Spontaneous Intracranial Hypotension and the Otolaryngologist
Case Study and Literature Review
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ABSTRACT

There has been a strong push in medical literature to shed light on an uncommon but increasingly diagnosed cause for headache that has escaped most Otolaryngology publications. Spontaneous intracranial hypotension (SIH) which has an estimated incidence of 5 per 100,000 is associated with an orthostatic headache, has gained attention in medical media, but almost exclusively within the fields of neurology, radiology and anesthesiology. It is important for the Otolaryngologist to be aware of this clinical entity to provide the utmost patient care in the management of headache. We present the case of a patient referred to our Otolaryngology practice for evaluation of orthostatic headache that was presumed to be secondary to a cranial base dehiscence, but found to be related to cerebrospinal fluid leak within the thecal nerve rootlets and subsequent SIH.

INTRODUCTION/BACKGROUND

The phenomenon of spontaneous idiopathic hypotension (SIH) is not new with the first case described in 1938 by the German neurologist Schaltenbrand; however, it has been increasingly studied and diagnosed over the past few decades, particularly with advent and advancement of various imaging modalities.1 Particular attention has been paid to this clinical entity within the fields of neurology, radiology and anesthesiology, but scant information and reports can be found within the otorhinolaryngology and ophtalmology literature. This lack of readily available information creates a diagnostic and therapeutic gap in patient management, as these fields frequently are involved in the diagnosis and treatments of headache patients. For the ENT surgeon, this can be from referrals for headache thought to be second not only to skull base erosion with CSF leak, but rhinogenic cephalalgia, sinusitis, and temporomandibular joint disorders, etc. that may cause confusion with the diagnosis of SIH. Additionally, many of the SIH patients experience visual changes, hearing loss, tinnitus, vertigo, neck pain and other symptoms that may generate a referral for these specialty services. The diagnosis of SIH can be elusive and frequently misdiagnosed as meningitis, migraine and psychogenic disorders prior to correct identification of the disease. Even with proper diagnosis there are typically extensive delays of weeks to many years for some patients. 2 We present the case of a middle aged woman who presented to the otolaryngology office for evaluation of headache and presumed sinusitis. This paper is designed to review the clinical entity as it pertains to otolaryngology and review the current diagnostic and treatment strategies.

CASE PRESENTATION

A 46 year old female referred to the Otolaryngology office at Drexel University Hospital for evaluation of headache and sinusitis. Patient had complaints of nasal congestion and post nasal drip for 7 months associated with a bilateral frontal headache, described as throbbing, for 1 month. The headache was noted to be more prevalent when standing and improved while lying flat. Additionally, she complained of persistent fatigue and nausea during this timeframe. There were no fevers, photophobia, phonophobia, diplopia or other vision changes, hearing loss, or tinnitus. She did not experience any significant clear rhinorrhea, otorrhea, or salty taste. On physical examination she had a septic relief to the left with mild macular edema and nasal endoscopy showed mucopus. Treatment was initiated with an antibiotic and tapering dose of steroids. Subsequently, the patient continued to show no improvement and was admitted to the hospital for further evaluation.

Sinus disease was ruled out on maxillofacial computed tomography, broad spectrum antibiotics were initiated to cover meningitis, but was completed after that demonstrated diffuse pachymeningeal thickening and enhancement. Lumbar puncture was performed and significant for low opening pressure of 4mH2O and mild elevation in protein. NM cisternography showed a slow CSF leak around multiple thoracic nerve roots. Based on these findings consistent with spontaneous idiopathic hypotension the patient underwent two blood patches with anesthesia with almost complete resolution prior to hospital discharge. On office follow-up 3 and 6 months after treatment, the patient has been doing well without refractory headache.

DISCUSSION

Etiology and pathophysiology

SIH is classified as an uncommon disorder, but appears to likely be more common than previously predicted and the cause for the CSF leak has yet to be fully elucidated. There is evidence to suggest that an underlying weakness of the connective tissue; however, less than 5% belong to a named and documented disorder of connective tissue such as Marfan, Ehlers-Danlos and autosomal polycystic kidney disease.3,4,5,6,7,8,9,10,11

The cause for the CSF leak has yet to be fully elucidated. There is evidence to suggest that an underlying weakness of the pachymeninges may contribute. Recent evidence suggests that at least some cases there might be a genetic defect of the connective tissue; however, less than 5% belong to a named and documented disorder of connective tissue such as Marfan, Ehlers-Danlos and autosomal polylysyl kidney disease.11

Clinical presentation

The chief complaint amongst patients with SIH is a positional headache that worsens on sitting up or standing in the upright position with some degree of relief on lying supine or flat. The headache is most often bilateral and can occur in any location but most common in the occipital region. Increases in intracranial pressure from laughing, sneezing, coughing or Valsalva may exacerbate the headache in certain patients.4 Symptoms that may accompany the headache can assist the practitioner in discerning the correct diagnosis. Most commonly these include cervicalgia, neck stiffness, nausea and vomiting. Many other miscellaneous symptoms can be present and are listed routinely seen by otorhinolaryngology and include alterations in hearing, tinnitus, vertigo, dysgeusia, photophobia, facial numbness and diplopia in a study by Chung et al, dizziness, neck stiffness, aural fullness, tinnitus and hearing loss were present in 30%, 17%, 20%, 20% and 3% respectively.12 The headache is described as most commonly low frequency in nature that can fluctuate depending on patient position and overall hydration status.12

BIBLIOGRAPHY

2. Cho et al performed a retrospective study on 56 patients comparing blind epidural blood patches (EBP) directed into the wide lumbar space, typically at the L3/L4 level, and targeted EBP aimed at the site of the defined leak. After one injection the blind EBP had a success rate of 52% compared to 87.5% for the targeted approach.12 Most of the literature states the thoracic and cervico-thoracic junction to be the most common locations for the leak.12,13

CONCLUSIONS

Our patient presented with the classic orthostatic headache and an initial brain MRI demonstrated pachymeningeal enhancement. Spinal MRI failed to discern a CSF leak and radionuclide cisternography was completed, which successfully localized the CSF leak to the multiple upper thoracic nerve roots. Definitive treatment was provided with epidural blood patches directed into the lumbar space with complete resolution of symptoms after two injections. During office follow up at 3 and 6 months the patient remained asymptomatic.

Management

Conservative measures including bed rest, hydration and abdominal binders are typically used first for a couple of weeks with variable amounts of caffeine, theophylline and steroids.13,14 The utilization of conservative measures must be weighed against the morbidity for the patient. Those patients more severely affected may elect to proceed to more invasive techniques. The main treatment modality is an injection of autologous blood into the epidural space to provide a blood patch to the leaking area.12 Cho et al performed a retrospective study on 56 patients comparing blind epidural blood patches (EBP) directed into the wide lumbar space, typically at the L3/L4 level, and targeted EBP aimed at the site of the defined leak. After one injection the blind EBP had a success rate of 52% compared to 87.5% for the targeted approach.12 Most of the literature states the thoracic and cervico-thoracic junction to be the most common locations for the site of the leak.12,13

The mnemonic SEEPS represents the 5 most commonly seen abnormalities on brain MRI and stands for (1) subdural fluid collections with variable mass effect, (2) diffuse enhancement of pachymeninges, (3) venous engorgement, (4) intracranial hypotension and (5) ligging of the brain.12,13

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In conclusion, it is important for the ENT physician to be aware of this clinical entity when evaluating headache patients, particularly when the headache is postural and they present with concomitant otorhinolaryngologic complaints and should be considered during the patient evaluation. It is important for the ENT physician to be aware of this clinical entity when evaluating headache patients, particularly when the headache is postural and they present with concomitant hearing loss, tinnitus, vertigo and other cranial neuropathies. Though previously perceived as an extremely rare entity, the prevalence of this disease process is likely underestimated and the number of patients with this diagnosis will rise in the future. Presumably with the increase in recognized cases there will be greater opportunity for randomized controlled trials to further elucidate the proper diagnostic and treatment regimens in evidence based fashion.

key words: Spontaneous, intracranial hypotension, cerebrospinal fluid leak, orthostatic headache, otorhinolaryngologist, idiopathic intracranial hypotension

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DIAGNOSIS

Much of the diagnosis is based on clinical suspicion and confirmed through various imaging modalities. According to the International Headache Society, recent revision of the International Classification of Headache Disorders (ICHD-3-F edition), the criteria include any headache that develops within close proximity to CSF hypotension or leakage. They also require a low CSF pressure of <40mm H2O and/or suggestion of CSF leakage on imaging and exclusion of alternate diagnoses within the ICHD-3 classification.