ORIGINAL ARTICLE

Vestibular Schwannoma in Patients with Sudden Sensorineural Hearing Loss

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ABSTRACT

Sudden sensorineural hearing loss (SSNHL) has several etiologies. It may be a presenting symptom of vestibular schwannoma (VS). This study aimed to establish the incidence of VS in patients with SSNHL, and we report several unusual cases among these patients. We reviewed retrospectively the charts and magnetic resonance imaging (MRI) findings of all adult patients who presented with SSNHL between 2002 and 2008. We utilized three-dimensional fast imaging with steady-state acquisition temporal MRI as a screening method. Of the 295 patients with SSNHL, VS was found in 12 (4%). All patients had intrameatal or small to medium-sized tumors. There were three cases with SSNHL in one ear and an incidental finding of intracanalicular VS in the contralateral ear. There were four cases of VS that showed good recovery from SSNHL with corticosteroid treatment. There were two cases that mimicked labyrinthitis with hearing loss and vertigo. A greater number of cases than expected of VS were detected in patients with SSNHL, as a result of increasing widespread use of MRI. Various unusual findings in these patients were identified. MRI would seem to be mandatory in all cases of SSNHL.

KEYWORDS: Sudden hearing loss, vestibular schwannoma, magnetic resonance imaging, steroid, vertigo

Sudden sensorineural hearing loss (SSNHL) can be caused by various etiologies, including infectious diseases, neurological disorders, ototoxic agents, immunologic causes, head injury, and neoplasms. Approximately 1% of cases of SSNHL are caused by retrocochlear lesions that may be related to neoplasms, demyelinating disease, or stroke.[1] Recently, as a result of the increasing widespread use of magnetic resonance imaging (MRI), more patients with vestibular schwannoma (VS) than expected have been detected among those with SSNHL.

The most common symptom of VS is hearing loss on the affected side. Typically, hearing loss develops gradually, but SSNHL is found in 7 to 20% of patients.[2–4] This study aimed to establish the incidence of VS in patients with SSNHL, and we report several unusual findings among these patients.

MATERIALS AND METHODS

We reviewed retrospectively the charts and MRI findings of all patients who were admitted to the Department of Otorhinolaryngology–Head and Neck Surgery at Soonchunhyang University Bucheon Hospital, Korea, with SSNHL between 2002 and 2008. We defined SSNHL as acute onset of hearing loss of 30 dB at three contiguous frequencies, which occurred instantaneously or progressively over several days. Patients with

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Skull Base 2011;21:75–78.

Received: January 23, 2010. Accepted after revision: April 12. 2010. Published online: July 21, 2010.
DOI: http://dx.doi.org/10.1055/s-0030-1261257.
ISSN 1531-5010.

Table 1 Summary of Patients with VS Presenting with SSNHL

Case no.	Sex	Age (years)	Size (mm)
1	M	55	14.93
2	M	56	3.87
3	M	47	9.61
4	F	55	15.42
5	F	33	16.07
6	F	67	8.78
7	F	69	intrameatal
8	F	44	8.83
9	M	59	intrameatal
10	M	32	5.98
11	F	39	intrameatal
12	F	53	12.72

F, female; M, male.

SSNHL were evaluated with MRI to rule out a retrocochlear lesion. We used three-dimensional (3-D) fast imaging with steady-state acquisition (FIESTA) temporal MRI as a screening method instead of contrast-enhanced MRI. Among these patients with SSNHL, we studied those with VS. According to the recommendations of the Consensus Meeting on Systems for Reporting Results in Acoustic Neuroma in Tokyo, intrameatal tumor and the largest extrameatal diameter were used.

RESULTS

Of the 295 patients with SSNHL, VS was found in 12 (4%). There were seven women and five men aged 32 to 69 years (mean, 50.7 years). All patients had intrameatal or small to medium-sized tumors. The patients' profiles are summarized in Table 1.

Patients with SSNHL in One Ear and VS in the Contralateral Ear

There were three cases of SSNHL in one ear and an incidental finding of VS in the contralateral ear (Table 2). Two patients (cases 1 and 2) had already nonserviceable hearing loss in ears with tumor and did not recover hearing in ears with SSNHL. Hearing rehabilitation involved hearing aids in the ears with SSNHL. Case 3 showed good hearing in the ear with a tumor and slightly recovered hearing in the ear with SSNHL. We observed his hearing carefully without any hearing rehabilitation.

Patients with VS Showed Hearing Recovery with Corticosteroids

There were four cases of VS that showed good recovery from SSNHL with corticosteroid treatment (Table 3). Cases 4 and 5 showed nonserviceable hearing initially but complete recovery after treatment with corticosteroids. Hearing preservation surgery via the suboccipital approach was undertaken. Although other patients (cases 6 and 7) showed hearing recovery, they were treated with a "wait and see" approach because of their old age.

Patients with VS Mimicking Labyrinthitis

There were two cases of VS that mimicked labyrinthitis; both patients had hearing loss and vertigo (Table 4). Spontaneous nystagmus was identified, and the direction of nystagmus was contralateral. Spontaneous nystagmus was sustained for several weeks in case 8. Tumor removal via the translabyrinthine approach was performed, and the symptom of vertigo improved rapidly after surgery.

Table 2 Summary of Patients with SSNHL in One Ear and VS in the Contralateral Ear

Case no.	Site (sudden)	Site (tumor)	PTA/SD (sudden)	PTA/SD (tumor)	F/U (sudden)	Tumor Treatment	Hearing Rehab
1	R	L	60/32	60/12	no response	GK	HA (R)
2	R	L	75/24	deaf	no response	GK	HA (R)
3	L	R	85/0	25/84	slight recovery	Observation	none

L, left; GK, gamma knife; HA, hearing aid; PTA, pure tone audiometry; R, right; SD, speech discrimination.

Table 3 Summary of Patients with VS who Showed Hearing Recovery with Corticosteroid Treatment

Case no.	Initial PTA (dB)	Initial SD (%)	Final PTA	Final SD	Treatment
4	45	44	25	82	Surgery (SOC)
5	56	24	20	96	Surgery (SOC)
6	70	4	30	64	Observation
7	65	4	30	68	Observation

SOC, suboccipital approach.

Table 4 Summary of Patients with VS Mimicking Labyrinthitis

Case no.	Chief Complaint	PTA (dB)	SD (%)	ENG	Canal Paresis	Treatment
7	Dizziness	65	4	SN(+) Rt	56% (L)	Observation
8	Dizziness	35	24	SN(+) Lt	100% (R)	Surgery (TLA)

ENG, electronystamography; SN, spontaneous nystagmus; TLA, translabyrinthine approach.

DISCUSSION

The symptoms of VS are highly variable and include hearing loss, dizziness, and tinnitus. Progressive hearing loss is the most common symptom of VS, and ~1% of patients with SSNHL are found to have VS. The higher percentage (4%) in our study seems to be attributed to widespread use of MRI. Gadolinium-enhanced MRI is recognized widely as the gold standard in the diagnosis of VS. However, we recommend 3-D FIESTA MRI for all patients with SSNHL because of its cost-effectiveness. Daniels et al[5] reported that nonenhanced fast spin echo (FSE) MRI was a more cost-effective screening method than contrast-enhanced MRI. Hatipoğlu et al[6] reported that 3-D FIESTA MRI was superior to FSE MRI for imaging the cisternal parts of the posterior fossa nerves.

Various mechanisms such as microvascular embarrassment in the cochlea, conduction block of the cochlear nerve, and endolymphatic hydrops have been proposed as a cause of SSNHL in patients with VS.[7] Yanagihara and Asai[4] have reported that small tumors tend to trigger SSNHL more frequently than medium and large tumors. However, Moffat et al[8] have reported that there is no tumor size difference between patients with SSNHL and others. Our finding that all patients had intrameatal or small- to medium-sized tumors is consistent with previous studies.

We found three patients with SSNHL in one ear and an incidental finding of VS in the contralateral ear. These patients did not actually have VS presenting as SSNHL. The otologist is confronted with a difficult situation if SSNHL does not recover. Hearing rehabilitation such as hearing aids or cochlear implants may be considered for such patients. The options available to treat VS include conservative management, radiosurgery and surgical excision, although it is acknowledged that either option may precipitate hearing loss on the tumor side. Although a cochlear implant should be considered in the ear with SSNHL once bilateral profound hearing loss has occurred, it may be considered in the tumor side first if the cochlear nerve can be preserved while removing the tumor.[9] Two patients who did not respond to corticosteroid treatment wore a hearing aid on the same side as their SSNHL. Since only one patient had serviceable hearing on the tumor side and poor word recognition in the ear with SSNHL, he did not wear a hearing aid following hearing level periodically. We think that audiological management is very difficult in patients with SSNHL in one ear and VS in the contralateral ear.

There have been several reports that hearing recovers with corticosteroid treatment in patients with VS presenting as SSNHL.[10–12] These studies have noted that hearing recovery does not exclude the presence of VS. We observed four patients with VS who showed good recovery from SSNHL with corticosteroid treatment. Two of these elected conservative management and the others underwent hearing preservation surgery. We recommend that corticosteroid treatment should be considered, even if SSNHL coexists with VS, because hearing conservation therapy may then become possible.

The vestibular symptom associated with VS is usually disequilibrium and true vertigo is far less common.[13] SSNHL with vertigo (cochleovestibular loss) can be caused by inflammation of the labyrinth. We experienced two cases of VS that mimicked labyrinthitis, which showed acute hearing loss on one side and vertigo. These cases showed horizontal nystagmus beating toward the healthy ear, and a caloric test revealed a decreased response in the affected ear. Godefroy et al[14] have suggested that surgical treatment should be considered in patients with small-sized tumors and persistent disabling vertigo. Our one patient with persistent vertigo recovered shortly after translabyrinthine surgery.

CONCLUSION

A greater number of cases of VS than expected were detected in patients with SSNHL, because of increasing widespread use of MRI. Our study identified several unusual findings in these patients with SSNHL. We also strongly recommend that all cases of SSNHL should be evaluated with MRI.

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