A Rare Cause of Fluctuant Hearing Loss: Cerebellar Peduncle Cavernoma

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Summary

Background: Cavernomas are an unusual type of intraparenchymal vascular malformations which are generally located in the cerebral hemispheres, brainstem and midbrain, but they occur rarely within cerebellum.

Case Report: A 41-year-old man with cavernoma of the middle cerebellar peduncle and complaints of tinnitus, hearing loss and vertigo was presented. Treatment was initiated with concurrent transtympanic and systemic corticosteroids. It was learned that, two years ago he had suffered from tinnitus, hearing loss and had been diagnosed as idiopathic high frequency hearing loss. Magnetic resonance imaging (MRI) revealed middle cerebellar peduncle localization of a 15x13 mm cavernoma with subacute hemorrhage. Auditory brainstem response showed lengthened in I-III and III-V interpeak latencies. The patient was managed conservatively. At the end of the third month hearing was improved significantly.

Conclusions: Consideration of clinical, audiological and imaging findings together is important for accurate diagnosis in sensorineural hearing loss.

Key words: Cavernoma, fluctuant hearing loss, cerebellar peduncle

Fluktuan İşitme Kaybının Nadir Bir Nedeni: Serebellar Pedinküler Kavernom

Özet

Giriş: Kavernomlar, genellikle serebral hemisferler, beyin sapı ve ortabeyine lokalize olan, ancak ender olarak serebellumda görülen, nadir rastlanan intraparanjimal vasküler malformasyonlardır.

Olgu: Kulakta çığlama, işitme kaybı ve vertigo şikayetleriyle başvuran ve orta serebellar pedinkülde kavernom saptanan 41 yaşında erkek hasta sunuldu. İki yıl önce işitme kaybı ve çığlama şikayetleri olan hastaya yüksek frekanslı işitme kaybı tansısı konulduğu ögrenildi. Tedavi eşi zamanlı transtimpanik ve sistemik kortikosteroid ile başlatıldı. Magnetik rezonans görüntülemede orta serebellar pedinkülde lokalize 15x13mm subakut hemorajı içeren kavernom görüldü. İşitme belirgin olarak düzeldi. Üçüncü ayın sonunda işitme belirgin olarak düzelendi.

Sonuç: Sensorinöral işitme kaybında klinik, odyolojik ve görüntüleme bulgularının birlikte değerlendirilmesi doğru tanı için önem taşır.

Anahtar Kelimeler: Kavernom, fluktuan işitme kaybı, serebellar pedinkül
INTRODUCTION

Cavernomas are the second most common intracranial vascular malformation after arteriovenous malformation and represent 15% of all cerebral vascular malformations(9). Intraaxial lesions locate in brainstem, cerebellum, and cerebral hemisphere. Histologically, they are composed of endothelial-lined sinusoidal vascular spaces without intervening neural tissue, containing essentially thrombosed blood. The etiopathogenesis of cavernomas is thought to be congenital. Male and female patients are affected equally. The peak incidence of symptomatic presentation is between the third and fifth decades. The estimated spontaneous haemorrhage rate for brainstem cavernous angiomas is 5% per lesion per year or less(12).

The incidence of the sudden sensorineural hearing loss (SNHL) has been reported to range from 5 to 20 per 100000 subjects per year, but precise incidence is estimated to be higher(11). Its cause being known in only 10-15% of the cases (13). Intrinsic brain tumor is an extremely rare cause of sudden hearing loss and may be easily overlooked, if not evaluated with MRI.

MRI is the most sensitive and specific imaging modality for the diagnosis and localization of cavernomas(1). They show characteristic mixture of low and high signal intensity in T2-weighted MRI due to repeated hemorrhage. Angiography is also useful for diagnosing cavernomas because it reveals the flecked lesion stain at middle arterial to late venous phase owing to slow blood flow, delayed stain, or an avascular mass(7).

In the literature, hearing loss due to internal auditory canal and extraaxial cerebellopontine angle cavernomas were reported. We describe a case of cavernoma located in the middle cerebellar peduncle manifesting as right fluctuant SNHL. To our knowledge fluctuant hearing loss due to peduncle cavernoma has not been reported before.

CASE PRESENTATION

A 41-year-old male patient referred from the emergency service with complaints of hearing loss in the right ear, vertigo and tinnitus. Neuro-otological examination revealed no abnormality. Right total SNHL was found in pure tone audiometry. Speech discrimination score was 68% with monosyllables and the Weber test was lateralized to the left side. His medical history revealed temporary hearing loss in his right ear at high frequencies with accompanied tinnitus two years ago. He was interned with the prediagnosis of idiopathic sudden hearing loss and temporal MRI was planned.

He received intravenous 250 mg prednisolone for 3 days and transtympanic dexamethasone was administered for five days. The dizziness gradually reduced, but hearing in the right ear did not improve.

MRI revealed approximately a 13x14 mm lesion compatible with acute-subacute haemorrhagic cavernoma on right middle cerebellar peduncle. T2 weighted images showed central hyperintensity and, peripheral hypointensity (Figure 1). T1 weighted and flair images demonstrated hyperintense lesion confined with hypointensity due to hemosiderin ring reflecting chronic blood degradation products (Figure 2). There was no extra contrast enhancement on T1 weighted images. MR angiography was normal. Neurosurgery consultants decided to follow-up with MRI, surgery was not planned.

After 10 days, right ear air conduction thresholds improved from 100 dB to 85 dB. Auditory brainstem response (ABR) was present at 60 dB one month after the onset of initial symptoms. At the end of the third month hearing improved considerably, 45 dB.
DISCUSSION

Cavernomas are angiographically occult vascular malformations of the central nervous system consisting of irregular sinusoidal vascular channels without intervening neural parenchyma\(^2\). The most common presenting features of intracranial cavernomas are headache, epilepsy and focal neurological deficits such as imbalance, hemiparesis, speech disorder, impairment of vision and hearing.
loss\(^5\). Especially after the routine use of MRI, in the number of cases reported as cavernoma were increased\(^8\). Symptoms referable to brainstem cavernous angiomas include progressive long tract and cranial nerve deficits occurring over the course of days or weeks. This is in distinction to supratentorial angiomas, which are more frequently associated with seizures. A relapsing and remitting course is not uncommon with recurrent bleeding as indicated in our case. Brainstem cavernomas within the pons lie in close proximity to the vestibulocochlear nucleus complex and may therefore result in hearing loss and dizziness\(^12\).

The most common etiology of fluctuant SNHL is Meniere’s disease, but may be associated with autoimmune inner ear disease or even with vestibular schwannoma\(^4\). Although patients with cerebellopontine angle tumors may present with steroid-responsive fluctuant hearing loss, hearing deterioration is expected in a short period. Fluctuating and worsening hearing levels over a prolonged period of time is compatible with vascular pathologies rather than solid tumors as in the presented case\(^3,6\). Many cases of SNHL remain “idiopathic” with no proven etiology. However, clinicians must always be aware that idiopathic sudden SNHL still remains an exclusion diagnosis after the multiple possible cases are eliminated\(^11\).

For patients with sudden hearing loss, the use of immediate MRI can be useful for establishing an appropriate differential diagnosis and to help guide proper management. Although several previous studies recommended evaluating patients with sudden SNHL loss by MRI to rule out underlying organic disease, few clinicians seem to be aware that even a small intrinsic lesion has the potential to produce sudden SNHL\(^11\).

Natural history for peduncle cavernomas and surgical morbidity are the defining factors in the decision to conservatively manage or to excise them\(^10\). There is no specific medical treatment for peduncle cavernoma. Anticoagulant therapy, platelet-dispersing medication and violent sports activities are contraindicated\(^3\).

A central lesion should be considered in fluctuant SNHL and MR imaging should be done before any treatment. As in this reported case, clinical findings should be correlated with radiological findings in order to exclude any central lesion before diagnosing the patient as “idiopathic” SNHL. In addition, recurrent hearing loss may be a clue for microhemorrhages due to peduncular cavernomas. Consideration of clinical and imaging findings together is important for accurate diagnosis.

**CONCLUSION**

Brainstem cavernous angiomas within the pons and especially in middle cerebellar peduncle lie adjacent to the vestibulocochlear nucleus complex and nervus vestibulocochlearis, hence may cause tinnitus and hearing loss. The otolaryngologist should be aware of this less common clinical entity and entertain this differential diagnosis in the evaluation of patients with such symptoms.

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**Received by:** 24 October 2014
**Revised by:** 14 December 2014
**Accepted:** 02 March 2015

**The Online Journal of Neurological Sciences (Turkish) 1984-2015**
This e-journal is run by Ege University Faculty of Medicine, Dept. of Neurological Surgery, Bornova, Izmir-35100TR as part of the Ege Neurological Surgery
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