Symptomatic intrathecal hypertension

Awss Zidan, MD

Deborah Young Bradshaw, MD

Practical Implications

Consider intrathecal hypertension as an etiology of radiating back pain in patients who have complete spinal block on neuroimaging or who report symptomatic relief after lumbar puncture.

The Monro-Kellie doctrine states that since the cranium is an incompressible space, the sum of its contents must remain constant. An increase in one component must be accompanied by a decrease in another component, and even a small increase in the intracranial contents can result in a marked increase in intracranial pressure, causing pain and neurologic dysfunction.¹ It is interesting that this concept has not been widely applied to the spinal canal, which is also an incompressible space. Theoretically, an increase in the intraspinal contents should result in increased pressure within the spinal canal with the potential to cause damage to the spinal cord and nerve roots. Here we describe a patient with complete spinal block due to arachnoiditis, elevated CSF pressure, and back pain whose symptoms resolved with CSF removal.

Case report

A 37-year-old man with a history of recurrent pneumonia and meningitis was evaluated for severe back pain. MRI demonstrated pachymeningitis in the midthoracic region. Thoracic dural biopsy demonstrated nongranulomatous chronic inflammation consistent with pachymeningitis. Serum immune studies revealed deficiency of IgG subclasses 1, 2, and 4 with normal IgM and IgA, compatible with isolated IgG deficiency. IV immunoglobulin therapy was initiated. Several months later, he presented with worsening back pain that was severe and sharp and radiated from the thorax to the cervical and lumbar regions with an electrical quality. There were bouts of superimposed excruciating pressure-like pain that prompted hospitalization. He also reported numbness and weakness in the legs. The neurologic examination showed antigravity power limited by pain. Lower extremity reflexes were depressed and Babinski signs were absent. Sensation was reduced to all modalities below T10. Thoracic spinal MRI showed adhesion of the cord anteriorly and linear enhancement with nodularity of the anterior and posterior margins of the thecal sac at T4 without nerve root enhancement or cord compression (figure, A). Lumbar puncture revealed an opening pressure of 32 cm H₂O. Twenty mL of CSF was removed, resulting in a closing pressure of 14 cm H₂O. Fluid analysis showed a white blood cell count of 75/hpf, red blood cell count of 0, and protein of 927 mg/dL. After the procedure, the patient experienced immediate improvement of the pain. Herpes simplex virus (HSV)-2 antigen was detected by PCR in the CSF. Treatment with IV acyclovir was initiated. However, the pain gradually recurred and became intolerable. CT myelogram showed complete obstruction of CSF flow at the T6 level despite 10 minutes in a 40° head-down position (figure, B).

Four months later, a temporary lumbar drain was placed. CSF showed an elevated opening pressure of 34 cm H₂O, protein of 100 mg/dL, and no evidence of HSV-2. Repeat MRI

Department of Neurology, SUNY Upstate Medical University, Syracuse, NY.

Funding information and disclosures are provided at the end of the article. Full disclosure form information provided by the authors is available with the **full text of this article** at Neurology.org/cp.

Correspondence to: zidana@upstate.edu

Figure Imaging of thoracic spine

(A) Sagittal MRI T2 shows dural adhesions (arrow) with accompanying cord edema and (B) sagittal CT myelogram shows abrupt cessation of contrast flow at a midthoracic level (arrow) implying a spinal block.

showed a newly developing syrinx in the area of adhesion. Lumbar drain provided substantial relief of the back pain, weakness, and numbness. A lumboperitoneal shunt was later placed and the patient reported resolution of the severest components of his pain.

DISCUSSION

The concept of intrathecal hypertension has been introduced by others. It was hypothesized to result in cauda equina flattening in a patient who had metastatic spinal lesions well above the cauda equina.² In a case series, 5 patients with ankylosing spondylitis who developed chronic arachnoiditis and dural fibrosis resulting in dural sac dilations had significant improvement of their pain, urinary function, and sensation after lumboperitoneal shunts.³

Our patient experienced severe back pain presumably due in part to elevated spinal CSF pressure. CSF dynamics are not completely understood. CSF is secreted by the choroid plexus (40%–70%) and pia mater (30%–60%).⁴ Resorption occurs mainly through the arachnoid villi. However, it has been postulated based on radionuclide studies that the spinal structures are responsible for ~10 mL/h of CSF absorption.^{5,6} Elevated CSF pressure in our patient was likely due to impaired clearance caused by the tight block at the T6 level. There may also have been impaired resorption due to scarring of spinal meningeal villi caused by recurrent meningitis. In addition, markedly increased protein may cause elevated CSF pressure by creating an osmotic gradient that draws in interstitial fluid. The precise role of these factors is unknown without detailed fluid dynamic studies.

It is possible that the back pain was caused by the arachnoiditis itself. However, pain repeatedly remitted with CSF removal, suggesting that the source of the symptoms was the pressurized intrathecal space.

Intrathecal hypertension should be considered in patients with spinal pain and radiculopathy in the absence of compressive spinal lesions. The response to drainage and CSF flow studies may provide further clues to the diagnosis and aid in the treatment.

REFERENCES

1. Mokri B. The Monro-Kellie hypothesis: applications in CSF volume depletion. *Neurology* 2001;56:1746–1748.
2. Lefresne S, Fairchild A, Bistriz A, Venner P, Yee D. A case of indirect cauda equina syndrome from metastatic prostate cancer. *Can Urol Assoc J* 2009;3:E31–E35.
3. Ea H-K, Lioté F, Lot G, Bardin T. Cauda equina syndrome in ankylosing spondylitis: successful treatment with lumboperitoneal shunting. *Spine (Phila Pa 1976)* 2010;35:E1423–E1429.
4. Artru AA. Cerebrospinal blood flow: physiology and pathologic conditions. In: Sperry RJ, Johnson JO, Stanley TH, editors. *Anesthesia and the Central Nervous System*. Dordrecht, the Netherlands: Springer Netherlands; 1993:27–49.
5. Kido D, Gomez D, Pavese A Jr, Potts D. Human spinal arachnoid villi and granulations. *Neuroradiology* 1976;11:221–228.
6. Edsbacke M, Tisell M, Jacobsson L, Wikkelso C. Spinal CSF absorption in healthy individuals. *Am J Physiol Regul Integr Comp Physiol* 2004;287:R1450–R1455.

AUTHOR CONTRIBUTIONS

A. Zidan drafted the content of the manuscript, including medical writing. D.Y. Bradshaw revised the content.

STUDY FUNDING

No targeted funding reported.

DISCLOSURES

A. Zidan reports no disclosures. D.Y. Bradshaw receives research support from Cytokinetics, Inc. Full disclosure form information provided by the authors is available with the **full text of this article at Neurology.org/cp**.

Practical Implications

Neurology® *Clinical Practice* is committed to providing clinical insights that are helpful to neurologists in everyday practice. Each Full Case will include a “Practical Implications” statement, a pearl of wisdom for the practicing clinician.