Recovery of sensorineural hearing loss following operative management of a posterior fossa arachnoid cyst

Case report

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Arachnoid cysts are benign, intraarachnoid cysts filled with cerebrospinal fluid that are usually encountered in the middle cranial fossa. If present in the posterior fossa, they usually produce nonspecific signs and symptoms such as headaches, dizziness and vertigo. We report the rare presentation of a young girl with right-sided sensorineural hearing loss and tinnitus secondary to a right cerebellopontomedullary arachnoid cyst. The patient underwent a sub-occipital retrosigmoid (retromastoid) craniectomy with fenestration of the arachnoid cyst. Subsequently, the patient experienced improvement in hearing with near-complete resolution of sensorineural hearing loss. To the authors’ knowledge, postoperative near-complete resolution of hearing loss secondary to posterior fossa arachnoid cysts in a pediatric patient has not been previously reported. The authors also review the literature with respect to posterior fossa arachnoid cysts and discuss their clinical features, diagnosis, and management. (DOI: 10.3171/2009.3.PEDS08416)

Key Words • posterior fossa • arachnoid cyst • hearing loss • cerebellopontine angle • tinnitus

A rachnoid or leptomeningeal cysts are benign, intraarachnoid cystic lesions that are filled with CSF.8 Likely developmental in origin and generally asymptomatic, these lesions can become symptomatic as a result of enlargement or intracystic hemorrhage. Although their exact etiology is unknown, various hypotheses have been considered. These hypotheses include active fluid secretion from the cystic wall,10 fluid accumulation secondary to changes in the osmotic gradient,6 and vascular pulsation causing CSF flow between the cyst and subarachnoid space.23 Arachnoid cysts occur throughout the CNS, and while generally no communication is demonstrable with the subarachnoid space, a more recent hypothesis has been suggested based on intraoperative reports of apparent one-way, slit-valve communications between the cyst and subarachnoid space.19,20

Arachnoid cysts represent approximately 1% of all intracranial space-occupying lesions.4 They are typically located in the middle cranial fossa, but other locations including the cerebellopontine angle, cerebellar hemispheres and posterior fossa have been described.5,12,13,17

Symptoms due to arachnoid cysts are generally secondary to mass effect, and consequently in the posterior fossa, these usually manifest as seizures, tremors, Ménière disease, lower cranial nerve deficits, and occasionally headaches secondary to hydrocephalus.2,4,15,16,24

We report the case of a patient with a cerebellopontomedullary arachnoid cyst who initially presented with unilateral sensorineural hearing loss and tinnitus, and who subsequently experienced improvement in hearing following surgical decompression. To our knowledge, this is the first time that recovery of hearing following decompression of a posterior fossa arachnoid cyst in a pediatric patient has been reported. Additionally, we review the relevant literature and discuss the clinical features, diagnosis, and management of posterior fossa arachnoid cysts.

Case Report

History, Presentation, and Examination. This 12-year-old girl, who had been in good health previously, presented to the Boston Medical Center Department of Otolaryngology–Head and Neck Surgery in August 2005 with right-sided hearing loss and tinnitus that had been discovered
during a routine medical examination at school. The patient had no history of any ear problems or vertigo. Family history revealed that her maternal aunt also suffered from unilateral sensorineural hearing loss, although information regarding the exact cause and nature of the condition was unavailable. Results of the patient’s otolaryngological, neurological, and vestibular examinations were normal. Audiometric evaluation revealed mild to moderate, right-sided, high-frequency (3000–8000 Hz) sensorineural hearing loss.

Due to the presence of unilateral sensorineural hearing loss and the patient’s familial history, a CT scan of the IAC, MR imaging of the brain and IAC, and genetic testing were performed. There was no evidence of cochlear dysplasia, nor were there any genetic markers associated with hearing loss. The MR imaging study revealed a 12-mm, nonenhancing, right cerebellopontomedullary cystic lesion that was hyperintense on T2-weighted images (Fig. 1). During the course of her work-up, repeat audiometric evaluation revealed progression of her right-sided high-frequency hearing loss and an increased hearing handicap (Fig. 2A). Given these findings and progression of hearing loss, a hearing aid was obtained to help ameliorate her hearing loss, and she was referred to the Department of Neurosurgery for evaluation of the arachnoid cyst.

**Operation.** The patient underwent a right suboccipital retrosigmoid (retromastoid) craniectomy. Intraoperatively, she was found to have a right cerebellopontomedullary arachnoid cyst that was compressing the right seventh and eighth cranial nerve complex superiorly with resulting stretching of the nerve fibers. The cyst was dissected off the cranial nerves and fenestrated into the adjacent, surrounding subarachnoid cisterns. Following fenestration, the cranial nerves appeared relaxed and free. Somatosensory evoked potential and brainstem auditory evoked response monitoring was conducted throughout the procedure, with no changes in latencies observed.

**Postoperative Course.** The patient recovered from the surgery well, and by October 2006, her tinnitus had resolved, although her sensorineural hearing loss was unchanged. An MR imaging study performed 15 months postoperatively demonstrated a decrease in the size of the arachnoid cyst (Fig. 3).

**Fig. 1.** Preoperative T2-weighted axial MR images demonstrating the hyperintense cystic lesion in the right cerebellopontomedullary region.

**Fig. 2.** Pure tone audiograms of the patient's affected (right) ear. Preoperative examination (A) revealed mild to moderate high frequency (3000–8000 Hz) sensorineural hearing loss which required the use of a hearing aid. A subsequent examination 2 years postoperatively (B) revealed significant improvement in sensorineural hearing loss at 3000 Hz and 6000–8000 Hz. The hearing aid was deemed not required and its use was therefore discontinued.
Recovery of hearing loss after management of an arachnoid cyst

The patient was routinely seen in clinic postoperatively, and in August 2008, presented at the Department of Otolaryngology–Head and Neck Surgery with a complaint that the hearing aid was “too loud.” Audiometric evaluation at this time revealed significant improvement in her sensorineural hearing loss at 3000 Hz and 6000–8000 Hz (Fig. 2B). A follow-up audiometric evaluation a month later confirmed this improvement, and the patient was deemed to have right-sided hearing that was adequate for normal communication. Use of the hearing aid was therefore discontinued.

Discussion

Intracranial posterior fossa arachnoid cysts are uncommon and are frequently asymptomatic or cause nonspecific symptoms such as headache or dizziness.4,5,15,17 On occasion, they may present with specific symptoms related to lower cranial nerve palsies, cerebellar, and pyramidal signs, and these symptoms can aid in localizing the lesion.4,5,16,17,24

Although the exact pathophysiological mechanism for hearing loss in patients with arachnoid cysts has not been well described, vascular compromise has been proposed.13,14 It is our opinion, based on the clinical presentation and response, that the patient in this report was symptomatic as a result of the arachnoid cyst exerting mass effect on the right seventh and eighth cranial nerve complex.

There have been other reported cases of posterior fossa arachnoid cysts involving patients who presented with hearing loss. Thinakara-Rajan et al.22 reported the case of a 29-year-old woman with a history of right-sided sensorineural hearing loss and otalgia. Magnetic resonance imaging revealed a 5 × 7–cm arachnoid cyst in the right cerebellum with associated mass effect. Ottaviani et al.14 reported on a 21-year-old man with left-sided tinnitus and hypacusia, who had a left cerebellar convexity arachnoid cyst that was displacing the cerebellar hemisphere and subsequently causing cerebellopontine angle compression. Likewise, the patient reported on by Cadoni et al.1 presented with sudden cochlear hearing loss. Haberkamp et al.4 presented 3 cases of arachnoid cysts in the posterior fossa, each with different presenting symptoms, including one with a unique presentation of bilateral cochlear hearing loss. Patients with posterior fossa arachnoid cysts presenting with seizures, tremors, Ménière disease, and facial paralysis have also been described.2,16,21,24

To our knowledge, there has been only one prior reported case of recovery of hearing loss in a patient with a posterior fossa arachnoid cyst. In the case report, Lanzino et al.9 described a 53-year-old patient with a unilateral cerebellopontine angle arachnoid cyst and associated hearing loss who experienced recovery of his hearing following decompression and marsupialization of the cerebellopontine angle arachnoid cyst.

Magnetic resonance imaging is the preferred diagnostic imaging modality in these cases due to its ability to precisely locate and define the arachnoid cyst. The differential diagnosis should include meningiomas, acoustic neuromas, cholesteatomas, metastases, loculated subdural hygromas, and other cystic lesions such as epidermoid cysts.13

The management of arachnoid cysts continues to remain controversial. In general, the accepted practice (and our practice, as well) is to not pursue any form of surgical management for patients with asymptomatic arachnoid cysts.5,11 Symptomatic arachnoid cysts may be treated by a variety of surgical procedures, including fenestration into the adjacent subarachnoid spaces, drainage, removal of the cyst, external shunting, and endoscopic decompression.3,4,7,18,20

With the exception of one case report involving an adult,8 in all previous reports of patients with hearing loss secondary to posterior fossa arachnoid cysts, minimal to no improvement in the hearing was appreciated postoperatively.1,14,22 This lack of improvement was attributed to irreversible damage being caused by long-standing compression of the cerebellopontine angle.14,22 To our knowledge, the case described in the present paper is the first known case of an arachnoid cyst in a pediatric patient in which there was postoperative improvement in sensorineural hearing loss and near-complete recovery. Nevertheless, restraint must be used in generalizing from the satisfying outcome of this case to cases involving other patients with similar presentations, as one cannot guarantee their recovery from hearing loss. It is our opinion that screen-
ing audiology should be considered early in patients with cerebellopontomedullary or cerebellopontine angle arachnoid cysts even if they do not complain of hearing loss, with treatment being recommended for those patients with progressive hearing loss and evidence of vestibulocochlear nerve compression. Further, as seen in this case report, long-term follow-up must be conducted, with particular attention to the patient’s hearing status.

Disclaimer

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

References

18. Raffel C, McComb JG: To shunt or to fenestrate: which is the best surgical treatment for arachnoid cysts in pediatric patients? Neurosurgery 23:338–342, 1988

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