Acute Deafness: A Rare Complication of Shunting

Rafael Martinez-Perez[1,2], Agustin Montivero[1], Natalia Rayo[3], Marco Ramirez[1], Cristian Naudy[1], Jorge Mura[1]

Key words
- Deafness
- Hearing loss
- Hydrocephalus
- Overshunting
- VP shunt

Abbreviations and Acronyms
CSF: Cerebrospinal fluid
MRI: Magnetic resonance imaging
VP: Ventriculoperitoneal

From the [1]Division of Skull Base and Vascular Neurosurgery, National Institute of Neurosurgery Dr Asenjo, Universidad de Chile, Santiago, Chile; [2]Department of Neurosurgery, Hospital 12 de Octubre, Universidad Complutense, Madrid, Spain; and [3]Western University, London, Ontario, Canada

To whom correspondence should be addressed: Rafael Martinez-Perez, M.D., Ph.D. [E-mail: rafa11safin@hotmail.com]

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BACKGROUND: **Mild hearing loss following shunting has been described; however, severe auditory impairment associated with ventriculoperitoneal (VP) shunt is an uncommon, rarely reported phenomenon. Treatment options and pathophysiologic considerations are discussed in this case report.**

CASE DESCRIPTION: **A 27-year-old man who was treated for an eighth cranial nerve schwannoma with complete resection and a VP shunt 10 years previously presented to the emergency department with acute severe hearing loss and headache. Imaging showed diminished size of the ventricles and dural contrast enhancement. The previous shunt was replaced with a programmable antisiphoning VP shunt. The patient's hearing and headache improved 48 hours later, as demonstrated in serial audiograms.**

CONCLUSIONS: **Hearing loss is an underestimated complication of shunting that in some cases may progress to severe impairment and deafness. Patients with a VP shunt who experience hearing loss should undergo further evaluation and possibly adjustment of shunt settings.**

INTRODUCTION

Ventriculoperitoneal (VP) overshunting[1] is a very rare condition produced by the siphoning effect caused by the column of the cerebrospinal fluid (CSF) tube when the patient is erect.[2] Auditory symptoms and mild hearing loss are uncommon complications associated with VP shunt insertion that have been rarely reported in the literature; however, acute deafness after a prolonged time of normal function of the VP shunt is not considered to be a clinical manifestation of overshunting in patients with hydrocephalus. We report a case of documented acute severe hearing loss associated with VP overdrainage and resolution of the symptoms after shunt replacement. Diagnostic and treatment options for this unusual condition are presented and discussed.

CASE DESCRIPTION

A 27-year-old man presented to the emergency department with a headache, tinnitus, and progressive severe hearing loss within the 72 hours before admission. He had undergone retrosigmoid craniotomy and internal acoustic meatus drilling 10 years previously for complete resection of a left KOOS grade IVB acoustic schwannoma and VP shunt placement (Figure 1). After the procedure the patient developed left facial palsy (House-Brackmann grade IV) and left ear deafness. During the follow-up period, the patient remained without changes in his neurologic condition and without any evidence of shunt dysfunction (Figure 2).

The headaches were bifrontal and remitted when the patient was in the recumbent position. They were occasionally so disabling that the patient could not attend school; however, the patient was more concerned about the right ear hearing loss and tinnitus. Results of limb examination were normal, and cranial nerve examination revealed left facial palsy and bilateral hearing impairment. Formal audiologic evaluation demonstrated severe right-sided sensorineural hearing loss (Figure 3).

Magnetic resonance imaging (MRI) suggested overshunting, with significantly smaller ventricles than on previous MRI and dural contrast enhancement (Figure 4). There was no evidence of tumor recurrence or any other lesion at the right internal acoustic meatus (Figure 4C). There was no meningocele, fluid in the middle ear, or any sign of CSF leak on cranial or spinal MRI. Computed tomography scan with contrast medium failed to demonstrate any CSF leak or bone defect along the anterior or middle fossa.

The patient underwent replacement of the VP shunt with a programmable VP shunt with antisiphoning effect. The opening pressure at the time of surgery measured through the proximal ventricular catheter was 1 cm H_2O. The VP shunt was tested following the removal and demonstrated anterograde flow through the device at small increases in positive pressure. Postoperatively, the patient immediately reported improvement of hearing and headaches. On day 2, formal audiologic evaluation demonstrated great improvement of hearing function, and computed tomography scan showed mild enlargement of the lateral ventricles. At the 3-month follow-up, the patient remained stable with no headaches or hearing loss. Follow-up MRI showed clearance of dural contrast enhancement.

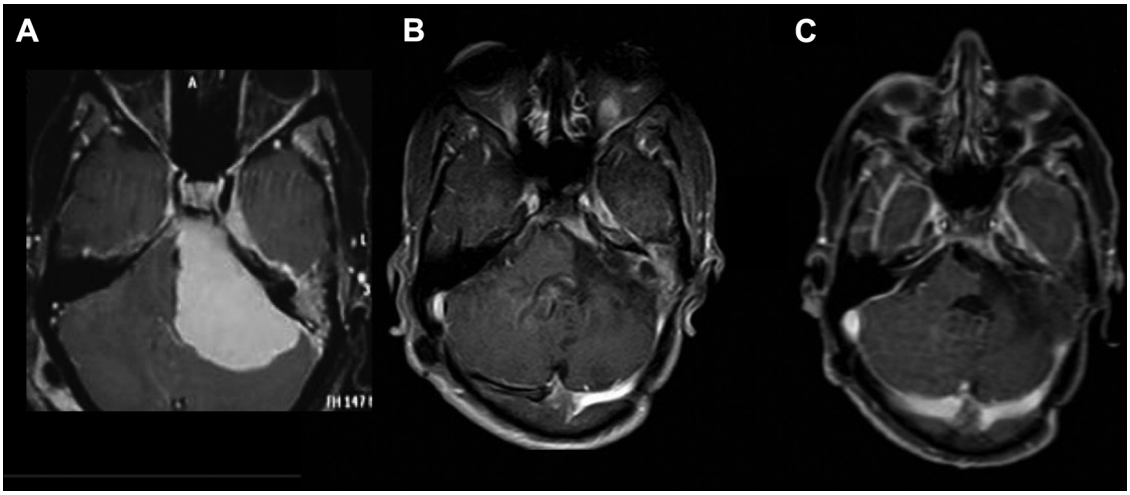

Figure 1. Preoperative (**A**) and immediate postoperative magnetic resonance imaging (MRI) (**B**) performed 10 years before the most recent admission. Preoperative MRI showed a sphenopetrous meningioma with complete occlusion of the fourth ventricle and mass effect over the brainstem. Postoperative MRI showed subtotal resection with adequate decompression of the brainstem and no other significant findings. Current MRI performed at admission (**C**) did not show evidence of meningocele or any other signs of fistula, although it was remarkable for diffuse dural contrast enhancement.

DISCUSSION

Although shunting or hydrocephalus is not traditionally considered in the differential diagnosis of hearing loss, some recent evidence indicates that CSF circulation abnormalities and changes in pressure can produce auditory dysfunction.[3-6] In 1 study, 10 of 12 patients treated with VP shunt for hydrocephalus experienced a high-frequency cochlear hearing loss.[6] Lim et al.[4] observed that 40% of ears of patients treated with VP shunt experienced a threshold elevation of at least 15 dB in ≥1 frequencies, and other studies have shown similar results.[3,5] Recent articles have suggested that this complication is more commonly associated with pediatric age[5-7] and that the hearing loss is more dramatic on the side where the VP shunt was deployed.[6] The presumed explanation for this phenomenon is that the decrease in intracranial pressure produced by shunting is transduced into the endolymph causing a small pressure difference between endolymph and perilymph. These pressure differentials may cause displacement of the Reissner membrane, affecting hearing.[7,8]

According to Lee et al.,[9] auditory impairment is an underestimated complication, rather than a rare event in

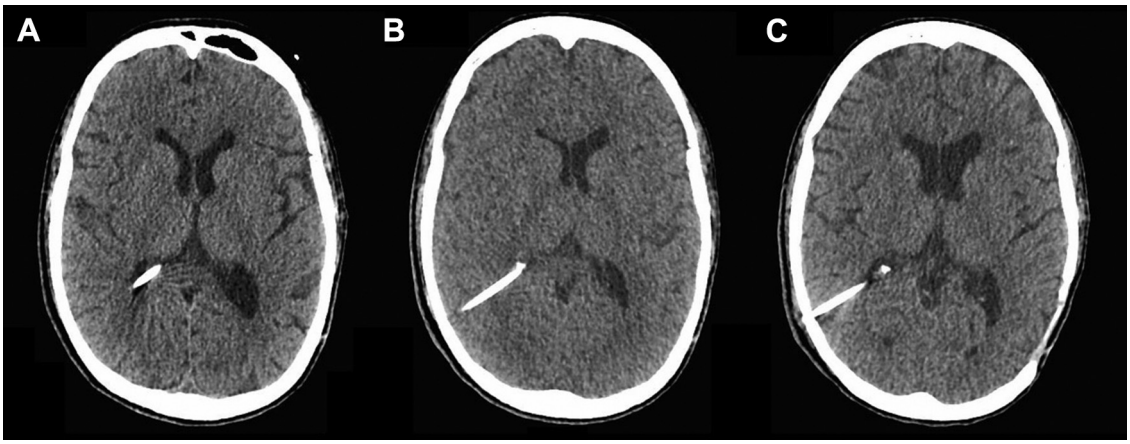

Figure 2. Head computed tomography scans performed over the follow-up period show changes in ventricle sizes. (**A**) Baseline computed tomography scan performed 5 years before present admission showed optimized position of the ventricular catheter and normal ventricle sizes without signs of shunt dysfunction. (**B**) Computed tomography scan at admission showed the typical images of slit ventricles, consisting in reduced size of lateral ventricles. (**C**) Postoperative computed tomography scan performed after ventriculoperitoneal shunt replacement demonstrated significant change in diameter of lateral ventricles with size comparable to the baseline computed tomography scan.

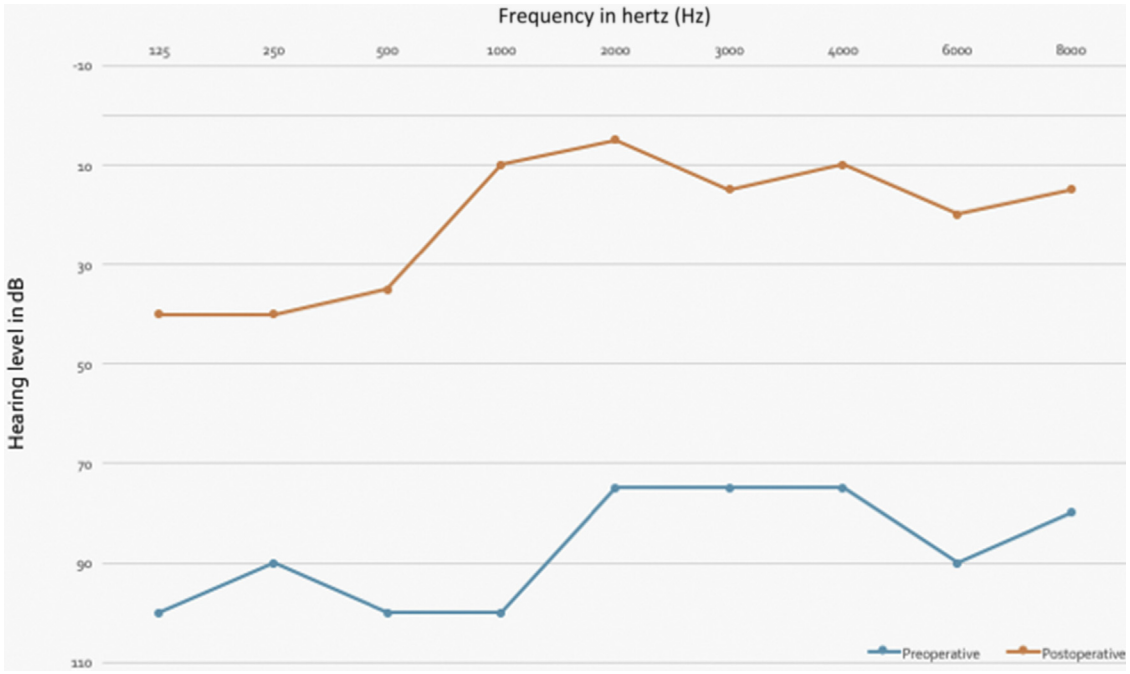

Figure 3. Preoperative (in blue) and postoperative (in red) audiograms of the right ear. Preoperative audiogram showed an average decline of 85 dB with a discrimination of 30% (level D American Academy of Otolaryngology—Head and Neck Surgery Foundation Hearing Classification System). After ventriculoperitoneal shunt placement, the postoperative audiogram reported an average decline of 12.5 dB with 100% word discrimination (level A American Academy of Otolaryngology—Head and Neck Surgery Foundation Hearing Classification System).

patients with VP shunt. However, severe hearing loss has been rarely reported, as the threshold elevations in the audiogram are usually not enough to be clinically significant.[4] Despite the fact that some degree of hearing loss after shunting has been described in a few reports and studies, this used to happen within the acute phase after VP shunt placement.[5,6,10] What is more remarkable about the current case is that hearing loss was severe and it had a sudden onset after 10 years of normal VP shunt function.

Sudden intracranial hypotension and therefore the auditory impairment could

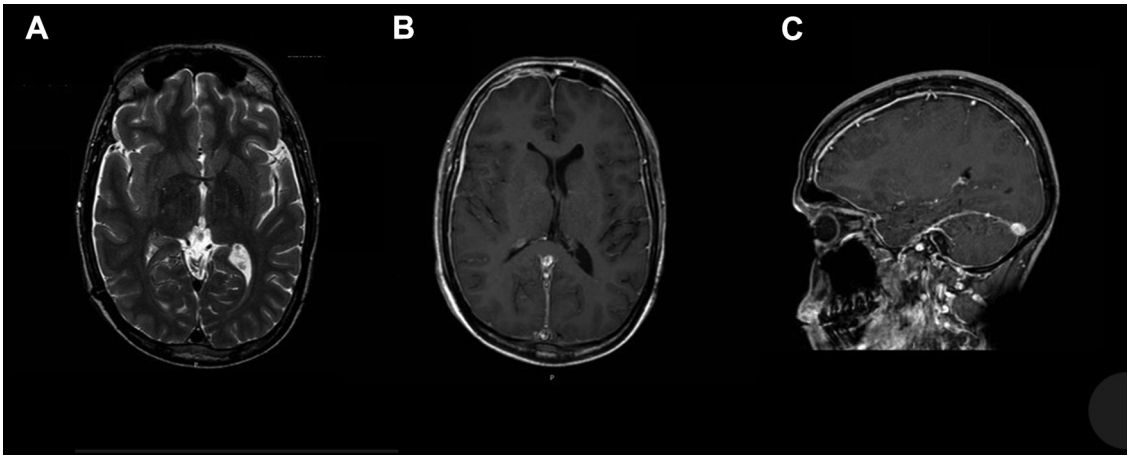

Figure 4. Magnetic resonance imaging of the brain at admission. Axial T2-weighted images showing bilateral increase of the extra-axial space (**A**) and dural contrast enhancement in T1-weighted axial (**B**) and sagittal (**C**) projections, both considered indirect signs of overshunting.

be explained in the current case by the presence of a spontaneous CSF fistula. This mechanism has been proposed in other uncommon sudden complications related to shunting.[11] The patient and family denied any trauma or iatrogenic procedures, and physical examination was normal. Complete spinal and cranial MRI did not reveal the presence of meningocele or CSF leak. We previously reported a case in which a defect in the tegmen tympani was responsible for spontaneous CSF leak causing intracranial hypotension. Likewise, in the current case, the patient underwent a retrosigmoid craniotomy for resection of an eighth cranial nerve schwannoma 10 years previously that required internal acoustic meatus drilling. However, helical computed tomography scan did not show any bone defect in the middle or anterior ears or any evidence of CSF leak.

In the absence of another more reasonable explanation for intracranial hypotension, dysfunction of the VP shunt should be considered as the main possibility. This hypothesis was reinforced after the VP shunt dysfunction was confirmed at the time of surgery. Within the first 6.5 years of initial shunting, 12% of patients may develop overshunting,[12] although it has also been reported at a later stage.[11]

The clinical presentation was also highly suggestive of intracranial hypotension and overshunting. The presence of a headache that is relieved by the recumbent position in the physical examination might support the diagnosis.[1] Slit ventricles and dural contrast enhancement are other important radiologic findings that are often associated with this syndrome.[12] Spontaneous subdural hematomas, pneumocephalus, and microcephaly are concurrent complications that have been described as manifestations of this phenomenon,[11,12] although acute deafness has not been considered in previous studies.

To our knowledge, this is the first case in the literature in which hearing loss occurred suddenly after a long period of normal VP function. In this case, our patient experienced a complete recovery of his previous auditory function following replacement of the VP shunt with a new programmable VP shunt with an antisiphoning effect. This confirms our hypothesis that overshunting was the cause of the auditory impairment.

CONCLUSIONS

Hearing loss is a potential complication of shunting that may lead to severe impairment and deafness. This case suggests that patients with a VP shunt should be monitored for hearing loss. Auditory impairment in these patients should prompt further evaluation and possibly adjustment of shunt settings.

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