CASE REPORT

Arachnoid Cyst of the Cerebellopontine Angle: A Case Report*

Hasan Yerli, Leyla Kansu, Celil Cabbarpur, Erdinc Aydin

From the Departments of Radiology (HY), and Otolaryngology-Head and Neck Surgery (LK, CC, EA), Baskent University, Ankara, Turkey

Arachnoid cysts of the cerebellopontine angle are rare. These benign cysts, which contain cerebrospinal fluid, develop in the intra-arachnoid space. Even the pathogenesis of those cysts is unknown; they are thought to be congenital. Symptoms are produced by the mass effect of the cyst on surrounding structures. The presenting symptoms are frequently nonspecific (headache, ataxia, gait disturbances, dizziness, generalized unsteadiness) or otologic such as sensorineural hearing loss, unilateral or bilateral hearing loss, vertigo, or tinnitus. The management of arachnoid cysts of the cerebellopontine angle is controversial. We report a 54-year-old man with an arachnoid cyst of the right cerebellopontine angle that led to progressive sensorineural hearing loss and tinnitus, and we discuss the therapy for and management of that disorder.

Submitted : 12 July 2008 Revised : 15 September 2008 Accepted : 25 March 2009

Lesions of the cerebellopontine angle account for 6% to 10% of all intracranial lesions. [1] Acoustic neurinomas and meningiomas represent approximately 85% to 90% of all tumors of the cerebellopontine angle, and the remaining lesions of this location are primary cholesteatomas and facial nerve schwannomas. Arachnoid cysts of the cerebellopontine angle are rare. [1,2]

Arachnoid cysts, which contain cerebrospinal fluid (CSF), are benign cystic lesions that develop in the intra-arachnoid space. Although the pathogenesis of those cysts is unknown, they are thought to be congenital; 60% to 90% of the reported patients with an arachnoid cyst are children. The other commonly accepted causes of arachnoid cysts are infection, trauma, splitting abnormalities of the arachnoid membrane, alteration of the CSF flow, and/or change of the CSF pressure. [3-5]

In this article, we describe a 54-year-old man with an arachnoid cyst of the right cerebellopontine angle that led to progressive sensorineural hearing loss and tinnitus, and we discuss the treatment policies of this disorder.

Corresponding address:

Leyla Kansu

Department of Otolaryngology-Head and Neck Surgery

Alanya Medical and Research Center Baskent University Alanya, Antalya, Turkey

 $Phone: +902425112522; Fax: +902425112350; \ E-mail: leylakansu@hotmail.com$

(*)This case report was presented for poster presentation in 8th International Conference on Cholesteatoma and Ear Surgery in 15-20 June 2008.

Copyright 2005 © The Mediterranean Society of Otology and Audiology

Case Report

A 54-year-old man with a hearing loss of approximately 8 years-duration and a 5-year history of tinnitus in his right ear was admitted to our hospital. Eight years earlier, he was referred to a private center seeking treatment for his hearing loss in right ear that was his only complaint at that moment. His audiometric evaluation revealed a 40-dB sensorineural hearing loss in his right ear. An internal acoustic canal magnetic resonance imaging (MRI) examination was recommended, but not performed. During subsequent 8 years, this patient's hearing level decreased and the severity of his tinnitus increased. He did not experience headache or vestibular symptoms during this period. The otolaryngologic, neurologic, and vestibular findings were within normal limits. There was no history of mumps infection and acute, chronic otitis media, or both. The audiometric evaluation showed a profound sensorineural hearing loss at high frequencies with a downward slope configuration in the right ear. The middle ear pressures were found to be normal bilaterally. Hearing thresholds were within normal limits in his left ear.

The results of magnetic resonance imaging (MRI) of the internal acoustic canal showed an extra-axial cystic lesion (15 x 15 x 8 mm) with a smooth surface. This lesion, which was localized in the anteromedial zone of the right cerebellar hemisphere in the posterocaudal region of the internal acoustic canal, caused an asymmetric enlargement in the cisterna of the cerebellopontine angle and demonstrated a CSF-like signal in all sequences (Figure 1). The content of the lesion was homogenous, and it showed no enhancement after gadolinium injection. The patient was was consulted with the the neurosurgery department but he refused the operation and agreed to to be followed-up with sequential MRIs.

Discussion

Usually, arachnoid cysts are asymptomatic. Symptoms from an arachnoid cyst are caused by an increase in the osmotic gradient of the liquid content of the cyst; the creation of a valve mechanism between the arachnoid cyst and the subarachnoid space, which leads to an increase in the size of the cyst; or the secretion of liquid from the cyst wall, which enlarges the cyst. The onset of the symptoms and signs are usually due to cortical irritation, compression of the cerebral parenchyma, or the obstruction of CSF circulation. [6] Symptoms are produced by the mass effect of the cyst on surrounding structures. When an arachnoid cyst is located in the cerebellopontine angle, the patient's presenting symptoms are frequently neurotologic, such as sensorineural hearing loss, vertigo, or tinnitus.

Those symptoms are due to the dysfunction of the eighth cranial nerve. Sometimes, nonspecific symptoms (headache, ataxia, gait disturbances, dizziness, generalized unsteadiness) develop. Compression of cranial nerves VII or V can cause facial palsy, hemifacial spasm and/or neuralgic pain. ^[2,4,7] Eslick and colleagues reported a patient with an arachnoid cyst of the cerebellopontine angle that caused diplopia via direct compression of cranial nerve VI. ^[6] Our patient exhibited only unilateral sensorineural hearing loss and tinnitus. Arachnoid cysts may also become symptomatic by compressing the brainstem. ^[8]

The differential diagnosis of an arachnoid cyst of the cerebellopontine angle includes other cystic lesions (epidermoid and neurenteric cyst, cystic acoustic schwannoma). MRI is helpful in differentiating arachnoid cysts from those cystic lesions. If a pathologic cause of a retrocochlear disorder is suspected in a patient with a unilateral sensorineural hearing loss and tinnitus, MRI should be performed to evaluate the cerebellopontine angle. arachnoid cysts appear as smooth-surfaced lesions that in all magnetic resonance sequences exhibit a signal characteristic of CSF. In contrast, epidermoid cysts show mixed signals on FLAIR images and high signals on diffusion weighted images. Neurenteric cysts present high signals on T1-weighted images and cystic schwannomas show some foci of contrast enhancement on T1-weighted postcontrast images. [1,2]



Figure 1. Axial T2-weighted (A) and coronal FSEIR (B) images show the signal of an arachnoid cyst (arrows), which is similar to the signal characteristic of cerebrospinal fluid. Post-contrast T1-weighted image (C) shows no enhancement of the lesion.

The management of arachnoid cysts of the cerebellopontine angle remains controversial. Asymptomatic arachnoid cysts do not require treatment, and such patients should be monitored clinically and radiologically with serial MRIs. If the patient demonstrates no significant compromise in local neural or vascular structures, no severe symptoms, and no suspected or proven rapid cyst growth, a watch-and-wait policy should be implemented. The risks of surgery are few, but complications (meningitis, hemiparesis, oculomotor palsy, subdural hematoma, grand mal epilepsy, and death) have been reported. [4-6] We discussed the risks and benefits of surgery with our patient, who preferred to undergo monitoring with MRI.

The surgical treatment of arachnoid cysts of the cerebellopontine angle consists of total resection and surgical drainage. To reduce the likelihood of complications, surgical drainage via the retrolabyrinthine or retrosigmoid exposure is the recommended therapy. Some authors recommend the placement of a cystoperitoneal shunt in patients with hydrocephalus. In recent years, endoscopic cyst decompression, has been shown to be safe and effective. [2,5,7]

In a review of the English literature, we found few reports in which arachnoid cysts of the cerebellopontine angle leading to sensorineural hearing loss. In these articles, the degree of hearing improvement following the surgery varied. [4,7,9]

Arachnoid cysts of the cerebellopontine angle are a rare clinical conditions that can cause various neurotologic symptoms. The benefits and risks of surgical treatment should be discussed with the patient. In most patients with an arachnoid cyst, a watch-and-wait policy can be safely applied.

References

- 1. Brackmann DE, Arriaga MA: Extra-axial neoplasms of the posterior fossa. In: Cummings CW, Fredrickson JM, Harker LA, Krause CJ, Richardson MA, Schuller DE (eds) Otolaryngology Head and Neck Surgery. 3rd ed. St. Louis, MO: Mosby-Year Book: 1998: 3294-3314.
- 2. Bonneville F, Sarrazin JL, Marsot-Dupuch K, Iffenecker C, Cordoliani YS, Doyon D, Bonneville JF: Unusual lesions of the cerebellopontine angle: a segmental approach. Radiographics 2001; 21: 419-43.
- 3. Jallo GI, Woo HH, Meshki C, Epstein FJ, Wisoff JH: Arachnoid cysts of the cerebellopontine angle: diagnosis and surgery. Neurosurgery 1997; 40: 31-38.
- 4. Alaani A, Hogg R, Siddiq MA, Chavda SV, Irving RM: Cerebellopontine angle arachnoid cysts in adult patients: what is the appropriate management? J Laryngol Otol 2005; 119: 337-341.
- 5. Ucar T, Akyuz M, Kazan S, Tuncer R: Bilateral cerebellopontine angle arachnoid cysts: case report. Neurosurgery 2000; 47: 966-968.
- 6. Eslick GD, Chalasani V, Seex K: Diplopia and headaches associated with cerebellopontine angle arachnoid cyst. ANZ J Surg 2002; 72: 915-917.
- 7. Ottaviani F, Neglia CB, Scotti A, Capaccio P: Arachnoid cyst of the cranial posterior fossa causing sensorineural hearing loss and tinnitus: a case report. Eur Arch Otorhinolaryngol 2002; 259: 306-308.
- 8. Greenberg MS: Primary brain tumors. Handbook of Neurosurgery. 6th edition. NewYork, Thieme 2006: 408-479.
- 9. Lanzino G, diPierro CG, Ruth RA, Helm G, Jane JA: Recovery of useful hearing after posterior fossa surgery: the role of otoacoustic emissions: case report. Neurosurgery 1997; 41: 469-473.