Spontaneous intracranial hypotension associated with transdural thoracic osteophyte reversed by primary dural repair

Case report

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Spontaneous intracranial hypotension (SIH) is an increasingly recognized syndrome associated with a specific set of clinical and imaging findings; however, determining the site of spinal cerebrospinal fluid (CSF) leakage in these patients is often difficult, and indications for surgical intervention need to be better defined. The authors report on a 55-year-old woman who presented with posture-related headache, disorientation, and memory impairment. Imaging features were consistent with SIH. Computerized tomography myelography demonstrated a large T2–3 anterior transdural osteophyte associated with a CSF fistula. After an unsuccessful trial of conservative therapy, the patient underwent median sternotomy, T2–3 discectomy and removal of osteophyte, which allowed adequate exposure for primary dural repair. Postoperatively, there was immediate and prolonged resolution of all of her symptoms. This case of SIH was caused by transdural penetration by an anterior osteophyte and CSF leakage in the upper thoracic spine, which was treated effectively by anterior exposure and primary dural repair. Aggressive surgical intervention may be required to treat upper thoracic CSF leaks refractory to other measures.

KEY WORDS • cerebrospinal fluid leak • intracranial hypotension • thoracotomy • median sternotomy • osteophyte • blood patch

Case Report

Presentation and Examination. This 55-year-old woman presented with a 6-week history of severe holocranial headache, which worsened when she was upright and improved when she was recumbent. She also experienced aural fullness, memory difficulties, and disorientation. On admission brain MR imaging demonstrated the classic imaging finding of SIH (Fig. 1A). A large-volume (30-ml) lumbar epidural blood patch was placed, but it only provided temporary symptomatic improvement. Postmyelography CT scanning demonstrated a CSF fistula centered in the anterior upper thoracic spine associated with a large ventral osteophyte (Fig. 2). An MR image of the spine was not obtained.

Operation. The patient was taken to the operating room where she underwent a median sternotomy and anterior T2–3 discectomy. This enabled visualization of the large ventral T2–3 osteophyte. The osteophyte was debrided using a high-speed drill. After completion of the osteophytectomy, the dural defect became apparent, although a small amount of arachnoid had herniated into the space.

Abbreviations used in this paper: CSF = cerebrospinal fluid; CT = computerized tomography; MR = magnetic resonance; SIH = spontaneous intracranial hypotension.
and had sealed off the leak. Gentle mechanical displacement of the arachnoid led to prompt egress of CSF under medium pressure. The linear dural defect resulting from the osteophyte was repaired by placing two nonabsorbable vertical mattress sutures and a small piece of interposed muscle, and fibrin glue was applied. No fusion procedure was performed.

Postoperative Course. Postoperatively, the patient was maintained on bed rest for 48 hours. Her headache, aural fullness, and neurological symptoms resolved completely within 24 hours of surgery. She was mobilized on postoperative Day 2 without difficulty. Repeated MR imaging on postoperative Day 4 demonstrated partial reversal of brain sag and other imaging manifestations of SIH (Fig. 1B). She was discharged on postoperative Day 7. Since discharge (> 1 year), she has suffered no recurrence of orthostatic headache or other neurological symptoms.

Discussion

History and Origin of the System

Schaltenbrand51,52 was the first to describe a condition of spontaneously low or even negative CSF pressures with orthostatic headaches, which he termed primary spontaneous intracranial hypotension or “essential aliquorrhea.” Subsequent investigators have further defined this syndrome of intracranial hypotension or hypoliquorrhea.4,27,28,72

It was originally controversial whether the low CSF volume state resulted from decreased CSF production, increased CSF resorption, or CSF leakage.21,26,35,51,52,69 In 1992, Rando and Fishman48 hypothesized that the mechanism of CSF leakage involved spontaneous rupture of a spinal arachnoid (Tarlov) cyst. Based on this report and on subsequent series, it is now clear that spinal CSF leaks are the most common cause of SIH.53,57 Most CSF leaks are spontaneous and probably result from weakness of the spinal meninges,32,40,57,59 but generalized connective tissue disorders (for example, Marfan syndrome),12,14,19,34,56,57,63 local trauma (for example, osteophyte piercing dura, as in our case),12,53,67 so-called nude nerve root syndrome,53 or iatrogenic overdrainage of CSF50 may contribute in some cases.

Clinical Manifestations

Spontaneous intracranial hypotension is nearly always41,60 characterized by orthostatic headache. The onset of headache is usually gradual or subacute, but patients may present with so-called thunderclap headache.62 The headache may be holocranial or localized to the frontal or occipital regions. It is probably caused by a loss of CSF volume and subsequent traction on the brain-sensitized intracranial dura due to downward brain displacement.

Associated findings are quite varied and may include neck pain or stiffness, interscapular pain, nausea, vomiting, and cranial neuropathies.4,7,17,30,34,37,57 Distinct cranial neuropathies in association with SIH include abducence (horizontal diplopia), oculomotor, trigeminal (facial pain/paresthesias), vestibulocochlear (dizziness and hearing changes, phonophobia), and optic (blurred vision, photophobia, and transient visual obscurations).15,24,37,41,45,57,68 Occasionally SIH may present with predominantly motor manifestations.41,42 In some cases, severe brain sag may lead to signs of central (transient) herniation and patients may suffer stupor due to dienchephalic compression.3,6,44,57

Diagnosis of SIH

Neuroradiological evaluation is essential for diagnosis and for accurate localization of the site of the CSF leak.17,22,35,43,49 The main imaging findings of SIH are brought about because the CSF volume has been reduced, which, according to the Monro–Kellie rule, induces venous engorgement;17,31 the imaging characteristics thus include the following: 1) pachymeningeal enhancement; 2) enlargement of the dural venous sinuses; and 3) descent of the brain into the posterior fossa (also called brain sag or pseudo-Chiari malformation).1,7,17,30,34,35,37,43,49,61,65 Other associated features include venous engorgement of the pituitary gland,1,31,38 chiasmatic drooping on the dorsum sellae, collapse of the superior ophthalmic veins,4 and spontaneous subdural hygromas. Meningeal biopsy sampling is unnecessary for establishing a diagnosis.4,16,18,25,36

Localization of the Site of a CSF Fistula

Localization of the site of a CSF fistula can be difficult
and requires expert neuroradiological/neuroimaging consultation. Most commonly, spontaneous CSF leaks occur in the lower cervical or upper thoracic region. The initial study of choice is fat-suppressed fast–spin echo MR imaging of the spine,\textsuperscript{10} which may demonstrate an extradural collection of CSF or a prominent perineural cyst. Supportive evidence includes CSF hygroma, epidural venous engorgement and dural enhancement, and paraspinal fluid collections behind the C1–2 segment.\textsuperscript{10} Spinal MR imaging, however, is often unable to reveal the actual site of CSF leakage.\textsuperscript{37,46,48,57} Furthermore, the location of an epidural collection does not necessarily correlate with the site of the CSF fistula.\textsuperscript{10} Postmyelography CT scanning is currently the study of choice to demonstrate extrathecal contrast accumulation or define anatomical abnormalities such as meningeal diverticula;\textsuperscript{10,11,57} however, it still may not demonstrate the site of the leak. In such cases, it may be necessary to perform dynamic myelography or dynamic postmyelography CT scanning in which contrast is instilled in the scanner and repetitive CT scans are obtained.

Radionuclide cisternography has also been used to evaluate spinal CSF leaks,\textsuperscript{9,26,39,48} but it is less sensitive than postmyelography CT scanning and has poorer spatial resolution.\textsuperscript{57} It is primarily applied in cases of cranial CSF leaks. The newer modality of MR myelography may prove useful.\textsuperscript{29,47}

Nonsurgical Treatment

In cases of SIH the patient’s headache usually resolves spontaneously after bed rest and provision of fluids. Pharmacological management is largely ineffective.\textsuperscript{18,23} In cases involving intractable persistent headache or other signs and symptoms of SIH, the initial treatment of choice is a large-volume (> 20-ml) epidural blood patch,\textsuperscript{5,11,20,37,57} following which the patient should be placed in a head-down reverse Trendelenberg position.\textsuperscript{2,66} Relief of symptoms is often immediate, secondary to thecal compression and increased CSF pressure, but symptoms may recur if leakage persists. Occasionally, several large-volume blood patches are necessary for permanent benefit.\textsuperscript{11,56} Accurate localization of the leakage by CT myelography can allow an epidural blood patch to be placed at the site of the leak. If the patient suffers mental status deterioration due to brain sag,\textsuperscript{3,6,44,57} lumbar intrathecal infusion with Elliott B solution (artificial CSF) may be required to arrest or reverse impending central (transient) herniation.\textsuperscript{6}

Surgical Management of Spinal CSF Leaks

Surgical exploration and repair have been successful in cases refractory to conservative management.\textsuperscript{13,44,55,58,59,67} In a retrospective analysis of 10 patients treated surgically for spontaneous spinal CSF leaks between 1992 and 1997, the authors found that although symptomatic relief occurred postoperatively in all cases, preoperative diagnostic imaging findings did not always correlate with intraoperative findings.\textsuperscript{58} During their mean follow-up period of 19 months, there was no recurrence of symptoms in any case.

Only a few previous cases of osteophyte-related SIH have been reported.\textsuperscript{13,58,67} For example, CSF leakage associated with cervical bone spurs has been reported.\textsuperscript{13,58,67} In one case, CT myelography demonstrated a high-flow CSF leak anterior to C5–6; this was confirmed on radionuclide cisternography. A midline bone spur protruding through the posterior longitudinal ligament into the thecal sac was successfully extracted via an anterior approach, and primary dural closure was then performed.\textsuperscript{57} Postoperatively, MR imaging demonstrated resolution of SIH.\textsuperscript{67}

Eross, et al.,\textsuperscript{13} reported on three patients with SIH caused by osseous pathological entities of the cervical spine. Their first patient had midline anterior bone spurs adjoining the C5–6 interface and CT myelography evidence of a leak starting at C-6 with contrast pooling to T-11. Efforts at conservative management had failed, and the patient underwent C-5 discectomy and partial C5–6 corpectomy. Intraoperatively, the C-5 bone spur was mobilized, which provided exposure but also led to enlargement of a dural tear.\textsuperscript{13,67} Multiple distinct procedures were performed to repair the dural defect, including further spur

![Fig. 2. A: Axial CT myelogram obtained at T2–3 demonstrating a 5-mm bone osteophyte penetrating the anterior dura (arrow) and extradural contrast collection (arrowhead). B: Sagittal reconstruction of CT myelogram revealing an osteophyte arising from superior endplate of T-3 (arrow).](image-url)
Thoracic dural cerebrospinal fluid leak repair
debridement, C6–7 corpectomy, and placement of a lumbo-peritoneal shunt. Because primary dural closure was not successful, this patient continued to be incapacitated by orthostatic headaches 24 months after onset.

Unlike the variety of case reports of cervical disease associated with CSF leakage and SIH, to our knowledge there is only one documented case of SIH secondary to a thoracic bone spur. In that case a calcified T7–8 disc indented the thecal sac, and myelography demonstrated pooling of extradural contrast at the same level. Primary surgical repair of the CSF fistula and removal of the herniated disc were considered, but instead the authors opted for localized T7–8 epidural blood patch. Despite clinical improvement, recurrent or increased size of a right subcortical collection prompted burr hole drainage, which was repeated 1 week later following recurrence of headache. The authors report that ultimately the patient became asymptomatic despite the fact that a direct surgical effort at primary spinal dural CSF leak repair was not performed.

Conclusions

In summary, we have detailed a case of symptomatic SIH due to an upper thoracic CSF leak resulting from a large transdural anterior osteophyte. An appropriate trial of conservative management was ineffective. Median sternotomy, anterior T2–3 discectomy, and osteophyte removal exposed a high-flow CSF fistula, and primary dural repair led to rapid reversal of symptoms.

To our knowledge, this is the first case report of median sternotomy for operative repair of a thoracic CSF leak causing SIH. The results of this case, together with our review of the literature, demonstrate that aggressive operative exposure may be necessary for primary dural repair in cases of SIH refractory to conservative therapy. Because most identified sites of spontaneous CSF leakage are in the cervical or thoracic region, at times extensive operative approaches may be necessary. We encourage all neurosurgeons to become familiar with the diagnosis and treatment of SIH as well as the potential for operative intervention to effect a cure.

References

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