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CASE REPORT

A rare case of orthostatic headache due to spontaneous intracranial hypotension

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Key Clinical Message

Headache is one of the most common clinical entities, and has a long list of differential diagnoses; however, one of the more uncommon causes of postural headache is spontaneous intracranial hypotension. It is important that clinicians be aware of this condition, as it is often overlooked, leading to invasive and unnecessary diagnostic testing. A good history and physical exam, paired with an MRI of the brain is sufficient to make the diagnosis of SIH, relieve the symptomology of the patient in a quick and efficient manner, and avoid costly invasive procedures.

Keywords

Intracranial hypotension, intractable headache, orthostatic headache, spontaneous.

Introduction

Spontaneous intracranial hypotension (SIH) is characterized by postural headaches secondary to low CSF pressures, which are not attributable to lumbar puncture (LP), trauma, surgery or any other inciting event. SIH is typically due to spontaneous spinal CSF leaks, and are an important and often misdiagnosed cause of new onset headache in young and middle-aged individuals, especially women.

Case

This is a case of a 31-year-old female with no significant past medical history, and no history of oral contraceptive use, who came to the emergency department complaining of sudden onset intense 10/10 frontal headache worsened by sitting upright and/or standing, and alleviated by lying supine, associated with nausea and vomiting.

On exam, vital signs were found to be blood pressure 101/70 mmHg, heart rate 61/min, respiratory rate 20/min, and temperature 98.7°F. Laboratory findings were found to be as follows: Hb 14.3 g/dL, Hct 42.3%, WBC 9.5 K/μL, Plt 297 K/μL, glucose 91 mg/dL, BUN 15 mg/dL, Cr 0.8 mg/dL, Na 138 mEq/L, K 4.4 mEq/L, Cl 101 mEq/L, and CO2 25 mEq/L. The rest of the physical exam, including neurological exam, was within normal limits. This patient denies any history of lumbar puncture in the past. Upon arrival to the emergency department, a lumbar puncture was offered to the patient; however, she refused at that time. A CT scan of the head without contrast was done in the ED as well and showed no acute intracranial processes.

The patient's headache was being treated with fioricet as needed for pain scale 2-5/10 and with morphine 1 mg IV as needed for pain scale 7-10/10. An MRA was done to rule out the possibility of Berry aneurysm, which was found to be unremarkable. An MRI with and without contrast showed dilatation of the right transverse sinus (Figs. 1 and 2) and paucity of CSF fluid in both optic nerve sheaths, compatible with intracranial hypotension.

As the patient's headache continued without any resolution unless the patient remained supine, a neurology consult was ordered, and the patient agreed to a lumbar puncture. The lumbar puncture was performed successfully under fluoroscopic guidance, which yielded an opening pressure of 0 mmH_2O.

Based on the all the above findings the patient was diagnosed with spontaneous intracranial hypotension, and morphine was discontinued, and fioricet alone was continued. The patient greatly improved with fioricet and

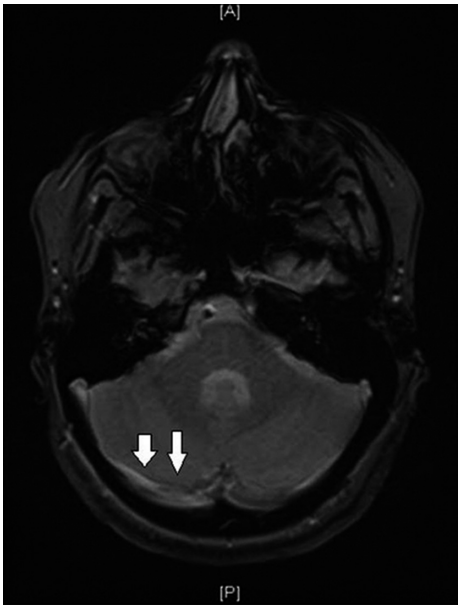

Figure 1. MRI Brain demonstrating mild dilatation of the dominant right transverse sinus, compatible with intracranial hypotension.

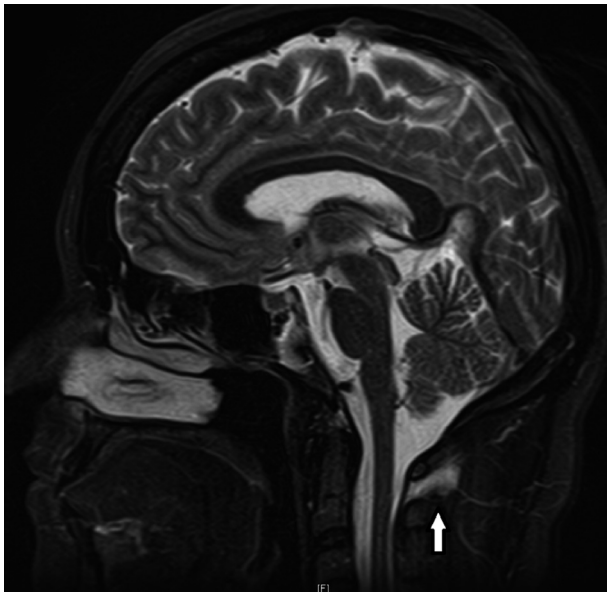

Figure 2. MRI Brain – Nonspecific fluid/edema within the C1-C2 retrospinal location. This finding has been reported in the setting of intracranial hypotension.

caffeine tablets. Patient was discharged home with follow-up in an outpatient setting.

Discussion

While postural headaches attributable to spontaneous intracranial hypotension in the postlumbar puncture setting is quite frequent, idiopathic spontaneous intracranial hypotension is an uncommon, but important cause of new onset postural headache in the young to middle-aged population, especially females [1].

Whether idiopathic or iatrogenic, spontaneous intracranial hypotension is classically described as an orthostatic headache that worsens when the patient is sitting upright or standing and resides when lying flat [1,2], and can be accompanied by a multitude of symptoms, including nausea, vomiting, anorexia, neck pain, dizziness, photophobia, etc. [3]. Idiopathic spontaneous intracranial hypotension is usually caused by a persistent occult CSF leak, which presents with a postural headache as the most prominent symptom [4].

Recognizing idiopathic spontaneous intracranial hypotension in patients is essential and may prevent costly unnecessary procedures. A retrospective study done by Schievink involving 18 patients with headache due to idiopathic intracranial hypotension demonstrated that 17, or 94%, were incorrectly diagnosed with a diagnostic delay ranging from 4 days to 13 years [1].

Diagnosis of idiopathic spontaneous intracranial hypotension is ideally obtained with MRI of the head with gadolinium, which often shows diffuse meningeal enhancement, engorgement of the venous sinuses, subdural fluid collections, or descent of the cerebellar tonsils, often misdiagnosed as a chiari malformation [2,4,6].

The aforementioned imaging findings are explained using the Monro-Kellie hypothesis which states that the relationship between intracranial pressure and CSF volume, blood and brain tissue form a constant [5]. When one of these variables is either increased or decreased, the other variables must compensate accordingly to maintain the constant pressure [5]. It is important to note that these imaging findings are only significant in the correct clinical setting of a postural headache not attributable to any other cause.

Lumbar puncture with opening pressures is confirmatory in the diagnosis of spontaneous intracranial hypotension and are expected to be 0–60 mm H_2O (normal pressures range from 60 to 120 mm H_2O) [4], however, it is important to note that lumbar puncture may worsen the patient's symptoms by removing more CSF and potentially causing another leak, leading to post-LP iatrogenic intracranial hypotension related headache [4].

Initial preferred management of idiopathic spontaneous intracranial hypotension is conservative treatment with bed rest, fluid replacement and caffeine, in the form of caffeine tablets and/or high caffeine dietary intake, which due to its vasoconstricive properties has proven to work well [4–6]. If conservative treatment is found to be ineffective, then a nonfocalized epidural blood patch may be

placed. Should this too prove to be ineffective, then a CT myelogram may be done to localize the leak and then corrected with a focal epidural blood patch [4–6].

Nonetheless, idiopathic spontaneous intracranial hypotension is a manageable condition, which if approached correctly may relieve your patient of significant distress, sooner rather than later, and may avoid costly invasive procedures.

Conclusion

While spontaneous intracranial hypotension is an uncommon diagnosis, misdiagnosis of it is quite commonplace and is often confused with migraine, chiari malformation, pseudotumor cerebrii or aneurysms, which subsequently leads to wrong and potentially harmful procedures including craniotomies and brain biopsies. It is therefore important that both the primary care physicians and the neurologists keep idiopathic SIH in their list of differential diagnoses when treating a postural headache in order to relieve symptoms in a quick and efficient manner as well as to avoid costly and invasive procedures.

Conflict of Interest

None declared.

References

1. Schievink, W. I. 2006. Spontaneous spinal cerebrospinal fluid leaks and intracranial hypotension. JAMA 295:2286–2296.
2. Gordon, N. 2009. Spontaneous intracranial hypotension. Dev. Med. Child Neurol. 51:932–935.
3. Goadsby, P. J., C. Boes, and C. L. M. Sudlow. 2002. Low CSF volume (Pressure) headache. Pract. Neurol. 2:192–197.
4. Greif, S., S. Mandel, D. J. Langer, and R. A. Ortiz. 2014. Spontaneous intracranial hypotension: spontaneous intracranial hypotension remains an underdiagnosed etiology of new-onset headache. Pract. Neurol. 6:24–27.
5. Zaatreh, M., and A. Finkel. 2002. Spontaneous intracranial hypotension. South. Med. J. 95:1342–1346.
6. Franzini, A., G. Messina, E. Mea, L. Chiapparini, and G. Bussone. 2004. Spontaneous intracranial hypotension: diagnostic and therapeutic implications in neurosurgical practice. Neurol. Sci. 32(Suppl 3):S287–S290.