

> THE PATIENT

48-year-old man

> SIGNS & SYMPTOMS

- Acute hearing loss, tinnitus, and fullness in the left ear
- Weber test lateralized to the right ear
- Positive Rinne test and normal tympanometry

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> THE CASE

A healthy 48-year-old man presented to our otolaryngology clinic with a 2-hour history of hearing loss, tinnitus, and fullness in the left ear. He denied any vertigo, nausea, vomiting, otalgia, or otorrhea. He had noticed signs of a possible upper respiratory infection, including a sore throat and headache, the day before his symptoms started. His medical history was unremarkable. He denied any history of otologic surgery, trauma, or vision problems, and he was not taking any medications.

The patient was afebrile on physical examination with a heart rate of 48 beats/min and blood pressure of 117/68 mm Hg. A Weber test performed using a 512-Hz tuning fork lateralized to the right ear. A Rinne test showed air conduction was louder than bone conduction in the affected left ear—a normal finding. Tympanometry and otoscopic examination showed the bilateral tympanic membranes were normal.

THE DIAGNOSIS

Pure tone audiometry showed severe sensorineural hearing loss in the left ear and a poor speech discrimination score. The Weber test confirmed the hearing loss was sensorineural and not conductive, ruling out a middle ear effusion. Additionally, the normal tympanogram made conductive hearing loss from a middle ear effusion or tympanic membrane perforation unlikely. The positive Rinne test was consistent with a diagnosis of idiopathic sudden sensorineural hearing loss (SSNHL).

DISCUSSION

SSNHL is defined by hearing loss of more than 30 dB in at least 3 consecutive frequencies with acute onset of less than 72 hours.^{1,2} The most common symptoms include acute hearing loss, tinnitus, and fullness in the affected ear.¹ The majority of cases of SSNHL are unilateral. The typical age of onset is in the fourth and fifth decades, occurring with equal distribution in both sexes. The annual incidence of SSNHL is 5 to 20 cases per 100,000 individuals worldwide.^{1,2}

■ Etiology. Identifiable causes of SSNHL include viral infections, vascular events, cochlear hydrops, head trauma, tumors (eg, vestibular schwannoma), and demyelinating disorders. Bilateral SSNHL can be seen in autoimmune diseases and rarely can be caused by medications, such as aminoglycosides or certain chemotherapy medications. However, 90% of cases of SSNHL are considered idiopathic because the etiology cannot be determined.¹

■ Diagnosis. The initial evaluation should include an otoscopic examination, tuning fork tests, and pure tone audiometry.¹⁻³ Weber and Rinne tests are essential when evaluating patients for unilateral hearing loss and determining the type of loss (ie, sensorineural

➤ **The most common symptoms of sudden sensorineural hearing loss include not only acute hearing loss, but also tinnitus and fullness in the affected ear.**

vs conductive). The Weber test (ideally using a 512-Hz tuning fork) can detect either conductive or sensorineural hearing loss. In a normal Weber test, the patient should hear the vibration of the tuning fork equally in both ears. The tuning fork will be heard in both ears in conductive hearing loss but will only be heard in the unaffected ear if sensorineural hearing loss is present. So, for instance, if a patient has a perforation in the right tympanic membrane causing conductive hearing loss in the right ear, the tuning fork would be heard in both ears. If the patient has sensorineural hearing loss in the right ear, the tuning fork would only be heard in the left ear.

The Rinne test compares the perception of sound waves transmitted by air conduction vs bone conduction and serves as a rapid screen for conductive hearing loss. In a positive Rinne test, the patient should be able to hear the vibrating tuning fork next to the pinna louder than when placed against the mastoid bone (ie, air conduction greater than bone conduction). In a negative Rinne test, bone conduction is greater than air conduction, and a conductive hearing loss is present. In our patient, pure tone audiometry, tympanometry, and speech audiometry results were consistent with SSNHL.

Magnetic resonance imaging (MRI) of the brain and brainstem with gadolinium contrast can reveal vascular events (thrombotic or hemorrhagic), demyelinating disorders, or retrocochlear lesions such as vestibular schwannoma and is indicated in all cases of suspected SSNHL.^{4,5} An MRI should be obtained within 6 weeks of the initial presentation—even if the patient's hearing returns to normal after treatment.⁴

■ **Treatment and management.** The current standard of care for treatment of idiopathic SSNHL is systemic steroids.^{1,2} Although the gold standard currently is oral prednisolone or methylprednisolone (1 mg/kg/d for 10 to 14 days with a taper,^{1,2} the evidence for this regimen stems from a single placebo-controlled trial (N = 67) that demonstrated greater improvement in the steroid group compared with the placebo group (61% vs 32%).⁶ A Cochrane review and other systematic analyses have not demonstrated

clear efficacy of corticosteroid treatment for the management of idiopathic SSNHL.^{7,8}

Because of the potential systemic adverse effects associated with oral corticosteroids, intratympanic (IT) corticosteroids have been advocated as an alternative treatment option. A prospective, randomized, noninferiority trial comparing the efficacy of oral vs IT corticosteroids for idiopathic SSNHL found IT corticosteroids to be noninferior to systemic treatment.⁹ IT treatment also has been advocated as a rescue therapy for patients who do not respond to systemic treatment.¹⁰

A combination of oral and IT corticosteroids was investigated in a retrospective study analyzing multiple treatment modalities.¹⁰ Researchers first compared 122 patients receiving one of 3 treatments: (1) IT corticosteroids, (2) oral corticosteroids, and (3) combination treatment (IT + oral corticosteroids). There was no difference in hearing recovery among any of the treatments. Fifty-eight patients who were refractory to initial treatment were then included in a second analysis in which they were divided into those who received additional IT corticosteroids (salvage treatment) vs no treatment (control). There was no difference in hearing recovery between the 2 groups. The authors concluded that IT corticosteroids were as effective as oral treatment and that salvage IT treatment did not add any benefit.¹⁰

The American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS) recently published guidelines on the diagnosis and management of SSNHL.¹¹ The guidelines state that IT steroids should be considered in patients who cannot tolerate oral steroids, such as patients with diabetes. It is important to note, however, that the high cost of IT treatment (~\$2000 for dexamethasone or methylprednisolone vs < \$10 for oral prednisolone) is an issue that needs to be considered as health care costs continue to rise.

■ **Antivirals.** Because an underlying viral etiology has been speculated as a potential cause of idiopathic SSNHL, antiviral agents such as valacyclovir or famciclovir also are potential treatment agents.¹² Antiviral medications have minimal adverse effects and are relatively inexpensive, but the benefits

have not yet been proven in randomized controlled trials, and they currently are not endorsed by the AAO-HNS in their guidelines for the management of SSNHL.¹¹

■ **Spontaneous recovery** occurs in up to 40% of patients with idiopathic SSNHL. As many as 65% of those who experience recovery do so within 2 weeks of the onset of symptoms, regardless of treatment.^{1,2} Treatment beyond 2 weeks after onset of symptoms is unlikely to be of any benefit, although some otolaryngologists will treat for up to 6 weeks after the onset of hearing loss.

A substantial number of patients with SSNHL may not recover. Management of these patients begins with referral to an appropriate specialist to initiate counseling and lifestyle changes. Depending on the degree of hearing loss, audiologic rehabilitation may include use of a traditional or bone-anchored hearing aid or a frequency-modulation system.^{1,2,11} Tinnitus retraining therapy might be of benefit for patients with persistent tinnitus.¹¹

■ **Our patient.** After a discussion of his treatment options, our patient decided on a combination of oral prednisolone (60 mg once daily for 9 days followed by a taper for 5 days) and intratympanic dexamethasone injections (1 mL [10 mg/mL] once weekly for 3 weeks). Additionally, antiviral treatment with oral valacyclovir (2 g every 8 hours for 7 days) was initiated per the patient's request (but is not currently recommended by AAO-HNS).

The rationale for this approach was the minimal adverse effects associated with short-term (ie, days to 1–2 weeks) use of high-dose (ie, > 30 mg/d) corticosteroids. Although steroid therapy has been associated with adverse effects such as aseptic necrosis of the hip, these complications usually arise after longer periods (ie, months to years) of high-dose steroid therapy with a mean cumulative dose much higher than what was used in our patient.¹³

Our patient noticed slight improvement within 48 hours of the initial onset of symptoms that continued for the next several weeks until full recovery was attained. An MRI performed 5 days after the onset of symptoms was negative for retrocochlear pathology.

THE TAKEAWAY

SSNHL is a medical emergency that requires prompt recognition and diagnosis. The steps in evaluating sudden hearing loss include: (1) appropriate history and physical examination (eg, otoscopic examination, tuning fork tests), (2) urgent audiometry to confirm hearing loss, (3) immediate referral to an otolaryngologist for further testing (eg, tympanometry, blood tests, MRI), and (4) initiation of treatment.

If a specific etiology is identified (eg, vestibular schwannoma), the patient should be referred to a specialist for appropriate treatment. If there is no identifiable cause (idiopathic SSNHL), the patient should be treated with oral and/or intratympanic steroids. Patients who do not recover following treatment should be offered audiologic rehabilitation. **JFP**

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References

- Schreiber BE, Agrup C, Haskard DO, et al. Sudden sensorineural hearing loss. *Lancet*. 2010;375:1203-1211.
- Rauch SD. Clinical practice. Idiopathic sudden sensorineural hearing loss. *N Engl J Med*. 2008;359:833-840.
- Paul BC, Roland JT Jr. An abnormal audiogram. *JAMA*. 2015;313:85-86.
- Aarnisalo AA, Suoranta H, Ylikoski J. Magnetic resonance imaging findings in the auditory pathway of patients with sudden deafness. *Otol Neurotol*. 2004;25:245-249.
- Cadoni G, Cianfoni A, Agostino S, et al. Magnetic resonance imaging findings in sudden sensorineural hearing loss. *J Otolaryngol*. 2006;35:310-316.
- Wilson WR, Byl FM, Laird N. The efficacy of steroids in the treatment of idiopathic sudden hearing loss. A double-blind clinical study. *Arch Otolaryngol*. 1980;106:772-776.
- Wei BPC, Stathopoulos D, O'Leary S. Steroids for idiopathic sudden sensorineural hearing loss. *Cochrane Database Syst Rev*. 2013. doi:10.1002/14651858.CD003998.pub3.
- Conlin AE, Parnes LS. Treatment of sudden sensorineural hearing loss: II. a meta-analysis. *Arch Otolaryngol Head Neck Surg*. 2007;133:582-586.
- Rauch SD, Halpin CE, Antonelli PJ, et al. Oral vs intratympanic corticosteroid therapy for idiopathic sudden sensorineural hearing loss: a randomized trial. *JAMA*. 2011;305:2071-2079.
- Lee KH, Ryu SH, Lee HM, et al. Is intratympanic dexamethasone injection effective for the treatment of idiopathic sudden sensorineural hearing loss? *J Audiol Otol*. 2015;19:154-158.
- Stachler RJ, Chandrasekhar SS, Archer SM, et al. Clinical practice guideline: sudden hearing loss. *Otolaryngol Head Neck Surg*. 2012;146(3 suppl):S1-S35.
- Westerlaken BO, Stokroos RJ, Dhooge JJ, et al. Treatment of idiopathic sudden sensorineural hearing loss with antiviral therapy: a prospective, randomized, double-blind clinical trial. *Ann Otol Rhinol Laryngol*. 2003;112:993-1000.
- Nowak DA, Yeung J. Steroid-induced osteonecrosis in dermatology: a review [published online March 30, 2015]. *J Cutan Med Surg*. 2015;19:358-360.

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