

## Posterior fossa arachnoid cyst presenting with isolated sensorineural hearing loss

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### Abstract

Arachnoid cysts are benign cysts occurring in the intra-arachnoid space and containing cerebrospinal fluid. They constitute approximately 1 per cent of all intracranial masses. They are uncommon in the posterior cranial fossa. Common presenting symptoms include headaches, seizures, focal neurologic signs and vague dizziness. Magnetic resonance imaging is the preferred method of investigation, and the treatment for symptomatic cysts is generally surgical drainage.

We report the unusual presentation of a young patient with a posterior fossa arachnoid cyst that manifested in the form of isolated unilateral sensorineural hearing loss. The patient underwent posterior fossa craniotomy and marsupialization of the cyst. To our knowledge, posterior fossa arachnoid cyst presenting with isolated hearing loss alone has not been reported in the English literature. A review of the literature pertaining to posterior fossa arachnoid cysts, including the clinical features, diagnosis and management, is also presented.

**Key words:** Arachnoid Cysts; Cranial Fossa, Posterior; Hearing Loss, Unilateral

### Introduction

Arachnoid (leptomeningeal) cysts are benign, space-occupying lesions occurring within the split layers of the arachnoid and are filled with cerebrospinal fluid (CSF).<sup>1,2</sup> The exact aetiology is unproven but they are believed to be congenital in most cases. They frequently communicate with the subarachnoid space.<sup>3</sup> Only 5 to 10 per cent of the cysts have been found to occur in the posterior cranial fossa.<sup>1</sup> There is no age predilection and the male to female ratio is 3:1.<sup>1</sup> The most common presenting symptoms of posterior fossa arachnoid cysts include headache, seizures and focal neurologic signs.

We report the case of a 29-year-old woman presenting with unilateral sensorineural hearing loss caused by a posterior fossa arachnoid cyst. To our knowledge, arachnoid cyst causing isolated hearing loss has not been reported before in the English literature. The clinical features, diagnosis and management of posterior fossa arachnoid cysts are discussed and the relevant literature is reviewed.

### Case report

A 29-year-old woman, who had been in good health previously, presented with a 12 months' history of right-sided hearing loss, otalgia and a feeling of ear blockage. There was no past history of any ear problems, tinnitus or vertigo.

The otolaryngological examination was normal, as were neurological and vestibular examinations. The patient underwent audiometric evaluation, which revealed a 50 dB sensorineural hearing loss in the right ear at 250 and 500 Hz, with an upward slope configuration (Figure 1).

In view of the unilateral sensorineural hearing loss, magnetic resonance imaging (MRI) of the internal acoustic meatuses was performed to rule out any cerebellopontine

angle pathology. Unexpectedly, this revealed a large arachnoid cyst in the right cerebellum, extending across the midline to the left, with diameters of 7 cm transversely and 5 cm anteroposteriorly. The brainstem was displaced anteriorly and the fourth ventricle was moderately compressed (Figures 2 and 3).

The patient was referred to the regional neurosurgical centre. She underwent posterior fossa craniotomy and marsupialization of the cyst communicating at the cervical subarachnoid space. Post-operatively, she made an excellent recovery without any neurological deficit. Histological examination of a portion of the excised cyst wall confirmed the diagnosis of an arachnoid cyst. Post-operative MRI scans performed one week after surgery confirmed a good decompression of the cyst. At three months post-operative follow up, the symptoms of earache and blockage had resolved. However, an audiogram showed the patient's hearing loss to be unchanged.

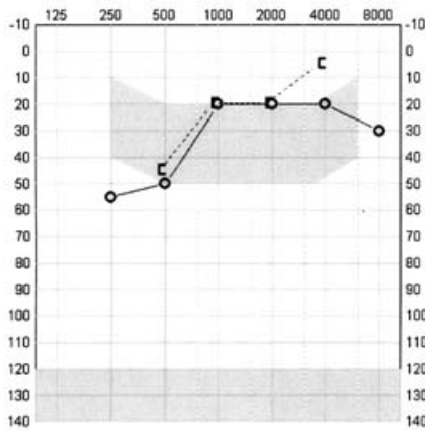
### Discussion

The precise aetiology of arachnoid cysts is poorly understood and remains controversial. The common consensus is that they are congenital in nature.<sup>4,5</sup> Minor aberrations of CSF flowing through the loose primitive perimedullary mesenchyme may result in focal splitting of the developing meninges. Formation of a pocket or diverticulum in the space thus created between the arachnoid and pia mater results in an arachnoid cyst. However, acquired aetiologies, such as inflammation and trauma,<sup>4,5</sup> have also been proposed. Arachnoid cysts range in size from small, incidental cysts to large, space-occupying lesions. Compression of the underlying brain tissue depends on the size and location of the arachnoid cyst.<sup>6</sup>

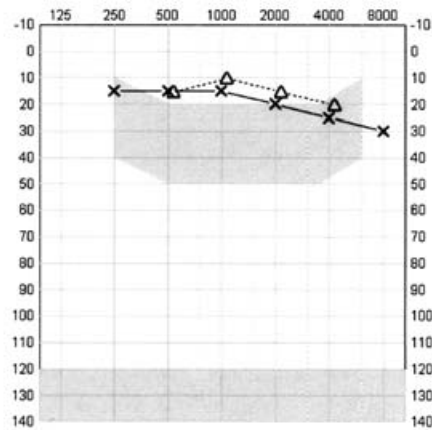
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AUDIOGRAM Clinic AuditData		Audiometer		Name							
Date		Doctor		Hospital no.: 5138061							
Diagnosis		Tone		Speech audiogram							
		Mean value		speech	Mask-Reflex	Max speech rec 1 & 2 (discrimination ability)					
Remark		BCL	HTL	tresholding	dB	%	dB	Mask.	%	dB	Mask.
		R	28	30							
		L	13	17							
Ear drum status		Comments									
<input type="checkbox"/> Normal <input checked="" type="checkbox"/> Note		tested 14/1/04									
Meas. result											
<input type="checkbox"/> Reliab. <input checked="" type="checkbox"/> Note											
Audiogram no		Signature									

Right



Left



Masking dB	125	250	500	750	1000	1500	2000	3000	4000	6000	8000	Ins. masked
HTL R:												
HTL L:												R <input type="checkbox"/>
BTL R:												L <input type="checkbox"/>
BTL L:												

**Audiogram notes:**

**Tympanometry results:**

	Canal Vol cm <sup>3</sup>	Compliance cm <sup>3</sup>	Pressure daPa
RIGHT EAR			
LEFT EAR			

**Tympanometry comments:**

FIG. 1

Pure tone audiogram showing 50 dB sensorineural hearing loss at 250 and 500 Hz in the right ear.

The middle cranial fossa is the most common site of location,<sup>7</sup> accounting for about 65 per cent of all arachnoid cysts. Up to 80 per cent of arachnoid cysts are symptomatic. Only 5 to 10 per cent of arachnoid cysts occur in the posterior fossa.<sup>1</sup> The cerebellopontine angles and cisterna magna are the most common infratentorial sites.

The hallmark of a posterior fossa arachnoid cyst is the variability in presenting signs and symptoms.<sup>4,7</sup> Posterior fossa arachnoid cysts are often unexpected, because they remain asymptomatic or can cause vague and non-specific symptoms such as headache, dizziness, hearing loss, tinnitus, lower cranial nerve palsies, facial numbness,

cerebellar and pyramidal signs, psychomotor retardation, seizures, and symptoms compatible with multiple sclerosis. No single symptom or group of symptoms define posterior fossa arachnoid cysts. These lesions should be considered as a potential diagnosis when the symptoms are long-standing, vague, fleeting or difficult to explain.

Ottaviani *et al.*<sup>8</sup> reported the case of a 21-year-old man with symptoms of left-sided tinnitus, followed by a mild, homolateral hypoacusia. An MRI revealed a large arachnoid cyst in the left cerebellar convexity which pushed the cerebellar hemisphere forward and caused homolateral cerebellopontine angle compression. Cartwright *et al.*<sup>9</sup>



FIG. 2

Axial posterior fossa magnetic resonance imaging scan; T2-weighted image showing a large arachnoid cyst in the cisterna magna, with displacement of the brainstem and fourth ventricle.

reported the unusual case, in a 74-year-old woman, of a posterior fossa arachnoid cyst presenting with an isolated, ipsilateral XIIth nerve paresis. Haberkamp *et al.*<sup>4</sup> presented three cases of posterior fossa arachnoid cysts, highlighting the extreme variability of the presenting symptoms, with a newly reported symptom – bilateral cochlear fluctuant hearing loss. O'Reilly and Hallinan<sup>7</sup>

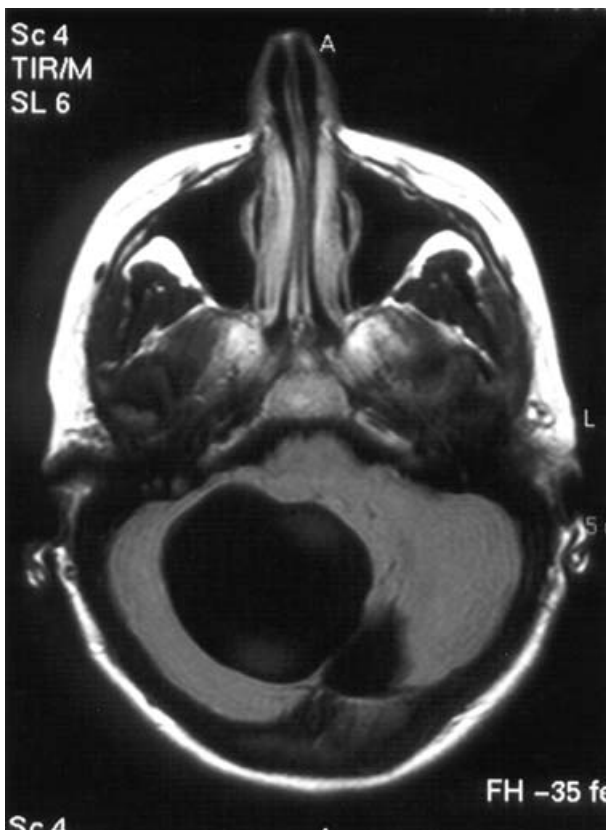


FIG. 3

Posterior fossa magnetic resonance scan; post-contrast image showing the large arachnoid cyst in the cisterna magna.

reported two cases of posterior fossa arachnoid cysts mimicking Ménière's disease, as well as the substantial resolution of symptoms in one patient after cysto-peritoneal shunt. Sumner *et al.*<sup>10</sup> described a case of facial paralysis in a three-year-old child, associated with hearing loss and enlargement of the internal auditory canal.

Magnetic resonance imaging is the preferred diagnostic procedure because of its ability to demonstrate the exact location, extent and relationship of the arachnoid cyst to the spinal cord. The differential diagnosis includes meningioma, epidermoid tumour, infarct, cystic tumour, loculated chronic subdural hygroma and metastasis.<sup>7</sup>

The treatment of arachnoid cysts is controversial.<sup>11</sup> The main objective of treatment is to relieve the symptoms. Periodic surveillance is recommended in asymptomatic patients. Symptomatic cysts are treated by various surgical procedures, such as drainage, total or partial removal of the cyst, and shunting or fistulization of the cyst to the subarachnoid space.<sup>4</sup> The posterior fossa arachnoid cyst can also be treated by endoscopic decompression.<sup>8</sup>

In patients presenting with unilateral hearing loss, cerebellopontine angle pathology is usually considered in the differential diagnosis; the most common lesions are acoustic neuroma, meningioma and cholesteatoma. However, in our patient, the underlying pathology turned out to be an arachnoid cyst in the right cerebellar hemisphere, displacing the brainstem anteriorly. The cyst was large in size, but there were no cerebellar symptoms or signs.

Although a cause and effect cannot be claimed with absolute certainty, we feel that the arachnoid cyst was probably responsible for the patient's hearing loss. A possible mechanism would be compression of the cerebellopontine angle due to the space-occupying lesion in the cerebellum. The exact pathophysiological mechanism for hearing loss in patients with arachnoid cysts is not understood. Vascular compromise has been proposed by some authors.<sup>7,8</sup>

Could the clinical picture in our patient be a manifestation of atypical Ménière's disease, with the arachnoid cyst as an incidental radiological finding? This is unlikely, as our patient had right-sided otalgia at the onset of the hearing loss, and this otalgia resolved after the surgical treatment. So far, 14 cases of posterior fossa arachnoid cysts mimicking Ménière's disease have been reported, and all these patients had a simultaneous onset of vertigo, imbalance, hearing loss and tinnitus. Surgical intervention improves vestibular symptoms and headaches but hearing loss is least likely to improve.<sup>7</sup> Samii *et al.*<sup>2</sup> reported that, in six of eight patients, vertigo or dizziness improved post-operatively. However, tinnitus and hearing loss were unchanged or worse in most of the patients (four out of five and five out of seven, respectively). Hearing loss due to arachnoid cyst is irreversible in most patients.

- Arachnoid cysts are benign cysts occurring in the intra-arachnoid space and containing cerebrospinal fluid. They constitute approximately 1 per cent of all intracranial masses
- Magnetic resonance imaging is the preferred method of investigation, and the treatment for symptomatic cysts is generally surgical drainage
- This paper describes a patient with a posterior fossa arachnoid cyst that manifested as isolated unilateral sensorineural hearing loss. A review of the literature pertaining to posterior fossa arachnoid cysts, including clinical features, diagnosis and management, is presented

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