Title

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Journal

Proceedings of the UCLA Department of Medicine, 16(1)

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Publication Date

2012-08-24

CLINICAL VIGNETTE

Spontaneous Intracranial Hypotension: Case Report

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Case report

A 53-year-old man presented to the outpatient clinic complaining of hearing loss and headaches. He described the headache as a dull ache of moderate intensity that persisted for several days in frontal and occipital regions. The headache improved with lying down. At the onset of the headache, his hearing became distorted and appeared to fluctuate over time. Certain sounds such as white noise had become very irritating to him. He also heard a constant, low-pitched ringing in his ears that sounded like an "old bathroom fan humming". He denied any fever, chills, nausea, vomiting, vertigo, dizziness, unsteady gait, blurry vision, diplopia, photophobia, or recent trauma. His past medical history was unremarkable.
On physical exam his vital signs were normal. Ophthalmologic exam showed normal eye movement, equal, round reactive pupils and normal funduscopic exam. Ear exam showed clear canals with normal tympanic membranes. Neck was supple with normal range of motion. Neurological exam demonstrated intact cranial nerves, normal sensation and muscle strength. His Romberg was negative and there was no cerebellar dysfunction. His audiogram showed a moderate mixed hearing loss in both ears with a mild conductive component in the lower frequencies more notable in the right ear. A MRI of the brain showed bilateral enhancing subdural fluid collections consistent with intracranial hypotension. No specific lesions of the cerebellopontine angle or inner ear were noted.
The patient was diagnosed with spontaneous intracranial hypotension and underwent an epidural blood patch. After treatment, the patient reported almost immediate relief from headaches. However, within three days, the headaches returned. Subsequently, he returned for a second epidural treatment which provided relief from the headaches, but his auditory thresholds did not improve, nor had the tinnitus abated.

Discussion

Spontaneous intracranial hypotension presenting with headaches is often misdiagnosed as migraines, tension headaches, or viral meningitis. The estimated annual incidence is 5 per 100,000. Women are twice as likely to have this condition compared with men with the onset usually occurring at age 40 to 50.[1] Spontaneous intracranial hypotension is caused by spontaneous spinal cerebrospinal fluid (CSF) leak. The cause of CSF leaks is often not known. Many patients with a CSF leak have been found to have connective tissue disorders including Marfan and Ehlers-Danlos syndromes. About one third of patients can recall some trauma, even if minor[1]. It is postulated that minor trauma could cause rupture of spinal epidural cysts or perineural cysts[2].

The classic headache associated with spontaneous intracranial hypotension is an orthostatic headache that worsens or starts 15 minutes after sitting or standing. The headache in spontaneous intracranial hypotension may be gradual or acute in onset, and may localize to the frontal or occipital regions[3].

Usually the headache improves by lying down. Patients may describe the headache as either diffuse or localized often to the occipital region and it is almost always bilateral. The headache is believed to be caused by the sagging brain pulling on pain-sensitive structures such as sensory nerves and bridging veins[2]. The most common associated symptoms are posterior neck pain or stiffness, nausea and vomiting. Hearing complaints are also very common and are believed to be caused by pressure changes that are transmitted to the cochlea. The sagging brain may also stretch the eighth nerve and other cranial nerves, which is felt to be the mechanism for visual changes such as blurred vision, diplopia, and photophobia[1]. Other common symptoms are dizziness, vertigo, unsteadiness, and hiccups[2].

Physical exam is usually normal, however patients can have abducens palsies or visual field defects[2]. The lumbar puncture opening pressure usually is low ranging from 0- 70 mmH20.

The International Classification of Headache Disorders requires the following to diagnose a

headache due to spontaneous spinal CSF leak: the orthostatic headache as described above along with at least one additional symptom(neck stiffness, tinnitus, hypacusia, photophobia, or nausea) and confirmatory evidence of low CSF pressure such as on magnetic resonance imaging (MRI) with gadolinium, conventional myelography, computed tomography (CT) myelography, cisternography, or CSF opening pressure[1]. The most helpful study is a MRI of the brain with the following characteristics: subdural fluid collections, enhancement of the pachymeninges, engorgement of the venous structures, pituitary hyperemia, and sagging of the brain. The meningeal enhancement has been described as continuous over the convexities, along the interhemispheric fissure and tentorium but not involving the depths of the sulci[4]. Venous hypervolemia is felt to be the cause for enhancement of the pachymeninges[5]. Of note, it has been estimated that up to 20 percent of patients can have a normal MRI. If this is the case, CT myelography is recommended as it is the best at localizing the site of the leak[2].

Although there have been no randomized clinical studies to evaluate treatment options, the initial treatment is either conservative treatment or an epidural blood patch which consists of autologous blood being injected into the epidural space. It is believed to work by sealing the leak and is effective in relieving symptoms in a third of patients[1]. If this fails, it can be repeated. Other treatments such as a directed epidural blood patch or percutaneous placement of a fibrin seal require knowledge of the exact site of the CSF leak[1]. Patients who fail these treatments should be referred for surgical repair. Despite treatment, some patients continue to have persistent symptoms. In 2005, Schievink conducted a study of patients with intracranial hypotension diagnosed by CT myelography. Out of 33 patients, 22% had a normal MRI. They found that patients with a normal MRI had poorer outcomes after treatment. The usual abnormal findings on MRI including pachymeningeal enhancement are felt to be compensatory reactions to the CSF leak. Schievink believed that the absence of these findings are likely to indicate the inability to compensate for the CSF leak and therefore the patient has a worse prognosis[6].

In the present case, the patient did have the typical orthostatic headache along with hearing loss and tinnitus, but he lacked the usual symptoms of neck pain, neck stiffness, nausea, and vomiting. Although spontaneous intracranial hypotension is not common, it should be considered in the differential diagnosis of new-onset persistent headaches, especially in middle-aged adults.

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Submitted on August 24, 2012