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Case Report

Overdrainage by Lumboperitoneal Shunting May Cause Intracranial Hypotension Resulting in Permanent Deafness

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Abstract

Intracranial hypotension (IH) can develop spontaneously or following a spinal intervention such as lumbar puncture, surgery and shunt procedure or a trauma. IH has a wide range of clinical presentation. Although orthostatic headache is the most frequent component of IH symptomatology, it might not be seen in some patients. We present a 27-year-old woman with nausea, vomiting and bilateral severe hearing loss following a lumboperitoneal shunt procedure after she was diagnosed with idiopathic intracranial hypertension. Her audiogram showed bilateral severe sensorineural hearing loss and magnetic resonance imaging showed displacement of cerebellar tonsils 10 mm inferior of foramen magnum, diffuse dural enhancement and pachymeningeal thickening. Cerebrospinal fluid (CSF) opening pressure was 0-1 cmH_2O. The patient was diagnosed with intracranial hypotension. After shunt removal surgery; her nausea and vomiting resolved completely while her hearing loss persisted. Permanent hearing loss is infrequent but one of the serious complications of CSF overdrainage after shunt procedures.

Keywords: Intracranial Hypotension, Permanent Deafness, Lumboperitoneal Shunting, Sensorineural Hearing Loss

Lumboperitoneal Şanttan Aşırı Drenaj Kalıcı İşitme Kaybı İle Sonuçlanan İntrakraniyal Hipotansiyona Sebep Olabilir

Özet

İntrakraniyal hipotansiyonun (İKH) en sık sebebi spontan İKH olmakla birlikte lomber ponksiyon, lomber cerrahi, şant operasyonu ya da travma sonrası da görülebilir. Çeşitli belirtilere sebep olabilir. Ortostatik baş ağrısı en sık görülen belirtisidir. Burada, 27 yaşında, bulantı, kusma ve bilateral işitme kaybı ile başvuran bir kadın hasta sunulmaktadır. Hastanın özgeçmişinde 2 yıl önce tıbbi tedaviye yanıt vermeyen idyopatik intrakraniyal hipertansiyon tanısı ile uygulanmış lumboperitoneal şant operasyonu vardı. Hastaya LP yapıldı, açılış basıncı 0-1 cmH_2O idi. Odyogramı bilateral ileri derecede sensorinöral işitme kaybı ile uyumlu idi. Kraniyal manyetik rezonans görüntülemesinde serebeller tonsillerin foramen magnumdan 10 mm inferiora doğru uzanım gösterdiği, ayrıca pakimeningeal kalınlaşma ve diffüz dural kontrastlanma olduğu saptandı. Hastaya İKH tanısı kondu. Lumboperitoneal şantı çıkarılan hastanın bulantı kusma şikayeti tamamen düzeldi, fakat işitme kaybında değişiklik olmadı. Hastamızda görüldüğü gibi lumboperitoneal şanttan aşırı drenaj sonucu İKH olabilir, ortostatik hipotansiyon olmaksızın işitme kaybı ve vestibüler semptomlar görülebilir. Bu

hastalarda erken tanı ile BOS basıncının düzeltilmesi işitme kaybının kalıcı olmasını önleyebilir.

Anahtar Kelimeler: İntrakraniyal Hipotansiyon, Kalıcı Sağırlık, Lumboperitoneal Şant, Sensörinöral İşitme Kaybı

INTRODUCTION

Intracranial hypotension (IH) is caused by a decrease in cerebrospinal fluid (CSF) pressure. It can be either spontaneous due to a possible connective tissue weakness or secondary to a CSF leakage caused by traumaand spinal interventions such as surgery or lumbar puncture[2,14]. Overdrainage due to shunting procedures is another underlying cause of IH[5,8,9]. The patients with IH usually present with orthostatic headache which worsen in erect position and ameliorated in supine position. Nausea, vomiting, neck and interscapular pain, blurred vision, dizziness, diplopia, cochleovestibular manifestations, and cranial nerve palsies are other manifestations of IH[6,12]. We here report a patient with a primary diagnosis of idiopathic intracranial hypertension who presented with severe and persistent hearing loss due to IH as a result of lumboperitoneal shunting.

CASE PRESENTATION

A 27-year-old woman was admitted with nausea, vomiting, and bilateral progressing hearing loss that started one year ago. In medical history, she was diagnosed with idiopathic intracranial hypertension at the medical center where she admitted with severe headache, diplopia and bilateral papiledema five years ago. She was unresponsive to the medical treatment, so she underwent lumboperitoneal (LP) shunt surgery two years ago. Brain magnetic resonance imaging (MRI) that was performed prior to surgery was unremarkable (Figure 1).

On admission, her physical examination was normal. The neurological examination revealed bilateral severe hearing loss. Neuro-ophthalmologic examination was unremarkable. There was no abnormality in routine blood investigations. Audiogram showed bilateral severe sensorineural hearing loss (Figure 2).

Brain magnetic resonance imaging (MRI) showed caudal displacement of cerebellar tonsils 10 mm inferior of foramen magnum. Diffuse dural enhancement and pachymeningeal thickening were also demonstrated in brain MRI (Figure 3). Lumbar drainage that was performed after cranial imaging revealed a cerebrospinal fluid (CSF) opening pressure 0-1 cmH_2O. Biochemistry, cell counts and cytology of CSF were normal. LP shunt removal surgery was performed based on the diagnosis of intracranial hypotension due to overdrainage. In a few days after surgery; nausea and vomiting were resolved completely while her hearing loss improved only partially. On the follow-up examination after six months, hearing loss was persisting.

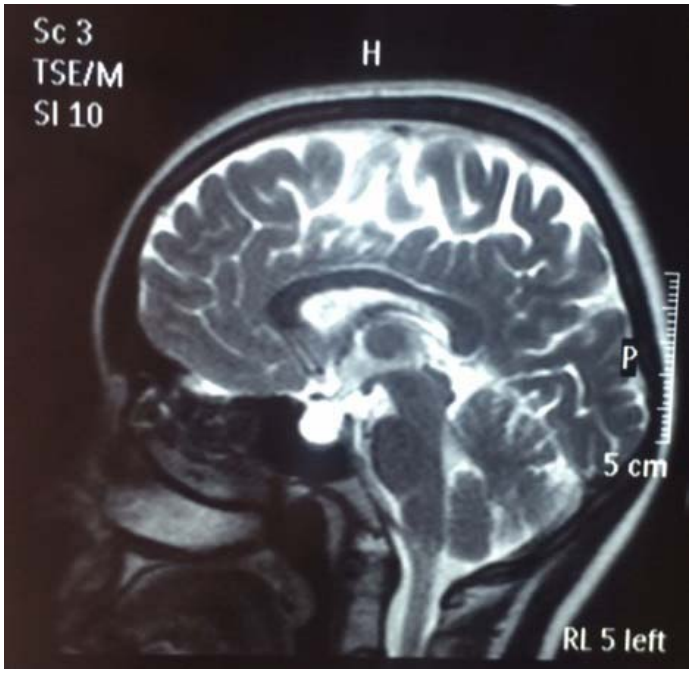

Figure 1: *Sagittal T2-weighted MRI image before the shunt procedure shows, partial empty sella with flattened pituitary gland, and extension of suprasellar cistern to the widened sella.*

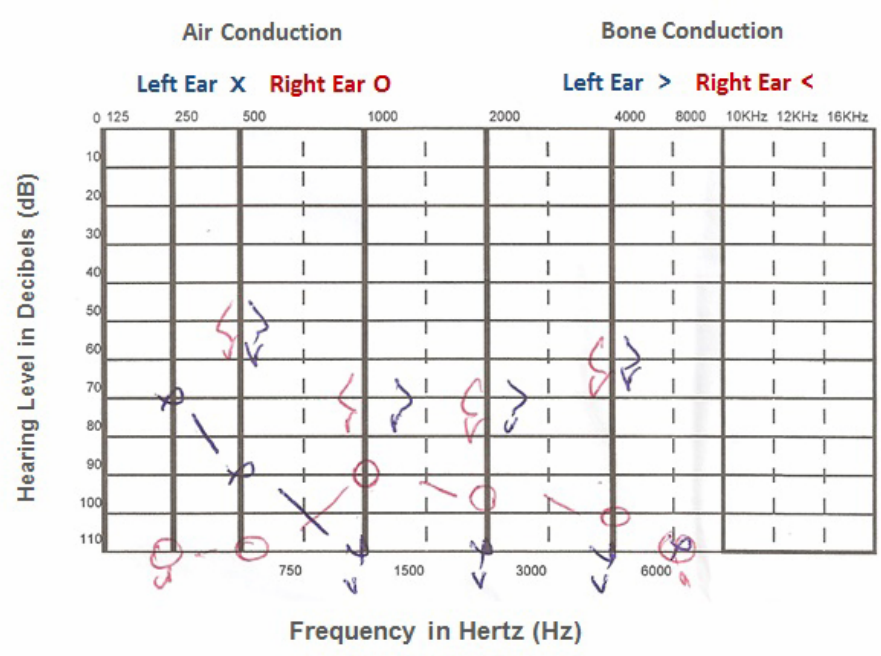

Figure 2: *The conductive hearing loss audiogram of the patient showed bilateral severe sensorineural hearing loss.*

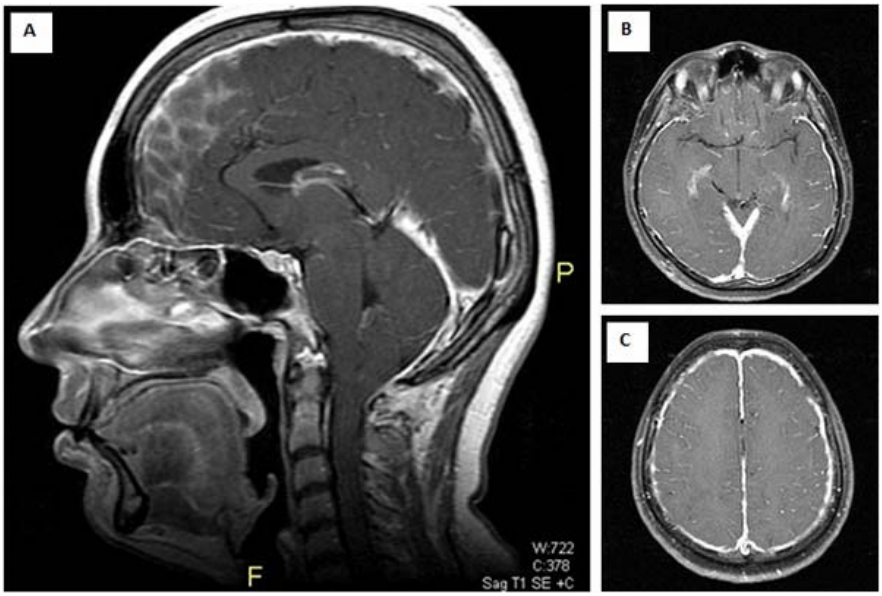

Figure 3: *(A) Sagittal, T1- weighted post-contrast MRI image shows displacement of cerebellar tonsils 10 mm inferior through the foramen magnum. Partial empty sella appereance is not persisting, and the height of the sella is in normal ranges, (B, C) Axial T1- weighted MRI image shows diffuse dural thickening and enhancement.*

DISCUSSION

According to pathophysiological considerations, IH can also be refered as CSF hypovolemia[11]. Orthostatic headache is a frequent and cardinal symptom of IH which results from meningeal irritation and displacement of pain-sensitive structures[1,13,14,16].Other clinical manifestations such as hearing loss, tinnitus, aural fullness, vertigo, dizziness, diplopia, neck pain, radicular upper limb syndromes and other cranial nerve palsies are related to the stretching of the cranial nerves and cervical nerve roots due to caudal displacement of the brain which can be well explained by intracranial hypotension[4,11,17].Although orthostatic headache is the most common symptom in the patients with IH, some patients do not exprience any headaches[11]as in our case. Some cases who presented with hearing loss and tinnitus but not with headache following lumber puncture have been reported previously[18].

Intracranial hypotension caused by CSF leakage which leads to hearing impairment has been reported after several spinal interventions such as surgery[7,15],lumbar puncture, myelography[10],and spinal anesthesia[10,20]. Hearing impairment resolves in most of the patients after the treatment of IH[19]. However, it may also remain permanently in some patients[7,10,15].CSF opening pressure less than 6cmH_2O usually supports the diagnosis of IH; however in some patients with spontaneous intracranial hypotension it might be normal[14].

The possible reason of hearing impairment is the relative endolymphatic hypertension and therefore endolymphatic hydrops caused by perilymphatic hypotonia which may result in inner ear dysfunction[19].Transmission of low CSF pressure to the perilymph of the cochlea via cochlear aquaduct gives rise to perilymphatic hypotonia. Other auditory symptoms such as tinnitus, aural fullness and vertigo might be explained by this theory which mimicks Meniere syndrome[1,6]. Thus, it is crucial for otolaryngologists to differentiate Meniere syndrome from IH since both have similar

auditory symptoms. Pure tone audiometry usually demonstrates low-frequency sensorineural hearing impairment in patients with IH[19].

A study of 16 consecutive patients with intracranial hypotension demonstrated that while 30% of the patients reported neuro-otological symptoms such as dizziness, hearing loss, earfullness and tinnitus along with the orthostatic headache; according to their laboratory evaluations 63% of the patients had audiovestibular impairments[3]. These finding might indicate that patients with intracranial hypotension may have audiovestibular impairments despite being asymptomatic and should be evaluated with laboratory tests also.

The same study also showed spontaneous and/or positional vertical nystagmus in 38% of the patients which was most prominent in supine and head hanging position when intracranial pressure was closest to the normal[3]. Since the patterns of nystagmus seen in those patients was consistent with the patterns of the patients with brainstem and cerebellar lesions, this clinical finding supports compression or traction of the brainstem and cerebellum along the cranial floor as the underlying mechanism. In addition to the vertical nystagmus, horizontal and torsional nystagmus were also seen in those patients which favors endolymphatic hydrops as an other underlying mechanism[3,15].

Typical MRI findings of IH are diffuse dural enchancement, pachymeningeal thickening and enchancement, brain descent, venous engorgement, pituitary hyperemia and subdural collections[1]. In contrast to meningeal enhancement seen in inflammatory and infectious diseases, the enhancement seen in intracranial hypotension involves only dura and is diffuse, linear, symmetrical and prominent rather than being focal or nodular[16]. Dural enhacement caused by dilatation of vascular bed can be well explained by Monroe-Kellie hypothesis in which a decrease in the pressure of an intact skull ends up in compensatory dilatation of the vessels to maintain a constant volume in cranium. Since the increased blood volume is drained by venous system, venous engorgement is also seen in MRI[16]. In our patient, we observed dural enchancement, pachymeningeal thickening and inferior displacement of cerebellar tonsils.

In conclusion, it has been shown in our patient that IH caused by LP shunt overdrainage can result in sensorineural hearing loss as one of the main symptoms. Early consideration and treatment of these patients may provide resolution of the symptoms and prevent them from permanent hearing loss.

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