

CASE REPORT

Arachnoid Cyst Presenting With Sudden Hearing Loss

Hsuan-Ho Chen, Chin-Kuo Chen*

Department of Otolaryngology, Chang Gung Memorial Hospital, Taoyuan, Taiwan, R.O.C.

Arachnoid cysts can occur at different intracranial sites, including the cerebellopontine angle (CPA). They often occur in childhood, in the posterior fossa. They may present with symptoms such as dizziness, tinnitus and hearing loss, or they may be asymptomatic. Presentation with sudden deafness is very rare. We report the unusual presentation of a 67-year-old male with CPA arachnoid cyst and the complaint of sudden-onset deafness. In this case, the cystic lesion at the CPA was found by magnetic resonance imaging of the brain. Pathology after retromastoid suboccipital craniotomy confirmed an arachnoid cyst. The treatment of this patient is discussed and the possible causes of CPA arachnoid cyst are briefly reviewed. [*J Chin Med Assoc* 2010;73(6):338–340]

Key Words: arachnoid cyst, cerebellopontine angle, sudden sensorineural hearing loss

Introduction

Sudden sensorineural hearing loss (SSNHL) is a common clinical disease in otolaryngology.¹ In many cases, the cause of sudden hearing loss cannot be determined, but known causes include the following: viral infection of the inner ear, toxin, immunologic causes, abrupt disruption of blood flow to the cochlear, and cerebellopontine angle (CPA) lesion (e.g. acoustic neuroma, meningioma or arachnoid cyst).¹ Arachnoid cysts constitute 1% of all intracranial space-occupying lesions.² The posterior fossa, especially the CPA, represents the 2nd most common site of arachnoid cysts.^{3–5} Such cysts may present with symptoms such as dizziness, tinnitus and hearing loss, or they may be asymptomatic.^{6–8} Presentation with sudden deafness is very rare.⁷ Herein, we describe a 67-year-old male with arachnoid cyst who manifested with SSNHL, which is rarely reported in the literature.

Case Report

A 67-year-old man came to our clinic with the complaint of sudden hearing impairment in his left ear,

a severe swaying sensation, and tinnitus. There was neither headache nor consciousness disturbance. Physical and neurological examinations were normal. All hematological findings were within normal limits. Skull radiography (Stenver's view) revealed normal diameter of the internal auditory canal. The patient had a 1-year history of hypertension. Panendoscopy was normal. Pure tone audiometry showed sensorineural hearing loss with an average of 110 dB on the left side (total deafness) (Figure 1A). The auditory brain stem response demonstrated an absence of all waves on the left side. The patient was diagnosed with sudden deafness and subsequently treated with oral prednisolone (1 mg/kg) and intravenous low molecular dextran (MW4000, 10% 500 mL/Bot) per day during his 1-week hospitalization. His dizziness and tinnitus were relieved during hospitalization.

The audiogram did not improve until 1 month later, when it showed marked recovery, with an average of 38 dB improvement (average dB of 0.5 k, 1 k, 2 k, 4 k) (Figure 1B). However, the auditory brain stem response still showed an absence of waves on the left side. Under the suspicion of retrocochlear lesion involving the cranial nerves or their nuclei, magnetic resonance imaging (MRI) of the brain was performed.



*Correspondence to: Dr Chin-Kuo Chen, Department of Otolaryngology, Chang Gung Memorial Hospital and Chang Gung University, 5, Fusing Street, Gueishan, Taoyuan 333, Taiwan, R.O.C.
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Surprisingly, MRI showed a fluid-containing cyst (measuring $3.9 \times 1.2 \times 2.1$ cm) occupying the left CPA with displacement of the 7th and 8th cranial nerves (Figure 2). After retromastoid suboccipital craniotomy, pathology confirmed an arachnoid cyst.

The audiogram was not significantly changed after 1 year of follow-up. The patient, however, did feel much relief from his tinnitus and dizziness.

Discussion

Arachnoid cysts are developmental collections of cerebrospinal fluid contained within the arachnoid membrane and subarachnoid space.⁵ Most arachnoid cysts

become symptomatic in early childhood, and 60–90% of reported patients are children.^{2,9,10} There are 2 pathways to development: primary anomalies and secondary acquired lesions due to trauma, hemorrhage or infection.¹¹ In our patient, the symptoms occurred at a late age and without any history of head injury or brain infection. What precipitated the onset of his symptoms is not clear.

The most common site of development of these cysts is the middle cranial fossa, followed by the posterior cranial fossa (5–11%).^{3–5,8,10} The incidence of CPA arachnoid cysts has been estimated to account for 1% of all intracranial lesions.^{3–5}

The symptoms of CPA arachnoid cyst include ataxia, gait disturbance, headache, nausea, vomiting, character change, and memory disturbance. Some symptoms mimic transient ischemic attack, dysesthesias, dysphagia, nystagmus, and oscillopsia.^{2,9,11} Jallo summarized 33 cases of CPA arachnoid cysts that were reported either as individual case reports or as a subgroup in a series of infratentorial arachnoid cysts of the 33 patients; the most common symptom was ataxia (37%) followed by headache (33%), and the most common sign was dysfunction of the 8th cranial nerve (44%).⁹ The 8th cranial nerve dysfunction caused by arachnoid cysts results in the symptoms of hypoacusia, tinnitus, and vertigo.^{2,3,7,10}

SSNHL is defined as a sensorineural hearing loss greater than 30 dB over at least 3 continuous audiometric frequencies, which develops in a period of less than 3 days.¹ SSNHL may result from causes affecting the cochlea, the 8th cranial nerve, or more central auditory tracts.¹² Arachnoid cysts rarely manifest with SSNHL.¹² Alaani et al reported 5 cases of CPA arachnoid cyst and only 1 initially presented with SSNHL.³

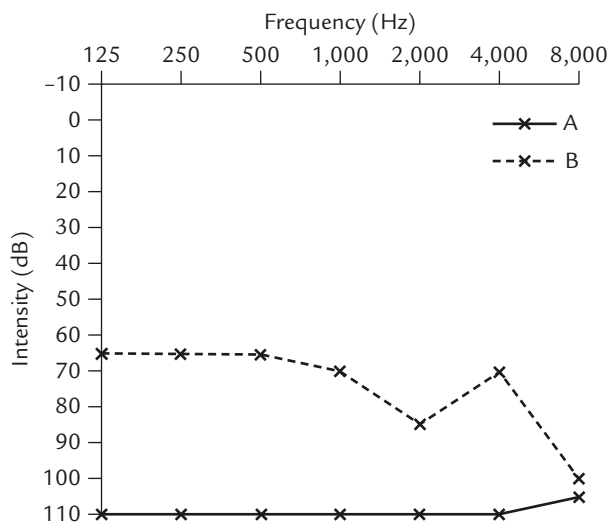


Figure 1. Audiogram shows total sensorineural hearing loss in the left ear (A) and marked recovery after medical treatment (B).

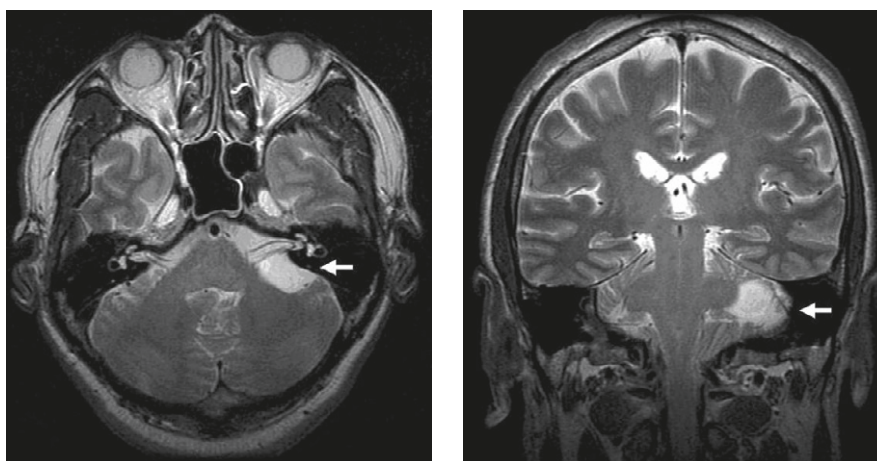


Figure 2. T2-weighted magnetic resonance imaging shows a fluid-containing cyst (white arrows), measuring $3.9 \times 1.2 \times 2.1$ cm, occupying the left cerebellopontine angle.

SSNHL evaluation should include a history and physical examination, contrast-enhanced MRI, and complete audiometry.^{1,8} MRI study of the audiovestibular nervous pathway and of the whole brain helps SSNHL evaluation because a previous study found that about 31 of 54 (57%) cases of SSNHL presented with MRI abnormalities.¹³

Corticosteroids are widely used because of their anti-inflammatory effect.¹⁴ They activate the glucocorticoid receptors, resulting in the suppression of proinflammatory molecules and reducing the quantity of inflammatory cells.¹⁴ Corticosteroids can be used in the treatment of SSNHL.¹ Our patient was treated with corticosteroids initially. Remarkable recovery (38 dB improvement) was demonstrated on the audiogram at the 1-month follow-up. A possible mechanism responsible for the effect may be a decrease in the degree of 8th cranial nerve edema caused by compression by the CPA arachnoid cyst.¹⁴ However, the extent of recovery depends on the degree and duration of the neural compression, and the medical treatment effect is thought to be only temporary.

Not all arachnoid cysts require surgical intervention.⁹ Patients with asymptomatic cysts should be followed-up clinically and radiologically with serial MRI.^{5,9} Indications for surgical intervention include any lesion that has demonstrated growth, hydrocephalus or refractory symptoms such as deafness, tinnitus and dizziness referable to a cyst in this location.^{8,9} The surgical choice may be craniotomy, such as drainage, total or partial removal of the cyst, shunting or fistulization of the cyst to the subarachnoid space.^{2,8,15} Minimally invasive endoscopic surgery has become the first-line therapy for arachnoid cysts because of the available endoscopic techniques.^{2,16} Operative management improves vestibular symptoms, but auditory deficits are less likely to respond to surgery.⁸

Our patient demonstrated that CPA arachnoid cysts may present with sudden onset of hearing impairment. The possibility of a CPA lesion should be considered in patients with SSNHL who are responsive to medical

treatment. Operative management is indicated if refractory symptoms are present.

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