Pathophysiological basis of radiological findings in cerebrospinal fluid hypovolemia- a case report and review of the literature

S Vadi, D Kumar

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Abstract

Post-dural puncture cerebrospinal fluid (CSF) leak tugs on the intracranial pain-sensitive structures leading to headache. This is characteristically postural in nature. MRI of the brain and cervical spine demonstrate dural enhancement, with low lying cerebellar tonsils, and venous distention.

CASE

A 20 years old pre-morbidly healthy lady presented to the ER with headache. This throbbing headache located over the frontal and temporal regions bilaterally was of 8-10/10 severity. It was aggravated with walking and standing, and relieved on lying down. Headache exacerbations were associated with blurring of vision, nausea, and vomiting. She denied any weakness or numbness. She denied any fever or chills. Her headache commenced on post-partum day 1. This was felt to be secondary to having received epidural anesthesia to aid her labor. An epidural blood patch for presumed CSF leak was placed by Anesthesia 1-2 days after delivery. However, this did not relieve her headache. Twenty days post-partum, an MRI Brain was performed, in view of persistent headache. This revealed thrombosis in the superior sagittal venous system, and of bilateral frontal cortical veins with extension into the right transverse sinus. Treatment consisted of Coumadin, for superior sagittal venous thrombosis, and Tramadol for headache.

She presented 3 months later for protracted headaches having missed her interim follow-up appointments. Other than mild difficulty with heel toe walking, her neurological examination was non-revealing. Indium 111 radioisotope cisternography was then performed to locate CSF leak, but this was negative at 24 and 48 hours. CT scan with contrast of the neural axis was unremarkable. With the MRI findings of thrombosis of superior sagittal sinus extending into transverse sinus, positive delta sign, pachymeningeal enhancement, subdural fluid collection, pituitary hyperemia, loss of pre-chiasmatic and pre-pontine cisterns, flattening of optic chiasm, and inferior migration of cerebellar tonsils, and a small ventricular size, a final diagnosis of low CSF pressure syndrome was established [Images 1-3], and treatment commenced with intravenous dexamethasone, and intravenous hydration. She was placed on complete bed rest. A second epidural blood patch was also placed. This partially relieved her headaches. Unfortunately the patient is lost to follow-up.

Figure 1

Image 1: Coronal image of MRI venogram demonstrating the venous distention sign (white arrow)
Figure 2
Image 2: Post-contrast, coronal MRI image of brain demonstrating: a) pachymeningeal enhancement, b) delta sign (filling defect suggestive of superior sagittal venous thrombosis), c) subdural hygroma, d) pituitary hyperemia, and cerebellar tonsillar herniation (downward arrows)

Figure 3
Image 3: Pre-contrast, midline sagittal MRI image of brain displaying a) loss of pre-pontine cistern, b) loss of pre-chiasmatic cistern, and c) downward herniation of the cerebellar tonsils (7mm) without evidence of syringomyelia (acquired Chiari I malformation)

DISCUSSION
Clino-pathological correlation of cranial mechanics can be explained on the basis of Monro Kellie hypothesis. 1 Cranium is a rigid and fixed structure. The sum of volumes of CSF, brain, and blood is constant. Per hypothesis, any reduction in CSF volume should lead to a compensatory increase in the brain and blood. Volume of brain being constant, this sum would thus be kept constant by a compensatory increase in the blood volume. Resultant dilatation of the venous system, a pain-sensitive intracranial structure leads to headache. Reduction of cerebral venous outflow during Valsalva maneuver leads to intracranial venous engorgement, and aggravates the headaches even in the supine posture. 2

A pressure gradient is created between CSF and the vasculature as a result of alterations in CSF volume. CSF hypovolemia results in the observed symptoms. 3 Spectrum of clinical presentation varies from the commonly presenting symptom of postural headache, to chronic daily headaches, acephalgia, or cranial nerve palsies (V, IX, X, or upper three cervical nerves), vomiting, photophobia, or confusion. 2 Normal CSF opening pressures vary from 60-200 mmHg. CSF opening pressures in CSF hypovolemia can be normal to low 4 , or even subatmospheric. 2 Rather than relying on opening pressures alone to help make the diagnosis, MRI is a better modality that helps diagnose the syndrome of CSF hypovolemia. Below is a flow-chart that depicts the correlation between pathophysiology, and radiological findings. 1,5,6

Identification of the site of dural leak by radionuclide cisternography would ease management issues by a targeted placement of an epidural blood patch. However, the site of leak is not always evident. Sometimes successive placement of several patches is needed before any significant response is experienced. 7 Several treatment options are available, such as bed rest, analgesics, caffeine, intravenous hydration, epidural blood patch, and parenteral corticosteroids. Corticosteroids are believed to help by their anti-inflammatory response, reducing vascular leakage, and fluid retentive properties. 2
CONCLUSION

CSF hypovolemia following a dural leak commonly manifests with postural headache. Knowledge of the MRI findings which have a pathophysiological basis will aid internists in the early diagnosis of CSF hypovolemia given an appropriate clinical setting, and facilitate the institution of relevant therapy.

References

Author Information

Sonali Vadi, MD, FNB
Department of Internal Medicine

Dharmendra Kumar, MD
Department of Radiological Imaging