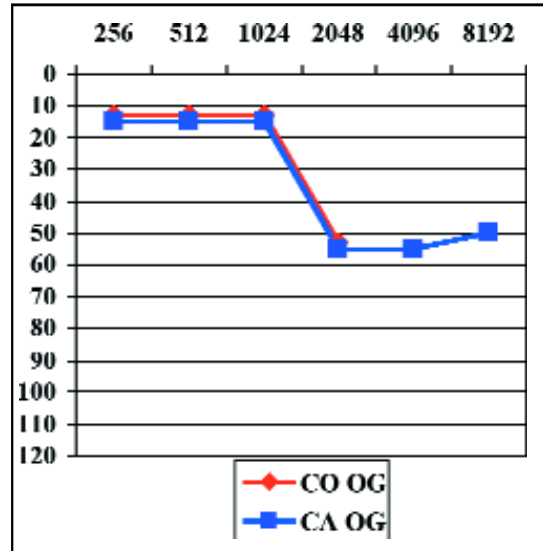
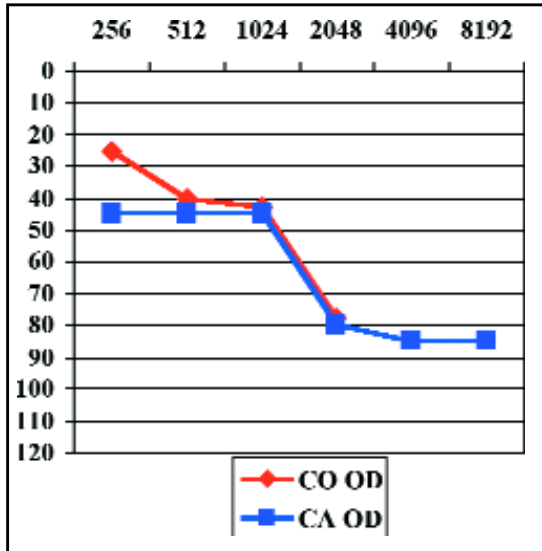
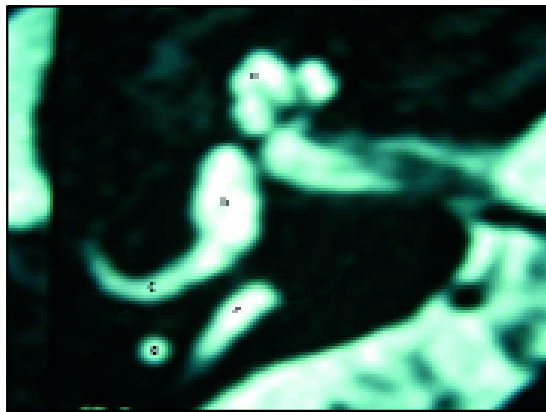


Figure 2: Magnetic resonance imaging in Case 1, axial section, T2-weighted sequence: dilation of the right endolymphatic canal (e). Visualization of the cochlea (a), utricle (b), horizontal semi-circular canal (c), and posterior semi-circular canal (d).



Case n°2

A 32-year-old woman presented with a 5-year history of frequent vertigo attacks. She reported deafness on the right since childhood, with no family history of deafness. Each vertigo attack lasted several hours and was accompanied with nausea and occasionally vomiting and diarrhea. She also described brief episodes of vertigo while coughing and sneezing, which she prevented by staring at an object or leaning against a

wall. Two attacks were so severe as to warrant emergency hospital admission.

During the first admission, the otologic and neurological examination showed a peripheral vestibular syndrome consistent with irritation: wearing Frenzel's goggles elicited right-beating horizontal and torsional nystagmus, and the patient deviated toward the left when she walked with the eyes closed. Findings were similar immediately after the second admission; on the next day, however, findings suggested a deficit, with a left-beating horizontal and torsional nystagmus while wearing Frenzel's goggles and deviation toward the right while walking with the eyes closed. No goiter was felt, and a thorough physical examination was normal.

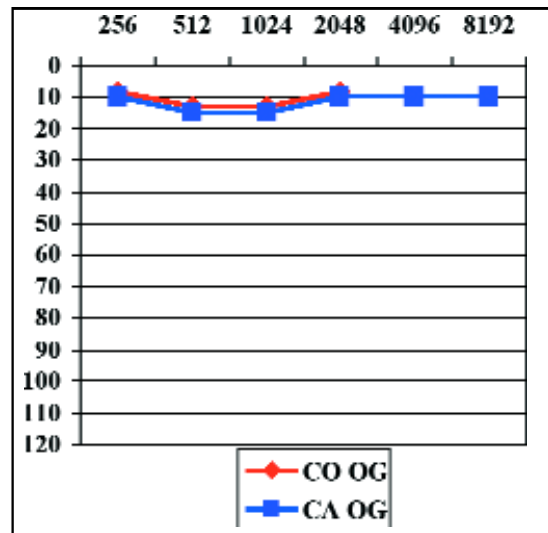
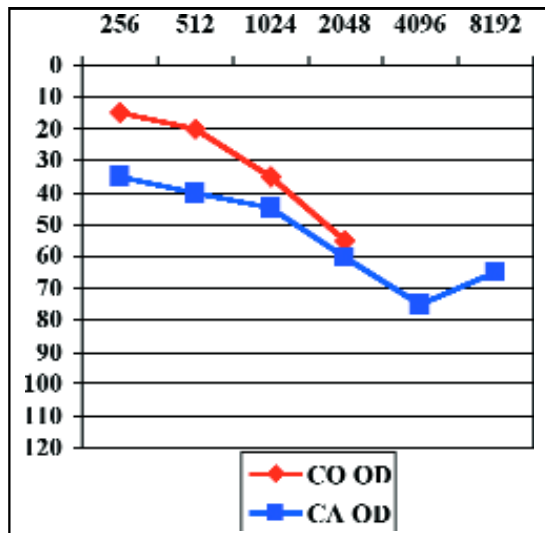
Pure-tone audiometry disclosed mixed hearing loss predominating in the high-frequency range (Figure 3). Speech audiometry findings were consistent with this pattern. Early evoked auditory potentials were normal. Findings were normal from videonystagmography, with symmetric caloric testing responses.

MRI disclosed unilateral dilation of the endolymphatic canal and sac on the right, with no other inner ear abnormalities (Figure 4). Thyroid function and other laboratory tests were normal.

Given the incapacitating vertigo with no response to pharmacotherapy, chemical labyrinthectomy was recommended. A tympanostomy tube was inserted under topical anesthesia and used to perform five

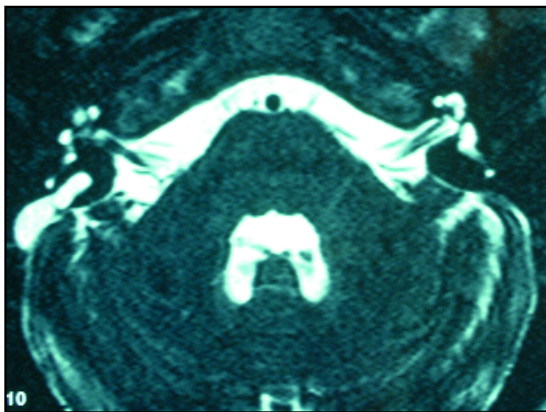
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Figure 3: Pure-tone audiometry in Case 2: *mixed hearing loss in the right ear predominating in the high-frequency range.*



injections of gentamicin, 20 mg/ml, over a 2-month period (0.5 ml; 0.4 ml; then 0.3 ml three times; for a total of 36 mg). At last follow-up 2 years later the patient was extremely satisfied; she reported no further vertigo attacks, despite brief minimally incapacitating discomfort upon rapid head movements.

Figure 4: Magnetic resonance imaging in Case 2, axial section, T2-weighted sequence: *dilation of the right endolymphatic sac and canal.*



DISCUSSION

Two points deserve discussion: the diagnosis, with vertigo revealing aqueduct enlargement in adulthood in both patients, and the treatment, with a good response to chemical labyrinthectomy in one patient.

Vestibular aqueduct enlargement usually manifests as sensorineural or mixed hearing loss [1, 3-5] starting in childhood. This symptom was present in both our patients. The hearing loss was bilateral in our case 1 despite the unilateral nature of the inner ear defect, in keeping with previous reports [2]. Radiologically visible vestibular aqueduct enlargement is simply a marker for more complex inner ear abnormalities [1]. The conductive component to the hearing loss, which was particularly marked in our case 2, may be related to sac enlargement and/or to sac communication with the subarachnoid spaces, which may act as a third window dissipating the sound waves [2-3]; this third window effect is the accepted mechanism for superior canal dehiscence syndrome. Although both our patients had long-standing hearing loss, the vestibular aqueduct enlargement was identified only in adulthood upon evaluation of recent-onset vertigo. The late onset of the vertigo (53 years in case 1) is surprising given the congenital nature of the inner ear defect and absence of previous head injury, although this time pattern has been reported previously [4]. The second patient experienced recurrent vertigo with a spinning sensation lasting several hours, and physical findings showed a peripheral vestibular syndrome consistent with irritation or a deficit, as seen in some patients with Menière's disease. Interestingly, our case 1 reported positional vertigo although her positional maneuvers showed no evidence of benign paroxysmal positional vertigo. Given the absence of arguments in favor of otolith migration, the most like-

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ly explanation to the positional vertigo is vestibular aqueduct enlargement. In our case 2, the vertigo occurred during coughing and sneezing, which can increase intracranial pressure; conceivably, this pressure increase may be transmitted to the inner ear via the enlarged vestibular aqueduct. None of the two patients reported Tullio phenomenon.

Vertigo related to vestibular aqueduct enlargement syndrome may be unresponsive to pharmacotherapy, as illustrated by our case 2. Surgical treatment of the enlargement is controversial, as endolymphatic sac decompression, arachnoid bypass, or endolymphatic sac occlusion may worsen the hearing loss, [1, 3-5]. Section of the vestibular nerve carries nonnegligible risks. These considerations led us to recommend chemical labyrinthectomy for our case 2. The outcome was favorable, although follow-up was only 2 years. We believe this technique can be recommended in patients with incapacitating vertigo due to unilateral vestibular aqueduct enlargement.

CONCLUSION

The combination of vertigo with sensorineural hearing loss predominating in the high-frequency range, or with mixed hearing loss, should prompt MRI. The scans should be examined for a cerebellopontine

tumor or an inner ear defect, such as vestibular aqueduct enlargement. In patients with incapacitating vertigo due to vestibular aqueduct enlargement, chemical labyrinthectomy can be performed, as with Menière's disease.

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