

Cerebrospinal Fluid Hypovolemia: A Case Report of a Red Herring

Abstracts

Mild intracranial hypotension can lead to classically recognizable symptoms such as positional headaches, nausea, vomiting, and occasionally blurred vision. Less commonly, severe cerebrospinal fluid (CSF) hypovolemia can lead to a life-threatening condition that mimics intracranial hypertension, including transtentorial herniation and subsequent rapid neurologic decline. In this report we present a unique case of severe intracranial hypotension from a thoracic tumor resection that led to symptoms initially mistaken for intracranial hypertension, however ultimately correctly diagnosed as severe CSF hypovolemia that improved with dural repair. Additionally, we describe a rare angiography finding associated with CSF hypovolemia, kinking of the basilar artery. Here we report a 47-year-old female with neurofibromatosis Type 2 found to have a T3 intradural extramedullary tumor. She initially presented with urinary incontinence and gait/balance difficulty. She underwent thoracic laminectomies at T3 and T4 for the excision of the lesion. She was discharged on postoperative day 4. On postoperative day 9, she was noted to have nausea, vomiting, and decreased consciousness. Head computed tomography (CT) demonstrated acute downward herniation. She was transferred to our institution from a community facility obtunded and was intubated for airway protection. She was placed in the Trendelenburg position with immediate improvement, and declined every time her head was raised. Angiogram showed significant kinking of her basilar artery. A CT myelogram revealed a CSF leak from her recent thoracic surgery. She underwent exploration of her thoracic wound, and a ventral durotomy was repaired. Following this, she began to tolerate the head of bed elevations and recovered back to her neurologic baseline. A postoperative head CT angiography obtained before discharge showed improvement of her basilar kink. Mild intracranial hypotension is a common finding in patients who undergo procedures that enter the CSF space. Severe intracranial hypotension can easily be missed diagnosed as the signs on the exam are similar to patients with signs of intracranial hypertension. It is of paramount importance that the clinician recognizes brain sag, as the treatment algorithms are vastly different from that of intracranial hypertension leading to transtentorial herniation.

Keywords: *Angiography, basilar artery kink, brain herniation, brain sag, cerebrospinal fluid hypovolemia*

Introduction

Intracranial hypotension or brain sag can lead to classically identifiable symptoms such as positional headaches, nausea, vomiting, and occasionally blurred vision [Table 1]. However, severe cerebrospinal fluid (CSF) hypovolemia can lead to a life-threatening condition that can mimic intracranial hypertension such as transtentorial herniation and poor neurologic exam.^[1] In this case report, we present a case of severe intracranial hypotension from a thoracic tumor resection leading to red herring symptoms of intracranial hypertension. Ultimately, the pathology was proven to be CSF hypovolemia from an

occult durotomy resulting in a CSF leak. We discuss the implications of this diagnosis and how to avoid misdiagnosis. In addition, we present a rare but well-reported finding of basilar artery kinking in the setting of severe intracranial hypotension which dramatically improved after durotomy repair.

Case Report

A 47-year-old female with neurofibromatosis Type 2, bilateral acoustic neuromas, deafness, multiple intracranial tumors requiring resection, and focal epilepsy presented with urinary incontinence and gait/balance difficulty. On work up the patient was found to have a T3 intradural extramedullary mass [Figure 1],

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L4-5 spondylolisthesis, and a stable C4-5 disc herniation. The patient underwent thoracic laminectomies at T3 and T4 for the resection of her thoracic mass. The immediate postoperative course was complicated by transient right lower extremity weakness, which subsequently returned to preoperative baseline, and the patient was discharged to an inpatient rehabilitation facility on postoperative day 4.

On postoperative day 9, inpatient rehabilitation staff found the patient to be experiencing nausea, vomiting, and decreased consciousness. Computed tomography (CT) head demonstrated acute downward herniation with cisternal effacement, including suprasellar, interpeduncular, and crural cisterns, however not ambient or quadrigeminal cisterns [Figure 2A and B]. On arrival to our institution, the patient was obtunded and intubated for airway protection. Urgent magnetic resonance imaging (MRI) of the brain demonstrated diffuse pachymeningeal enhancement and significant deformation of the brain stem [Figure 2C and D]. This imaging in conjunction with her clinical presentation raised some concern for possible intracranial hypertension. Despite this a central line was placed to provide hypertonic saline, which was complicated by inadvertent injury to the right vertebral artery. After further review, discussion of a potential durotomy from her recent thoracic surgery raised serious consideration for an occult CSF leak and resultant intracranial hypotension. Placement of the patient in Trendelenburg position showed immediate clinical improvement. Expectedly, the patient would decline neurologically when the head of bed was

elevated. An angiogram was performed to evaluate the iatrogenic vertebral artery injury encountered during central line placement, and ultimately the right vertebral artery was sacrificed to allow for safe catheter removal. Incidentally this angiogram revealed significant kinking of the basilar artery [Figure 3B]. Based on clinical concern for CSF hypovolemia, further supported by kinking of the basilar artery, a CT thoracic myelogram was performed, which did in fact show a CSF leak at the prior thoracic surgery site [Figure 4]. Subsequently, the patient underwent exploration of the thoracic wound, and a ventral durotomy was found and repaired. Two small artificial dural patches were placed laterally and ventrally to the thoracic cord at the level of the durotomy. Following this repair, the patient was able to once again tolerate sitting up, with eventual recovery back to her neurologic baseline. CT Angiography (CTA) head and neck was obtained prior to discharge which showed interval improvement of the basilar kink with resolution of her CSF hypotension [Figure 3A and 3C].

Discussion

Intracranial hypotension, or “brain sag,” leading to extreme changes in neurological exam remains an uncommon occurrence even after intradural neurosurgical procedures. The well-known signs easily recognizable as indicative of intracranial hypotension are postural headaches in addition to occasional nausea, emesis, photophobia, and blurry vision. These signs are related to a decrease in the already negative CSF vertex pressure in the erect posture.^[1] The headache easily resolves with the placement of the patient in a recumbent position and proceeding with a blood patch or CSF leak repair usually results in a permanent cure. It is the loss of overall CSF volume with resultant brain sag and pressure on pain-sensitive structures (located in the dura?) that leads to postural headaches and the radiologic findings that have been well described in the literature.^[2] These findings include subdural hematoma, pachymeningeal enhancement, cerebellar tonsillar herniation, tentorial herniation of the splenium and cingulate gyrus, and midbrain

Table 1: Common clinical and radiographic signs of cerebrospinal fluid (CSF) hypovolemia

Clinical Signs	Radiographic Signs
Postural headaches, worse upright	Pachymeningeal enhancement
Nausea	Cerebellar tonsillar herniation
Emesis	Transtentorial herniation
Photophobia	Midbrain sagging
Blurry vision	Convexity subdural hematoma



Figure 1: (A) Pre-op contrast enhanced thoracic T1 and T2 weighted MRI of the T3 intradural extra medullary mass. (B) Post resection contrast enhanced (C) T1 and (D) T2 weighted MRI showing gross total resection

sagging.^[1-7] Komotar, *et al.*,^[8] described the radiologic findings deemed important in the diagnosis of brain sag as well as clinical signs of transtentorial herniation including CT head revealing effacement of the basal cisterns with an oblong brainstem, and improvement of symptoms after placement of the patient in the Trendelenburg position. Although CSF hypovolemia is known to lead to intracranial hypotension, there have been reports of patients with spontaneous intracranial hypotension syndrome with normal CSF pressures.^[1,9] Brain sagging leading to poor neurologic exams in aneurysmal subarachnoid hemorrhage patients has been well described in patients who underwent preoperative placement of lumbar drains leading to significant drainage of CSF intraoperatively.^[8] Factors such as longer surgery time and global cerebral edema on admission cranial imaging are risk factors for brain sag after aneurysm surgery.^[8] There have also been reports of brain sag after subdural hematoma evacuation.^[6] Similar to the case presented here, patients present with symptoms of transtentorial herniation including lethargy, unilateral blown pupil, and poor motor exam. The importance of recognizing and differentiating CSF hypovolemia from true increased

intracranial pressure (ICP) cannot be overemphasized as the management algorithms are vastly different.

Patients with brain sag, or CSF hypovolemia, will improve with placement of the patient in the Trendelenburg position. Our patient presented here arrived overnight with a very poor neurologic exam. The patient's symptoms were not immediately recognized as relating to CSF hypovolemia, and the patient was initially treated for refractory high ICPs. When the patient did not improve with these interventions, the consideration for occult CSF leak was discussed, and the patient was immediately placed in Trendelenburg position with resulting improvement in the neurologic exam within minutes. Unlike most reports of brain sag/CSF hypovolemia presenting 1–4 days postoperatively, the patient described here presented in an unusually delayed manner 10 days postoperatively. We attribute this delayed presentation to a slower flow CSF leak. Given this patient's history being confounded by neurofibromatosis and multiple significant intracranial lesions, the presentation was a red herring for the patient's true pathology. Ultimately an ICP monitor was placed and confirmed normal ICPs.

Furthermore, given the iatrogenic vertebral artery injury, we were able to uniquely obtain a cerebral angiogram of the patient's brain while in this condition. The angiographic findings of brain sag have been well described in the literature. Alaraj, *et al.*,^[2] described five patients who presented in extremis from brain sag after clipping of the patient's aneurysm with the preoperative placement of a lumbar drain. These patients were described as having downward displacement of the basilar artery that was severe enough to cause basilar kinking, coined the "cobra sign" on cerebral angiogram. These findings were described as similar to those seen in transtentorial herniation with the exception of the presence of a mass lesion.

If unrecognized, brain sag can theoretically lead to worsening kinking of the basilar artery and posterior circulation vasculature with resulting brain stem ischemia and permanent neurologic deficits. If recognized and treated, Komotar, *et al.*^[8,10] showed that there is no difference in morbidity or mortality at discharge or at 3-month follow-up. Low ICPs and improvement in our patient's exam in Trendelenburg position verified the

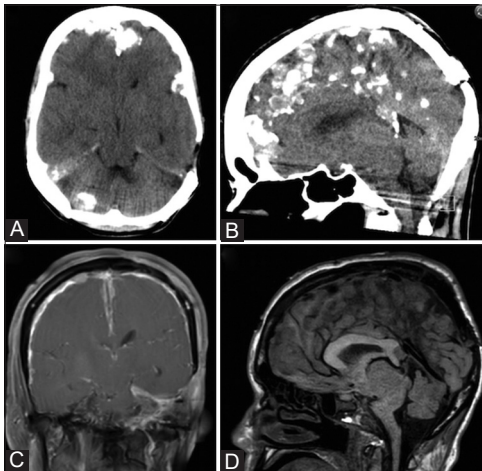


Figure 2: CT head without contrast (A) axial and (B) sagittal slices demonstrating downward herniation with effaced cisterns concerning for acute intracranial hypotension. MRI brain (C) contrasted T1 coronal slice demonstrating diffuse pachymeningeal enhancement, and (D) non-contrast T1 sagittal slice again demonstrating downward herniation with significant deformation of the brain stem. Imaging findings support concern for intracranial hypotension contributing to clinical presentation

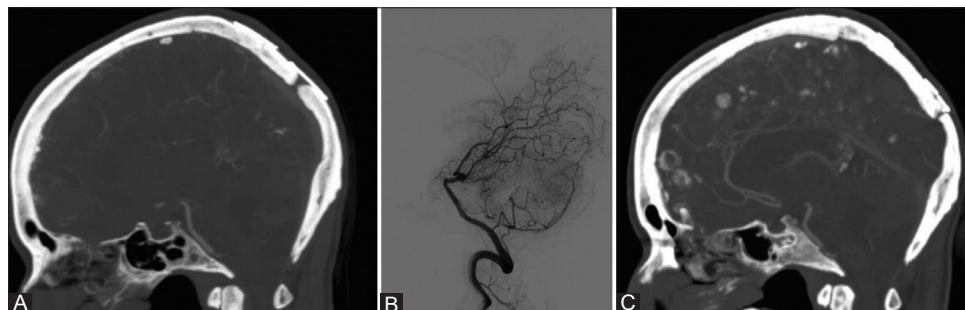


Figure 3: CT angiography (CTA) with (A) cerebrospinal fluid hypotension and (C) after cerebrospinal fluid leak repair. (B) Diagnostic cerebral angiogram (DCA) demonstrating profound basilar kinking with concurrent cerebrospinal fluid hypotension

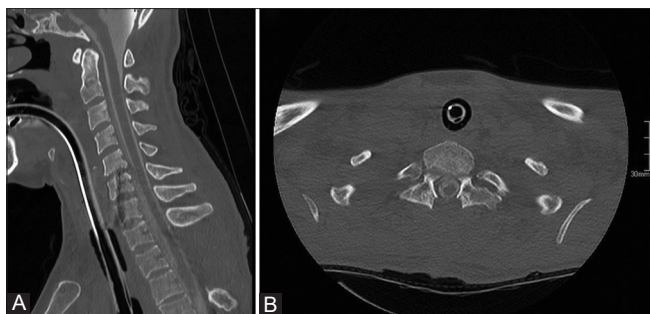


Figure 4: CT myelogram of the cervical and upper thoracic spine (A) sagittal and (B) axial slices revealing ventral contrast extravasation concerning for occult durotomy and cerebrospinal fluid leak

diagnosis of intracranial hypotension/brain sag. Once the diagnosis was made, steps were obtained to identify and treat the offending culprit, ultimately an occult CSF leak after thoracic intradural surgery.

Conclusion

Mild intracranial hypotension continues to be a common finding in patients who undergo procedures that require CSF diversion or CSF loss. Symptoms are easily recognizable including classic postural headaches. However, more severe cases of CSF hypovolemia can be misdiagnosed as intracranial hypertension given overlapping presenting symptoms that can act as a red herring. It is of paramount importance for the clinician to be aware of this diagnostic conundrum and evaluate both potential diagnoses fully as the treatment algorithms are vastly different.

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Conflicts of interest

There are no conflicts of interest.

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