Narrow Internal Auditory Meatus

An Idiopathic Case Confirming the Origin and Pathway of Vestibular Evoked Myogenic Potentials in Humans

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Objective: To confirm the origin and pathway of vestibular evoked myogenic potentials (VEMPs) in humans.

Design: Case study.

Setting: University hospital.

Patient: A patient with a narrow internal auditory meatus (IAM).

Main Outcome Measures: Imaging studies and functional studies concerning the seventh and eighth cranial nerves.

Results: Of the 4 nerves in the IAM, all but the cochlear nerve had normal function and normal courses, despite the pronounced narrowing of the IAM. The facial nerve had a normal diameter, but the vestibular nerves were thinner. Imaging revealed that the cochlear nerve was absent or extremely thinned. Both the cochlea and the cochlear nerve showed no function in the affected ear, although the VEMPs were evoked normally.

Conclusion: Our results definitively support the vestibular origin of VEMPs in humans.


A CONGENITALLY narrow internal auditory meatus (IAM) is rare, especially as an isolated finding without inner, middle, or external ear anomalies.1-3 Cases with thorough neurological and physiological examinations have not been found in the literature. Acquired stenoses of the meatus, such as osteomas, exostoses, and fibrous dysplasias, were more frequently reported.6-8 Herein, we present the findings of imaging and functional studies in a rare case of a unilateral narrow IAM. Of the 4 nerve bundles in the IAM, only the cochlear nerve was atrophic and showed no measurable function, mimicking the conditions of cochlear deafferentation. Therefore, the results of functional studies in this case serve as direct evidence for the origin and the pathway of the vestibular evoked myogenic potentials (VEMPs) in humans.

IMAGING FINDINGS

High-resolution computed tomography revealed no abnormalities in the external auditory meatus or in the middle ear on either side (Figure 1). The IAM, cochlea, vestibule, and semicircular canals were normal on the left side. The inner ear structures were also normal on the right side. However, the right IAM was very nar-
row, and small, branched canals for the 4 nerves were identified at the periphery. The roof of bone overlying the superior canal was intact on both sides.

T2-weighted magnetic resonance images were more informative than T1-weighted images (Figure 2). On the left side, the inner ear structures were normal, and the 4 nerves in the IAM were clearly identified. On the right side, the cochlea, vestibule, and semicircular canals were normal. At the cerebellopontine angle, where the seventh and eighth cranial nerves leave the brainstem, the right eighth nerve was obviously smaller in diameter than the other side. On T2-weighted images, routes for the facial, superior vestibular, and inferior vestibular nerves were not identified. This was probably because of the little fluid space between the stenotic canal wall and the nerve. However, the route for the cochlear nerve was clearly identified to its entrance to the cochlea, which, paradoxically, suggests the atrophy of the nerve, because of the sufficient fluid space in the stenotic canal. On T1-weighted images, the right eighth cranial nerve was thin, and the 3 branches were not clearly identified, indicating the thinning of these branches or the absence of certain branches.

A summary of the imaging studies is shown in the Table.

NEURO-OTOLOGICAL FINDINGS

Auditory

Pure-tone audiometry revealed a profound sensorineural hearing impairment in the right ear and normal hearing in the left ear. Thresholds for sound sensation were unmeasurably high in the right ear. In the frequency range of 500 to 1000 Hz, at higher tone levels above 100- to 105-dB hearing level (HL), the patient had a feeling that her whole head was shaking as though she were experiencing an earthquake. Repeated distortion product otoacoustic emission and transiently evoked otoacoustic emission studies confirmed normal responses in the left ear, but very poor or no responses in the right ear, indicating severe cochlear impairment. On click evoked electrocochleography, action potentials were not recorded in the right ear, whereas they were normally evoked in the left ear. Auditory brainstem response tests also showed a normal response in the left ear but no response in the right ear. Promontory stimulation tests were performed on the right ear, using a needle electrode placed on the promontory mucosa through the ear drum. When the electric current was applied, the patient had a sensation of vibration but not sound, which indicated severe to total atrophy of the cochlear nerve.

Vestibular

Caloric responses were normal on both sides. Vestibular evoked myogenic potentials were evoked by click stimuli and recorded from the sternocleidomastoid muscles ipsilateral to the stimulation. Rarefaction clicks of 95-dB normal HL were used. The stimulation rate and the analysis time were 5 Hz and 50 milliseconds, respectively. Two hundred responses were averaged to obtain 1 recording. The VEMPs were normal both in wave latencies and in amplitudes (Figure 3). Although the P13-N23 amplitudes of the VEMPs were higher on the left, the difference (26%) was within the normal range (<34%).

A summary of the imaging studies is shown in the Table.
Other

Type A tympanograms were recorded on both sides. Stapedial reflexes were normally recorded in the right ear when the tones were delivered to the left ear, indicating normal facial nerve function on the right side. No responses were found in the left ear with stimulation of the right ear, owing to the profound deafness in the right ear. The results of physical examination of facial nerve functions were normal. Other cranial nerve tests also revealed no abnormalities. The patient had no nystagmus, dysequilibrium, or ataxia. Nystagmus was not evoked, even when an intense sound was delivered to the right ear.

A summary of the function of each nerve and sense organ is shown in the Table.

### Summary of Imaging and Functional Test Results

<table>
<thead>
<tr>
<th>Organ</th>
<th>Imaging Findings</th>
<th>Functional Test Results</th>
</tr>
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<tbody>
<tr>
<td></td>
<td>CT</td>
<td>MRI</td>
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<tr>
<td></td>
<td>Function</td>
<td>Method of Evaluation*</td>
</tr>
<tr>
<td>Inner ear</td>
<td>Cochlea</td>
<td>Normal</td>
</tr>
<tr>
<td></td>
<td>Semicircular canals</td>
<td>Normal</td>
</tr>
<tr>
<td></td>
<td>Otolithic organ</td>
<td>Normal</td>
</tr>
<tr>
<td>Cochlear nerve</td>
<td>Superior</td>
<td>Separate narrow canal</td>
</tr>
<tr>
<td></td>
<td>Inferior</td>
<td>Separate narrow canal</td>
</tr>
<tr>
<td>Vestibular nerves</td>
<td>Superior</td>
<td>Separate narrow canal</td>
</tr>
<tr>
<td></td>
<td>Inferior</td>
<td>Separate narrow canal</td>
</tr>
<tr>
<td>Facial nerve</td>
<td></td>
<td>Normal</td>
</tr>
</tbody>
</table>

*CT indicates computed tomography; MRI, magnetic resonance imaging; TEOAE, transiently evoked otoacoustic emission; DPOAE, distortion product otoacoustic emission; VEMP, vestibular evoked myogenic potentials; PST, promontory stimulation test; and SR, stapedial reflexes.

†Audiometry, electrocochleography and auditory brainstem response tests were also used to confirm hearing, although they are not capable of distinguishing between cochlear and auditory nerve functions.

### Comment

Vestibular primary afferent neurons of certain mammals respond to high-intensity sounds. Part of these neurons were also shown to respond to natural vestibular stimuli. The counterpart of these phenomena in humans is believed to be VEMP, which are the potentials recorded on the tonically contracting sternocleidomastoid muscle when loud monaural clicks are supplied to the ipsilateral ear. In humans, the presence of VEMP has been suggested that VEMP are of vestibular origin because of (1) the disappearance of the responses after vestibular deafferentation surgery despite the preserved hearing in 1 patient, (2) the preservation of the responses in patients with severe sensorineural hearing loss (>80 dB HL), and (3) the independence from the pure-tone hearing results in vestibular neuritis. However, there are objections to each theory: (1) because pure-tone thresholds remain normal until more than 80% of spiral ganglion cells, ie, cochlear nerve fibers, have disappeared, the integrity of the cochlear nerve is not guaranteed; (2) as the sound intensity delivered to the ear during VEMP recordings is very high, responses could have been improved because of the loudness recruitment phenomenon if the primary cause of deafness resides in the cochlea and not in the cochlear nerve; and (3) different patterns of the results can serve only as indirect evidence. To confirm the vestibular origin of VEMP, cochlear deafferentation, or section of the cochlear nerve, would be essential, which could not be done in human subjects. Substitution for cochlear deafferentation could be found in congenital anomalies in the IAM, in combination with the nerve atrophy.

With the exception of the cochlear nerve, the nerves in our patient’s IAM had normal function and normal
courses, although the superior and inferior vestibular nerves were thinner on the right side. The functional defects in hearing were not restricted to the nerve, but involved the sensory organ, i.e., the cochlea. Therefore, our patient simulated the state of selective and complete cochlear deafferentation in that no signals could be transmitted through the cochlear portion of the inner ear. This state is rarely found among human subjects, because it is very unlikely that an acquired disease process can impair only the cochlear nerve and because no treatment cuts the cochlear nerve selectively. If only the cochlear nerve were impaired, eg, by a viral infection, it would be impossible to diagnose this condition by imaging. The normal VEMPs that were recorded in our patient support the hypothesis that the origin of this response resides outside the cochlea and that the afferent pathway for this response runs in the vestibular nerve in humans. This confirmation reinforces the importance as well as the validity of using VEMPs to test vestibular function in humans.

Because of the smooth outlines of the narrow bony canals in our patient, the narrowing was considered to be congenital, formed by the excessive bone proliferation around the atrophic nerves in the course of ontogeny, because in cases of acquired stenosis, such as fibrous dysplasia or osteoma, the outlines of the stenotic canal are irregular, being constricted only at the portion where the lesion exists.6,7 The normally shaped fluid space in the cochlea revealed by the imaging studies indicates that the cochlea was normally formed at least once during ontogeny. If that was the case, the cochlear nerve also might have been formed once, since the developing cochlea, the otic vesicle, releases an analog of a nerve growth factor, which is essential for the differentiation of the afferent nerve fibers.21 The existence of the bony route corresponding to the cochlear branch, connecting to the cochlea, supports this hypothesis. Therefore, a question remains as to the onset of our patient’s hearing loss. The possibility is that the once-formed cochlea and the thinner cochlear nerve might have been damaged postnatally, for unknown reasons.

The results of pure-tone audiometry in our patient are of particular interest. A sensation of vibration, rather than sound, was invoked. One hypothesis is that this sensation may have been formed by the saccule, conducted via the vestibular nerve.14,22 A fact that may support this hypothesis is that the frequency range (500-1000 Hz) that invokes this sensation during pure-tone audiometry corresponds with the best frequency of the acoustically responsive vestibular afferent neurons in cats23 as well as with the optimum frequency of the Tullio phenomenon.23-25

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REFERENCES