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Acute sensorineural hearing loss resulting from cerebellopontine angle arachnoid cyst

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We present the case of a 49-year-old woman who presented with acute, nonprogressive left sensorineural hearing loss and benign positional vertigo that was associated with an arachnoid cyst of the cerebellopontine angle. The presence of the lesion was documented by MRI examinations that were obtained 7 years apart. Arachnoid cysts at the cerebellopontine angle are usually found incidentally on MRI performed for unrelated reasons. However, if the arachnoid cyst displaces or compresses adjacent cranial nerves, symptoms may result. We review the salient imaging features of arachnoid cysts that allow their differentiation from other lesions of the cerebellopontine angle.

Case report

A 49-year-old woman presented with a seven-year history of nonprogressive, acute left-sided hearing loss. Upon initial presentation, she denied vertigo, headaches, visual disturbance, or other symptoms. Physical examination was normal. Audiology examination demonstrated sensorineural hearing loss in the left ear with no word-recognition abilities. Magnetic resonance imaging (MRI) of the brain without contrast at an outside hospital was dictated as high-signal-intensity T2 mass of the cerebellopontine angle (CPA) representing either an epidermoid cyst or arachnoid cyst. The patient returned 7 years later with new positional vertigo and dizziness. The vertigo waxed and waned, and appeared after activities such as rolling over in bed, looking up to reach a shelf, and bending. On physical examination, the patient had new rotary nystagmus in supine left-ear dependent position.

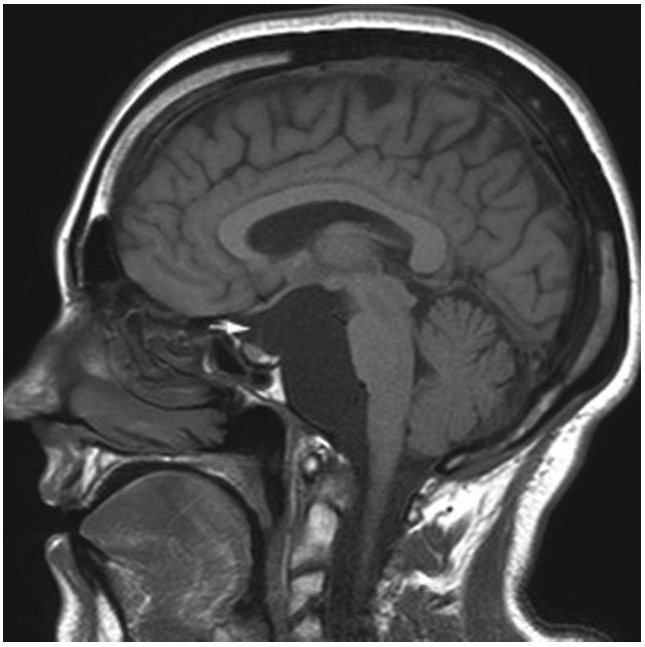

Figure 1. 49-year-old woman with sensorineural hearing loss. Sagittal T1 image demonstrates a mass isointense to CSF in the prepontine cistern that extends into the suprasellar region. The mass posteriorly displaces the pons, anteriorly bows the pituitary stalk (white arrow), and superiorly displaces the optic chiasm.

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Repeat MRI with and without contrast showed no change in a 2.5 x 4.9 x 4.4-cm low-T1 and very-high-T2-signal-intensity mass (Figs. 1 and 2) centered in the prepontine cistern. Signal characteristics mirrored those of cerebrospinal fluid (CSF) on all sequences. The lesion extended into both CPA cisterns and the suprasellar region. The mass superiorly displaced the optic chiasm and optic nerve, with anterior bowing of the pituitary stalk (Fig. 1), and lateral bowing of the trigeminal nerve, facial nerve, and vestibularcochlear nerves bilaterally (Fig. 3). There was mild adjacent-mass effect seen as mild flattening of the anterior pons. There was no postgadolinium enhancement (Fig. 4) or restricted diffusion (Fig. 5). Findings were most consistent with an arachnoid cyst.

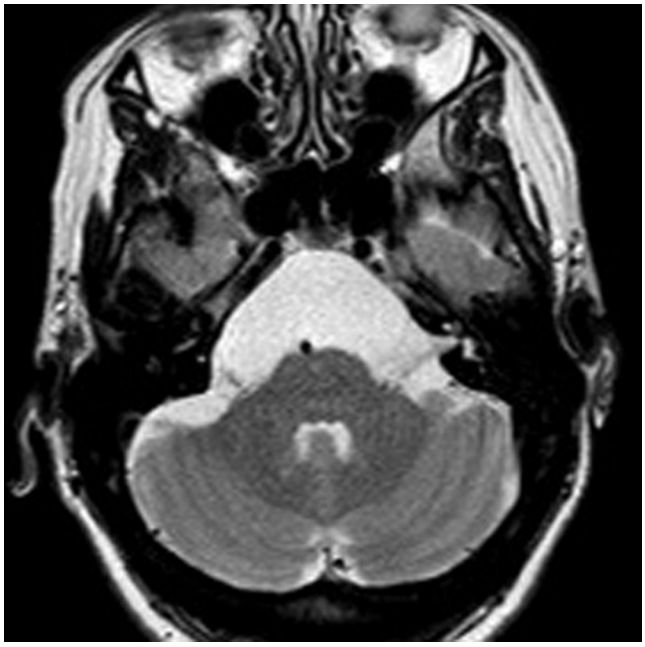

Figure 2. 49-year-old woman with sensorineural hearing loss. Axial T2 image demonstrates a high-signal-intensity mass of CSF intensity in the prepontine cistern and CPA cisterns. There is mild flattening of the anterior pons due to the mass but no edema.

Due to patient acceptance of the sensorineural hearing loss over the 14-year period and unchanged size of the arachnoid cyst, the patient was referred to a vestibular specialist to assist with vertigo. No surgical intervention was planned.

Discussion

Arachnoid cysts are benign, pouch-like congenital lesions that are postulated to occur from the splitting of embryonic meninges and thus are filled with CSF (1). They account for approximately 1% of all intracranial masses, with the majority located in the middle cranial fossa (2). Only 10% are found in the posterior fossa, most in the CPA (3). Although

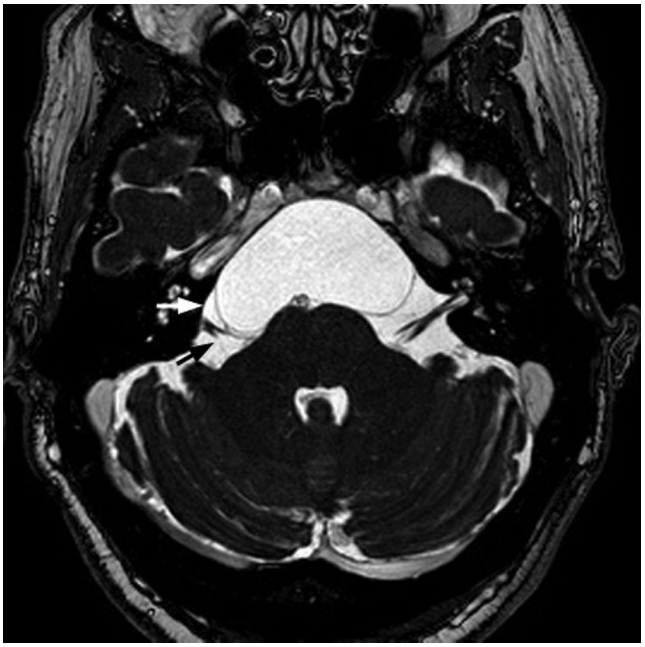

Figure 3. 49-year-old woman with sensorineural hearing loss. Axial MR cisternogram image demonstrates the high-signal-intensity mass, following CSF signal characteristics, in the CPA and prepontine cisterns. There is a thin membrane marginating the cisternal lesion (white arrow), and mild lateral and posterior displacement of the vestibulocochlear nerves (black arrow). The mass is not encasing the nerves or vascular structures, which would be more typical of an epidermoid cyst than arachnoid cyst.

they are usually asymptomatic, focal neurological symptoms may arise when the cysts exhibit displacement of adjacent cranial nerves or other cisternal structures, such as in our patient. Differentiation of a symptomatic arachnoid cyst from a vestibular schwannoma, the most common CPA tumor to result in sensorineural hearing loss, can easily be performed with contrast administration. Arachnoid cysts are nonenhancing lesions, while vestibular schwannomas exhibit homogeneous, heterogeneous, or(less commonly) cystic enhancement (4-6).

Bonneville et al. (3, 7) present a schema to characterize and limit differential diagnosis of CPA tumors based on presence or absence of enhancement. Nonenhancing lesions are further divided into those that have high or low signal on T1-weighted MR images. The three nonenhancing CPA lesions that are low signal on T1-weighted MR images are epidermoid cysts, arachnoid cysts, and neurocysticercosis (7).

Arachnoid cysts follow the signal intensity of CSF on all sequences. Thus, they are hypointense signal on T1 and hyperintense signal on T2. This pattern most commonly mimics an epidermoid cyst, which is the most common nonenhancing CPA mass (8). Epidermoid cysts arise from accidental inclusion of ectodermal epithelial tissue during neural tube closure in the first weeks of embryogenesis (3).

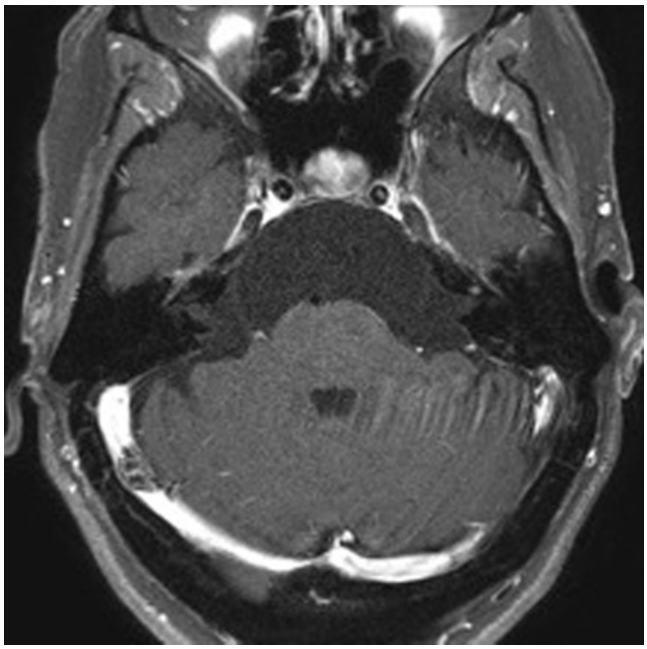

Figure 4. 49-year-old woman with sensorineural hearing loss. Axial T1 fat-suppressed postcontrast image demonstrates no enhancement of the CSF-signal-intensity mass in the CPA.

Due to a composition different from arachnoid cysts, several advanced imaging features can provide reliable differentiation of these entities.

On fluid-attenuated inversion recovery (FLAIR) sequences, arachnoid cysts are isointense to CSF. Epidermoid cysts alternatively demonstrate iso- to hyperintense signal with poor demarcation on FLAIR sequences (9). However, similar FLAIR-signal characteristics may be commonly seen in the posterior fossa cisterns due to CSF-flow artifact. Diffusion imaging is even more specific for discrimination of these lesions. Epidermoid cysts show restricted diffusion relative to CSF, whereas arachnoid cysts do not (10). Differential diagnosis for high signal on diffusion imaging of CPA masses includes epidermoid cysts and most of the solid tumors, except for hemangioendotheliomas, which show low signal on diffusion and high ADC signal (11). Thus, diffusion imaging is mainly helpful to differentiate between epidermoid cysts and arachnoid cysts.

Another available imaging tool for the CPA region is MR cisternography. This technique provides more precise anatomical depiction of lesion margins and relationship to other CPA structures for surgical planning compared to the low resolution of diffusion imaging. On MR cisternography, which is a heavily T2-weighted 3D sequence, epidermoid cysts show hypointensity to CSF, since (unlike arachnoid cysts) they are of mixed composition (12).

In our patient, diffusion-weighted imaging was key to diagnosing the lesion as an arachnoid cyst. Although conservative management was chosen in our case, it is suggested that symptomatic arachnoid cysts be surgically removed (13). However, vestibular symptoms are more likely to resolve, while auditory abnormalities may persist (14). Thus, our patient's sensorineural hearing loss likely would not have benefited from surgical intervention. Ultimately,

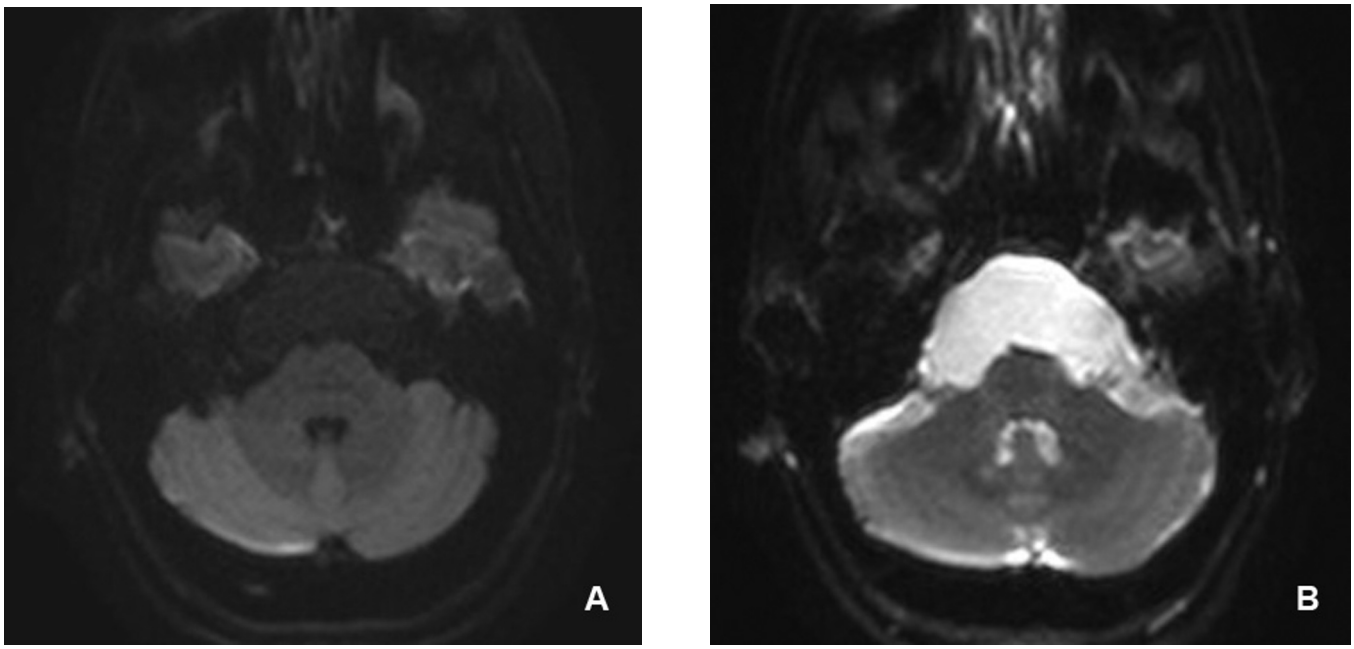

Figure 5: 49-year-old woman with sensorineural hearing loss. A. Diffusion-weighted image demonstrates low signal intensity and (B) ADC image demonstrates high ADC values within the cisternal mass matching those of CSF, confirming no restricted diffusion. Findings rule out an epidermoid cyst and confirm the diagnosis of arachnoid cyst.

arachnoid cysts can grow and encroach on adjacent structures, and initially they may be difficult to distinguish from other CPA masses. Use of diffusion-weighted imaging, MR cisternography, and gadolinium can provide a more precise diagnosis for patient management.

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