Case Report

**Isolated Sensorineural Hearing Loss due to Enlargement of Pontine Neuroglial Cyst**

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**Summary**

Isolated sensorineural hearing loss due to posterior fossa benign cystic lesions is extremely rare. Although they are benign in the natural course, rarely they may cause progressive neurological symptoms. Radiologic and audiometric examinations are useful methods for initial diagnosis and follow-up evaluations.

A 48 year old man suffered from left-sided progressive hearing loss for six months. Cranial magnetic resonance imaging showed that intrapontine cystic mass measuring 9 mm, enlarged 3 mm during the seven years period. In addition, auditory brainstem response revealed that prolonged 1-5 interpeak latency on the right side and wave 5th was absent on the left.

In this report, we described a patient, who has progressive unilateral neural sensorineural hearing loss without any neurological symptoms, because of the enlarging intrapontine cystic mass, which compress to the lower auditory pathways. Although, isolated sensorineural hearing loss has been reported in intraaxial cystic lesions in consequence of cerebellopontine angle compression, but we do not encountered in the English literature that isolated sensorineural hearing loss due to compression of lower auditory pathways by intrapontine cystic mass.

**Key words:** Auditory brainstem response, brainstem auditory evoked potential, intracranial auditory pathway, posterior fossa cyst, sensorineural hearing loss

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**Özet**

İyi huylu posterior fossa kistik lezyonlarına bağlı izole sensörinöral işitme kaybı oldukça nadirdir. Doğal seyirlerinde iyi huylu olmalarına rağmen, onlar nadiren ilerleyici nörolojik şikayetlere neden olabilirler. Radyolojik ve odyometrik incelemeler başlangıç tanı ve takip değerlendirme için kullanılabilecek yöntemlerdir. 48 yaşında erkek hasta 6 aydır solتراflı işleyici işitme kaybından yakınmaktaydı. Kraniyel manyetik rezonans görüntüleri 7 yıllık süreçte 3 mm genisleme gösteren 9 mm boyutunda pons içerisindeki kistik kitleyi gösterdi. Ek olarak, işitsel beyinsapı cevabı sağтарafa uzamsız 1-5 interpeak latansı ve solda 5. dalgânın kaybını ortaya koydu. Bu raporda, bir başka nörolojik yakınlıklar olmadan alt işitsel yolları baskılayan pons içerisindeki kistik kitlenin gelişmesine bağlı ilerleyici tek tarafı nöral sensörinöral işitme kaybı bulunan hastaı tarifieldik. Her ne kadar, serebellopontin köşe başına bağlı izole sensörinöral işitme kaybı intraaksiyel kistik lezyonlarda bildirilmişse de, biz inglizce literatürde intrapontin kistik kitle tarafından alt işitsel yolların basısı nedeniyle oluşan izole sensörinöral işitme kaybına rastlamadık.

**Anahtar Kelimeler:** İşitsel beyinsapı cevabı, beyinsapı işitsel uyarılmış potansiyeli, intrakraniyal işitme yolu, posterior fossa kisti, sensörinöral işitme kaybı
INTRODUCTION

Neural (retrocochlear) sensorineural hearing loss (SNHL) may be caused by lesions of internal auditory canal or cerebellopontine angle (CPA) or lesions of the intraaxial auditory pathway. In patients presenting with unilateral hearing loss, the most common CPA pathologies are acoustic neuroma, meningioma and cholesteatoma. In these pathologies, a possible mechanism of SNHL would be compression of the CPA due to the space occupying lesion in the posterior fossa, but a cause and effect cannot be claimed with absolute certainty. Besides these common CPA pathologies, isolated sensorineural hearing loss as the initial symptom of the disease had been rarely reported in isolated eight cranial nerve or CPA metastasis in consequence of cranial nerve dysfunction or CPA compression. Any intraaxial lesions that disrupt the normal function of the ascending fibers with the central acoustic pathway may also rarely cause SNHL, as in our case. These are ischemic, traumatic, demyelinating or neoplastic origin. In these cases, a possible mechanism of hearing loss would be a focal injury in the brainstem.

In addition to well-known pathologies, various kinds of cysts can occur in the posterior cranial fossa, but neuroglial cysts are rare lesions. Also, neuroglial cysts typically present brainstem dysfunction or signs and symptoms of raised intracranial pressure due to hydrocephalus. Especially, brainstem neuroglial cyst presenting with isolated hearing loss alone has not been reported in the English medical literature.

CASE PRESENTATION

A 48 year old man suffered from increasing left sided hearing loss for six months and was readmitted ENT clinic after 7 years. He had no history of trauma, otorrhea, pain, associated vertigo, exposure to noise or ototoxic medications, previous ear disease or surgery, and no family history of hearing loss.

The first admission was in January 2006 and the patient's otologic examination was normal and pure tone audiogram showed left-sided mild sensorineural hearing loss (Figure 1a). Magnetic Resonance Imaging (MRI) of brain revealed a well-circumscribed cystic lesion at the pontomedullary junction and centered in the pons (Figure 2a). It was a purely intraaxial localization, measured 6mm in its largest diameter, with no contrast enhancement on T1 weighted and only minimal mass effect on T2 weighted MRI (Figure 2c-d). Since the patient had no additional neurological symptoms, he preferred to a wait-and-see period. He had no progression of his complaints until second admission.

On second admission in August 2013, otologic examination was normal. Pure tone audiogram showed left-sided moderate sensorineural hearing loss and speech audiometry revealed moderately discrimination loss (Figure 1b). The appearance of the lesion on CT was consistently that of a low density lesion that did not enhance following administration of intravenous contrast medium had no surrounding edema. This time, repeated head MRI showed only a previous cystic mass, which had the same MR signal characteristics, but had 3 mm enlargement of its size (Figure 2a1- d1). For differential diagnosis, we needed additional MR sequences, laboratory and neurophysiological tests. The lesion was isointense with cerebrospinal fluid(CSF) on T1 weighted, T2 reversed and diffusion-weighted images. Also, early stage of neurocysticercosis was thought, but anticysticercosis antibodies were not detected in blood and CSF. Moreover, auditory brainstem response (ABR) was performed and revealed that wave 5th was absent on Figure 3a-left and prolonged 1-5
interpeak latency (IPL) on Figure 3b-right. The recent ABR findings suggested that the cyst affected the lower auditory pathways in brainstem. In contrast, we could not compare previous findings, since ABR test was not available during 2006 at our hospital. Also, patient had no physical or radiological evidence of any other embryological maldevelopment, and evidence of cerebral hemorrhage, infarction, infection, neoplasm. As a result of these findings, a presumptive diagnosis of brainstem neuroglial cyst was then made.

Thus, we offered stereotactic biopsy to the patient, but he refused surgery and preferred a wait-and-see period again.

**Figure 1:** Pure tone audiograms of the patient are demonstrated. Left sided mild sensorineural hearing loss (1a), and moderate hearing loss after 7 years (1b).

**Figure 2:** Left (a-d) and right (a1-d1 ) semicolon show initial and last MRIs, respectively. Sagittal (a,a1) and axial with gadolinium (b,b1) T1 images show an intrinsic cystic lesion arising at the pontomedullary junction (PMJ). The cyst appears isointense to CSF on T1 and T2 images (c,c1,d,d1).
DISCUSSION

Causes of neural SNHL, according to anatomic location, can be classified into two main groups: intraaxial and extraaxial (internal auditory canal or CPA) auditory pathway. In the former, most common lesions are ischemic, traumatic, demyelinating or neoplastic origin\(^{(13,19)}\). Additionally, various kinds of cysts can occur in the posterior fossa. In origin, they can be epithelial, such as endodermal (neuroenteric) or neuroepithelial (ependymal and choroidal epithelial); mesenchymal, such as arachnoid cyst; or mixed, such as teratomatous cysts\(^{(2,10)}\).

In rare instance, neuroglial cysts occur in a wide variety of sites throughout the neuroaxis. In the posterior fossa, they may occur in the CPA region, within the cavity of the fourth ventricle, or entirely within the parenchyma of the brainstem or cerebellar hemisphere. Also, these lesions typically present brainstem dysfunction or signs\(^{(18)}\).

Clinically, an isolated syndrome indicating a pathological process involving the cranial nerves or the posterior fossa may constrict the clinician due to a difficult diagnostic challenge\(^{(23)}\). While diplopia, vertigo and ataxia are often initial symptoms of pathological process in posterior fossa\(^{(6,9)}\), facial paralysis, spasm, neuralgia or hearing loss are unusual in clinical practice\(^{(2,6,10,18)}\).

In patients presenting with unilateral hearing loss, CPA pathology is usually considered in differential diagnosis; the most common lesions are acoustic neuroma, meningioma and cholesteatoma\(^{(7,20)}\). In few patients, authors have reported cases of arachnoid cyst in posterior fossa whose first clinical symptom was a neural hearing loss without any other symptoms typical of a cerebellar and CPA cistern localization\(^{(15)}\). The exact pathophysiological mechanism for hearing loss in these patients is not understood. In general, a possible mechanism would be compression of the CPA due to the space occupying lesion in the posterior fossa. However, vascular compromise has been proposed by some authors. In these few cases, the absence of improvement in otoneurological parameters after decompressive surgery that the long-lasting compression of the CPA led to irreversible damage\(^{(15,20)}\), whereas sometimes, the same compression may cause reversible damage. This situation is explained by compression neuropathy due to cyst, since hearing returned to normal responses\(^{(6)}\).

On the other hand, any intraaxial pathological process disrupts the normal
function of the ascending fibers within the central acoustic pathway. This process may result in a focal injury in the brainstem and cause SNHL\(^{(12)}\). In general, this injury involves one or more cranial nerve nuclei plus the corticospinal and/or the spinothalamic pathways in variable patterns\(^{(14,16,17,23)}\). In particular, intraaxial lesions proximal to the cochlear nuclei on the dorsolateral medulla generally present with bilateral SNHL, worse on the side opposite the lesions\(^{(12)}\), whereas isolated hearing loss alone has not been reported in the English medical literature. In our opinion, compression lower auditory pathways due to an enlarging intraaxial pontine neuroglial cyst, as in our case, has never been described.

To the best of our knowledge, excluding extraaxial pathologies, description of intraaxial lesion for hearing loss is not easy to find. In contrary, any intraaxial process affected lower auditory pathways can be explained with either radiological or neurophysiological evidences. Anatomically, the lower auditory pathways are situated in brainstem: superior olivary complex (SOC) and cochlear nuclear complex (CNC). The former is a set of nuclei located in brainstem, near the trapezoid body, and between the lateral lemniscus nuclei anteriorly and the facial nucleus posteriorly. The latter is located peripherally in the upper medulla-oblongata adjacent to the inferior cerebellar peduncle\(^{(3)}\). Firstly, the surface and sectional landmarks of CNC have been defined anatomically on MR imaging. As a result of these findings, MR-detectable focal CNC abnormalities causing the SNHL can be shown\(^{(3,8,9,13)}\). Secondly, ABR are routinely used in clinical practice to evaluate the normality of lower auditory system. ABR are considered abnormal when I to V interpeak latency is longer than 4.4 ms and the interaural latency difference for V is longer than 0.3 ms. Also, involvement of brainstem auditory structures by demyelinating diseases produce loss of synchronous cells activity.

For example, V waveform can be absent in multiple sclerosis\(^{(4,5,22)}\). In our opinion, based on radiologic and ABR evidence, we can speculate a compression of the lower auditory pathways, because of increasing mass effect of the enlarging neuroglial cyst in the brainstem. A possible mechanism of SNHL in our case can be explained that cyst distended with fluid may cause directly stretching or pressure effect on lower auditory pathways between SOC and CNC\(^{(21)}\).

Consequently, intraaxial neurogial cyst in brainstem are benign, but are not always stable lesions. Rarely, they may cause progressive neurological symptoms, such as unilateral neural SNHL.

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REFERENCES


6. Chu CK, Tseng HM, Young YH. Clinical presentation of posterior fossa epidermoid cysts Eur Arch Otorhinolaryngol 2006; 263: 548-51


20. Thinakara-Rajan T, Janjua A, Srinivasan V. Posterior fossa arachnoid cyst presenting with isolated sensorineural hearing loss J Laryngol Otol 2006; 120: 979-82

