

SPONTANEOUS INTRACRANIAL HYPOTENSION (SIH) PAIN RELIEF WITH LUMBAR EPIDURAL BLOOD PATCH (EBP) IN A PATIENT WITH CANCER: A CASE REPORT

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Background: Spontaneous intracranial hypotension (SIH) is a challenging chronic headache pain condition that can severely debilitate patients. It can be caused by any etiology in which intracranial cerebrospinal fluid (CSF) outflow surpasses CSF production. Conservative first-line therapy typically involves caffeine, hydration, medications, bed rest, and/or abdominal binding. Refractory cases involve epidural blood patch and surgical repair. To date, there have been no published studies or case reports on the treatment of SIH using epidural blood patch (EBP) in patients with cancer. We present a case demonstrating the utility of EBP in the treatment of refractory SIH in a patient with cancer.

Case Report: A 65-year-old woman with lung adenocarcinoma presented with refractory SIH of 2 years. She failed conservative first-line therapies and was evaluated by multiple services without relief. She then received an EBP with 75% relief of symptoms, and 100% relief of all symptoms with a repeat EBP.

Conclusions: Given its overall safety and effectiveness, EBP should be considered as a treatment option in patients with cancer who present with SIH.

Key words: Cancer, cerebrospinal fluid, chronic pain, epidural blood patch, headache, oncology, spontaneous intracranial hypotension

BACKGROUND

Intracranial hypotension is a syndrome that is caused by etiologies in which cerebrospinal fluid (CSF) outflow exceeds CSF production. The majority of etiologies are attributed to traumatic etiologies, such as procedures including but not limited to lumbar punctures, neuraxial anesthetics, and back surgeries. Atraumatic etiologies include CSF venous fistulas, hereditary malformations, spinal dural diverticula, and idiopathic causes (1,2). The headache may last for days to weeks if secondary to traumatic etiologies, and for years if spontaneous in origin.

Albeit highly variable, the symptomatology classically

consists of pain in the posterior skull base, posterior neck and upper thoracic spine, mild-to-moderate stiffness and pain of the neck and shoulders, dizziness, imbalance, photophobia, phonophobia, hearing changes such as ringing or muffing, and nausea (3). Less common symptoms include visual changes, facial pain, numbness, and changes in ability to taste (3). Symptoms are also classically alleviated with recumbent positions and exacerbated with upright positions. It is typically associated with a CSF opening pressure of less than 6 cm H₂O (4). Magnetic resonance imaging (MRI) of the brain with contrast medium features qualitative findings such as pachymeningeal thickening

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and enhancement, dural venous engorgement, tonsillar herniation and sagging due to decreased CSF volume, and subdural effusions (5,6). Computed tomography myelography may be used as an adjunct if MRI findings are equivocal and symptomatology is consistent with intracranial hypotension.

The diagnosis of spontaneous intracranial hypotension (SIH) has been established by the International Classification of Headache Disorders, 3rd edition, as having the following attributes: any headache with temporal relation to low CSF pressure or CSF leakage, either or both low CSF pressure and evidence of CSF leakage on imaging, absence of a procedure or trauma known to cause CSF leakage, and no other diagnosis that better accounts for these attributes (4).

In patients with cancer, pain symptoms require thorough workup as delayed diagnoses of malignancy can have devastating consequences. To date, there have been no published studies or case reports on the treatment of SIH with lumbar epidural blood patch (EBP) in patients with cancer. We present a case of a 65-year-old woman with lung adenocarcinoma presenting with refractory SIH of 2 years who received significant relief with EBP. A written HIPAA authorization to use and disclose existing protected health information in this case report was obtained and saved in the patient's medical record.

CASE

A 65-year-old woman with left upper lobe lung adenocarcinoma status post lobectomy presented with a 2-year history of a frontal and occipital headache. It was described as a stabbing pain to the right eye with radiation to the back of her head, and a concurrent engine humming sensation with left ear fullness and muffling. The frequency progressed from sporadic to daily. Upright positions, coughing, and loud noises exacerbated the pain. Lying recumbent and ibuprofen as needed alleviated her headache.

She initially presented to the otolaryngology service as an inpatient consult for left ear pain, for which she was subsequently referred to neurology. MRI brain with and without contrast medium was notable for dura-arachnoid pachymeningeal enhancement, and bilateral extra-axial fluid collections in the subdural space with more fluid on the left side; metastatic disease and lymphoma were less likely due to lack of nodularity and mass effect. MRI spine findings were unremarkable for findings consistent with metastatic causes. One year after the onset of headaches and prior to the aforementioned MRI brain, she had an

MRI brain that also revealed pachymeningeal enhancement without signs consistent with malignancy. After failure of symptom resolution with conservative therapy and a low index of suspicion for malignant etiologies, neurology referred the patient to our pain management service for further evaluation. Physical exam was notable for improvement of symptoms upon lying recumbent, and no neurologic deficits were found. Of note, the patient has never received a lumbar puncture or other spinal procedures.

A lumbar EBP was performed. The patient was placed in a prone position and her lumbar spine was sterilized, draped, and anesthetized subcutaneously. Under fluoroscopy, the L4-L5 disc space was identified in the anterior-posterior view, and a 17-gauge Tuohy needle was advanced through the anesthetized tract. The loss-of-resistance technique with saline was employed to localize the epidural space. Contrast medium was used to confirm placement in both lateral and anterior-posterior views. A total of 20 mL of autologous blood was drawn from the patient's 20-gauge peripheral intravenous catheter in the right antecubital vein. All 20 mL of blood were slowly administered into the epidural space without resistance.

At one week post EBP, the patient endorsed 75% relief of her headache. She only noted one headache over the entire week, and resolution of her left ear pain for 2 days with ibuprofen. The patient then elected to repeat EBP at 3 weeks post EBP for her residual headache and left ear pressure pain. She subsequently received 100% relief from all symptoms with her second EBP. She denied adverse effects including but not limited to weakness, numbness, rash, swelling, nausea, changes in bowel habits, fever, and erythema at her EBP site.

DISCUSSION

SIH can commonly occur without a precipitating event and can occur after traumatic etiologies resolve. SIH has an annual incidence of 5 per 100,000 (7). This incidence, however, is likely an underestimation because variability in presenting signs and symptoms and low awareness of the disorder contribute to delayed diagnoses. Although men, women, and children of any age may develop SIH, it is diagnosed most often in women with a peak diagnosis age of 40 years (7).

The goal of SIH treatment is to increase CSF volume and limit CSF leak. Conservative first-line therapy typically involves oral hydration, caffeine, acetaminophen, nonsteroidal anti-inflammatory drugs, and rest. Secondary conservative therapy involves steroids, acetazolamide,

gabapentin, pregabalin, theophylline, methylxanthines, sumatriptan, adrenocorticotrophic hormone, cosyntropin, desmopressin, and abdominal binding (8). Advanced interventional therapies consist of EBP and surgical repair. Of note, SIH is less responsive to first-line conservative therapy and more amenable to EBP when compared to postdural puncture headache, and there is a lack of conclusive evidence for its effectiveness secondary to conservative therapies (8,9). EBP has been found to provide relief in 80% to 90% of intracranial hypotension headaches, and patients sometimes require multiple EBPs before complete resolution of symptoms (10). EBP treats intracranial hypotension by 2 theories: the “pressure patch” hypothesis in which the injected blood increases epidural pressure, which subsequently elevates subarachnoid CSF pressure by compressing the dura rostrally; and the “plug” hypothesis in which the injected blood seals the dural hole and prevents further CSF leak into the epidural space. The “pressure patch” hypothesis is likely to be the predominant mechanism in SIH because there is no identifiable source of CSF leak.

Until now, there are no reports discussing the use of EBP for cancer patients with SIH. The only studies pertaining to cancer and EBP involve a limited number of studies that assessed risk of cancer seeding from EBP in patients with leukemia or lymphoma. EBP was found to bear a low cancer-seeding risk when used to treat postdural puncture headache unresponsive to conservative treatments (11). It is imperative to rule out cancer as a cause of pain in patients with a history of cancer, as early diagnosis is a strong predictor of survival (12). Upon ruling out cancer as a cause of intracranial hypotension symptoms, it is important to

treat the underlying etiology of pain to maximize patient quality of life, functionality, and patient impact on the social environment and health care system (13).

Our patient with a history of cancer presented with signs, symptoms, and imaging consistent with SIH that was resistant to conservative treatment for 2 years. A unique consideration for our patient was the MRI brain findings of pachymeningeal enhancement given her history of lung cancer and therefore her predisposition to malignancy in the brain. This MRI finding supported the diagnosis of SIH although it can also be found in malignant etiologies. The index of suspicion for a malignant primary or metastatic etiology was lower given the lack of nodularity, mass effect, leptomenigeal enhancement, constitutional symptoms, and her prior MRI brain that also showed pachymeningeal enhancement. Our patient received significant relief with her first lumbar EBP and was subsequently able to minimize her residual symptoms with an adjunct conservative first-line medication. Her repeat EBP was then ultimately able to provide 100% relief of all symptoms without adverse effects. Given its overall safety and effectiveness, EBP should be considered as a treatment option in patients with cancer who present with SIH.

Author Contributions

J.L. — First author. Involved in writing, reviewing, editing, and patient data collection.

M.C. — Second author. Involved in writing, reviewing, editing, and patient data collection.

C.L.S. — Primary investigator. Involved in writing, reviewing, editing, and patient data collection.

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