

The Case of the Confounding Reflex Pattern

By Nina Brennan and Brad A. Stach

WELCOME BACK to an ongoing article series that challenges the audiologist to identify a diagnosis for a case study based on a listing and explanation of the nonaudiology and audiology test battery. It is important to recognize that a hearing loss or a vestibular issue may be a manifestation of a systemic illness. Being part of the diagnostic and treatment “team” is a crucial role of the audiologist. Securing the definitive diagnosis is not only rewarding for the audiologist, but enhances patient hearing and balance health care.

—Paul Pessis, Investigator-in-Chief

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Case History

Following a particularly brutal bout of spring allergies in 2011, a 61-year-old woman developed a sinus infection, which coincided with perceived hearing loss in her right ear. She had been feeling slightly off-balance and noticed a hissing sound—only in her right ear. She was referred to an otolaryngologist when her hearing had not returned nearly six months later.

Past Medical History

In April of 2011, the patient was diagnosed with breast cancer. She did not receive chemotherapy or radiation treatment. In August, she had a right mastectomy, followed

by reconstruction of the breast with an implant. Complications following the surgery, including a persistent infection that ultimately exposed the implant, resulted in its removal nearly one month later. The patient was also taking levothyroxine for hypothyroidism, Maxzide for high blood pressure, and simvastatin for hypertension.

Physical Examination

Otologic examination revealed normal external auditory canals, normal tympanic membranes, and normal mobility by pneumo-otoscopy, bilaterally. Nasal, oral, oropharyngeal, and neck examinations were normal.

Findings

Audiometric testing revealed an asymmetric sensorineural hearing loss. Pure-tone and speech audiometric results are shown in **FIGURE 1**. The patient had a mild-to-moderate high-frequency sensorineural hearing loss in the left ear, with a speech recognition threshold of 5 dB and word recognition score of 96 percent. For the right ear, the loss ranged in degree from minimal in the low frequencies to severe in the high frequencies. Thresholds were poorer in the right ear at all frequencies, with differences ranging from 10 to 35 dB. Of note, word recognition testing showed “rollover” of the intensity function, with scores of 96 percent at 70 dB HL, worsening to 72 percent at 90 dB.

Immittance results are shown in **FIGURE 2**. They are consistent with normal middle-ear function bilaterally, characterized by normal Type A tympanograms, normal static immittance, normal ear canal volumes, and normal ipsilateral (uncrossed) acoustic reflexes bilaterally. Interestingly, the contralateral (crossed) reflexes with sound presented to the right ear are absent across the frequency range. All other reflexes are present. These results are consistent with a disorder in the more central portion of the reflex arc, implicating crossing fibers in the auditory brain stem.

Initial Treatment and Follow-Up

In addition to referrals for further evaluation, the patient was prescribed a course of prednisone, a steroid often given in an attempt to treat sensorineural hearing loss that might be caused by inflammation of the auditory system.

Audiometric testing was again carried out three months later. Results showed no significant change in pure tone sensitivity or immittance results. In contrast, word recognition scores were dramatically reduced on the right, with a score of 28 percent at a presentation level of 80 dB HL.

Balance Function Testing

Videonystagmography test results indicated vestibular hypofunction on the right side. Caloric responses from the right ear were significantly weak. Rotary chair testing could not be completed due to patient claustrophobia.

Consider the Facts

The patient presents with

- Unilateral tinnitus on the right
- Asymmetric sensorineural hearing loss, right poorer than left
- Rollover of the word recognition intensity function on the right
- Absent contralateral, but not ipsilateral, acoustic reflexes, stimulus right
- Vestibular weakness on the right side

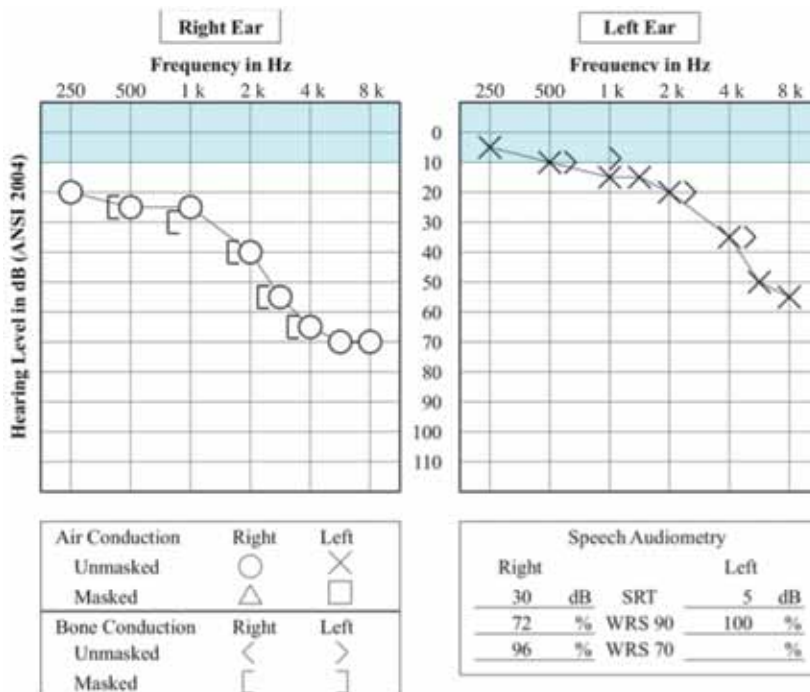


FIGURE 1. Pure-tone and speech audiometric results from a 61-year-old female with a history of cancer.

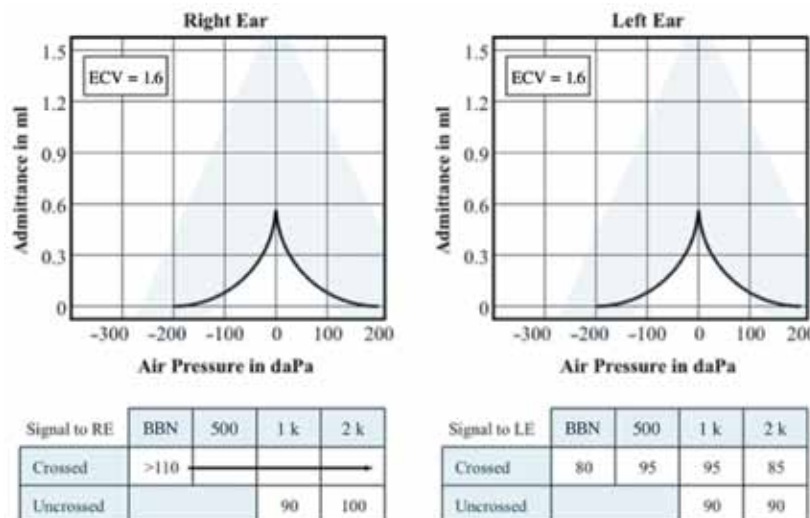


FIGURE 2. Tympanometry and acoustic reflex results.

Differential Diagnosis

You be the detective. Possible diagnoses could be

- Idiopathic sensorineural hearing loss, right ear
- Ménière's disease
- Metastasis of breast carcinoma

- Vestibular schwannoma
- Multiple sclerosis

Radiological Studies

The MRI report states:

- In the right internal auditory canal, there is a lesion measuring 13 × 9 mm primarily within

the internal auditory canal with a small component within the cerebellopontine angle that extends nearly to the fundus. There is smooth remodeling and expansion of the right internal auditory canal. There is diffuse enhancement of this lesion. There is no abnormal enhancement in the labyrinths.

- Nonspecific T2 FLAIR signal foci in the periventricular and deep white matter that likely relate to chronic ischemic white matter changes of mild burden.



FIGURE 3. MRI scan of a tumor of the right VIIIth nerve with extension into the cerebellopontine angle.

And the Diagnosis Is...

Vestibular schwannoma

Discussion

To a seasoned audiologist, this case likely brims with salient indications of VIIIth nerve disorder. The patient had unilateral symptoms, asymmetric hearing loss, asymmetric word recognition scores, rollover of the speech intensity function, unilateral vestibular weakness, and absent contralateral acoustic reflexes with sound presented to the suspect side. But what can we make of the ipsilateral reflexes with sound presented to the right ear? Let's walk through that story for a moment. The classic reflex pattern in a case like this is for the afferent portion of the reflex arc to be affected (Jerger and Jerger, 1977; Stach and Jerger, 1987). That is, a tumor on the right VIIIth nerve should theoretically interfere with the input side of the reflex arc and, eliminate both the

right ipsilateral and right contralateral reflexes. Presence of the left ipsilateral reflex suggests a normal efferent pathway on the left, eliminating that as a possible explanation for the absence of the right contralateral reflex. Similarly, presence of the right ipsilateral reflex suggests a normal afferent pathway on the right, eliminating that as a possible explanation for the absence of the right contralateral reflex.

Let's take a deep breath. This patient presented with normal right ipsilateral reflex thresholds and absent right contralateral reflexes. Theoretically, the pathology that eliminated or elevated the right contralateral reflex *should also* have eliminated or elevated the right ipsilateral reflex, if, in fact, the tumor was impacting the afferent portion of the reflex arc. By definition, vestibular schwannomas arise from the vestibular portion of the VIIIth nerve. Within the internal auditory canal, lesions of the vestibular nerve can compress the cochlear division, causing reduced nerve function and corresponding impairment of hearing and reflex responses. The facial nerve, primarily a motor nerve, is less commonly affected.

There is another possibility, of course, and that is that the tumor was having more of a secondary effect on the cochlea (Mahmud et al, 2003), explaining the hearing sensitivity loss and word recognition reduction, and less of a primary effect on VIIIth nerve function, explaining the preservation of the ipsilateral reflex. In other words, the portion of the tumor on the VIIIth nerve might not have been affecting the afferent portion of the reflex arc, while the extension of the tumor into the brain stem might only have been affecting the crossing pathway of the contralateral reflex.

Treatment

The patient's relative youth, good health, and residual hearing made her a viable candidate for surgical intervention. Risks, such as facial nerve weakness/paralysis, recurrence of tumor, infection, and decreased hearing sensitivity were explained to the patient. Coincidentally, the patient's son-in-law developed facial nerve weakness as a result of a vestibular schwannoma removal, and as a result, the patient was familiar with the potential adverse effects of the surgery. There are generally two favored approaches to surgical removal of a vestibular schwannoma. A translabyrinthine approach is considered the more accessible route to the tumor, but it destroys hearing in the affected ear. A suboccipital retrosigmoid approach is more complex but can often spare hearing in the affected ear. In an effort to preserve the hearing of the patient, the surgical team in this case opted for the suboccipital retrosigmoid surgery.

Results

In March 2012, the surgical team removed over 90 percent of the schwannoma. The precarious location of the tumor limited their ability to fully remove the growth. According to the operative note, the growth "could be seen to be coming from the superior vestibular nerve, but it extended significantly beneath (medial to) the facial nerve and splayed the nerve notably." The surgeon also noted "some extension into the CP angle."

Following surgery, the patient presented with facial weakness, which prevented her eye from closing completely, as well as a perceived further decrease in hearing on the right side. Postoperative MRI results indicate a residual mass in the cerebellopontine

angle and slight trauma consistent with neurosurgery. The patient is to return for another audiologic assessment upon surgical recovery.

Conclusion

In a perfect world of medical diagnosis, signs and symptoms of retrocochlear disorder align with textbook tenets. In this case, the audiometric outcomes were not classic, serving to remind us of the subtleties in the relation between structure and function and the uniqueness of every clinical encounter.

In short, it is rewarding to be a diagnostic detective. "Case" closed until the next issue of AT. 🎧

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