Because of cerebrospinal fluid (CSF) leak, orthostatic headaches sometimes follow neuraxial blockade or diagnostic lumbar puncture. Etiology and treatment are straightforward in those settings, since a site of dural disruption is known.1,2 With patient consent, we present a case in which intracranial hypotension resulted from dural puncture by a vertebral osteophytic spur. This case scenario illustrates the diagnostic dilemma of spontaneous intracranial hypotension (SIH), the role of imaging studies, and the value of empiric lumbar epidural blood patch (LEBP).

Case Report

A 36-yr-old woman came to our emergency department with a 1-week history of headache. She described a minor event 2 weeks previously, when she fell forward onto her outstretched hands. Although this fall caused some minor shoulder and neck pain that quickly resolved, she did not develop a headache until a week later. The pain was located bilaterally in the frontal and temporal areas as well as the neck. It did not initially interfere with her activities of daily living, but it became progressively more severe over 2 to 3 days. The headache also became postural, and she became unable to sit up or walk. She experienced nausea with attempted sitting, but she denied photophobia, neck stiffness, fever, or chills.

She had been evaluated at another hospital, where noncontrast head computed tomography (CT) scan, motion-degraded brain magnetic resonance imaging (MRI), and magnetic resonance angiogram were reported as normal. Lumbar puncture revealed an opening pressure of 9 cm H2O and was otherwise unremarkable. She went home but came to us several days later.

Magnetic resonance imaging of the cervical, thoracic, and lumbar spine without contrast demonstrated mild degenerative changes. Otherwise, the study was initially considered normal. After conservative treatments and medication failed to relieve her pain, the patient underwent an epidural blood patch via the L4 to L5 interspace with injection of 17 ml of autologous blood. She reported immediate and complete relief of her headache and returned home the same day. Her headache resumed at its original intensity later that evening and she was readmitted, prompting further diagnostic evaluation. MRI of the brain without and with contrast demonstrated diffuse meningeal thickening and enhancement, distension of dural venous sinuses, enlargement of the pituitary gland, and mild brain descent (brain sagging), findings consistent with intracranial hypotension (fig. 1). CT myelogram confirmed a suspected spinal CSF leak and identified its location (fig. 2). Contrast extravasated from the subarachnoid space into the anterior epidural space adjacent to a posterior osteophytic bone spur at C7 to T1. In association with the brain MRI and CT myelogram findings, close reexamination of the initial noncontrast spine MRI revealed distension of the cervical epidural veins and epidural fluid collection at the cervicothoracic junction, consistent with a CSF leak in that region (fig. 3).

The imaging results prompted surgical repair. Under general anesthesia, via an anterior approach, she underwent C7 and T1 partial corpectomy, C7 to T1 discectomy, excision of the bone spur, and decompression of the anterior epidural space. There was an 8-mm-long vertical rent in the dura opposite the bone spur, and CSF was actively leaking. After microsurgical primary repair of the dural tear, spinal arthrodesis was performed using a structural allograft with anterior plate and screws. She returned home on postoperative day 3. At a 2-week visit, she was experiencing only mild limitation in lateral movement of the neck, which had resolved when she returned for a 9-week postoperative visit. She experienced no further headaches.

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Spontaneous Intracranial Hypotension

**Discussion**

**Etiology**

A constant volume of CSF, approximately 150 ml that is replaced three to four times daily through the choroid plexus system, serves to cushion the brain and spinal cord. SIH results when the loss of CSF due to a dural tear outpaces the ability of the plexus to replace it. An altered distribution of craniospinal compliance is also a likely important factor explaining orthostatic intracranial hypotension and headache in spinal CSF leaks. Minor trauma, such as the fall reported by our patient, is reported in about a third of patients.

**Epidemiology**

The incidence of SIH has been estimated at 5 of 100,000 person-years. The peak incidence of SIH is in the fourth and fifth decades although it also occurs in children and older adults. Women are affected twice as often as men.

**Diagnosis**

Since 1938, diverse presentations have been recognized. Our patient presented with the most classic clinical manifestation of SIH, postural headache, likely related to dilation of pain-sensitive intracranial veins or to brain descent with associated traction. However, the headache may be gradual in onset, thunderclap in onset, less positional over time,

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Fig. 1. Brain magnetic resonance findings consistent with intracranial hypotension. (A) Axial T2-weighted fluid-attenuated inversion recovery image demonstrating pachymeningeal thickening (arrow) and distension of the superior sagittal sinus (arrowhead). Axial (B) and coronal (C) T1-weighted images after gadolinium contrast demonstrating diffuse smooth pachymeningeal enhancement (arrow) and distension of the superior sagittal and transverse sinuses (arrowheads). (D) Sagittal T1-weighted image demonstrating signs of brain descent (sagging): effacement of the interpeduncular cistern (dotted ellipse), enlargement of the pituitary gland (dotted arrow), depression of the corpus callosum (curved arrow), inferior pointing of the splenium of the corpus callosum (asterisk).
more positional over time,\(^\text{11}\) nonpositional,\(^\text{12}\) paradoxical (worse when lying down),\(^\text{13}\) exertional,\(^\text{14}\) intermittent,\(^\text{11}\) or even absent.\(^\text{15}\) Our patient experienced neck pain and nausea, which occur in about half of patients with sIH.\(^\text{5}\) Other signs of meningeal irritation include neck stiffness, vomiting, photophobia, and phonophobia.\(^\text{5,11}\) Dysfunction of cranial nerves, the pituitary gland, or the brainstem may reflect mechanical deformation due to brain descent. For example, altered hearing, dizziness, and vertigo are common, caused by tension on the vestibulocochlear nerve, or by transmission of abnormally low CSF pressure to the perilymph.\(^\text{16}\) There can be blurred vision, field defects, diplopia,\(^\text{17}\) facial pain or numbness,\(^\text{18}\) and altered taste.\(^\text{5,19}\) Stretching of the pituitary stalk may cause hyperprolactinemia and galactorrhea.\(^\text{20}\) In severe cases of brain descent, deformation may result in ataxia, Parkinsonism, bulbar weakness,\(^\text{21}\) and cognitive changes.\(^\text{22,23}\) Indeed, fluid leaks through dural defects can precipitate brain herniation.\(^\text{24}\) Symptoms and signs referable to the spine can occur, including back pain and radiculopathy. In 338 patients with SIH, 6% had a myelopathy or radiculopathy, typically presenting at time of headache onset.\(^\text{25}\) Symptoms were attributable to an epidural CSF collection compressing the spinal cord or nerve roots.

Various imaging techniques help to diagnose SIH and guide treatment. MRI of the brain and spine identify signs of SIH (figs. 1 and 3).\(^\text{5}\) CT myelography (CTM) and intrathecal gadolinium magnetic resonance myelography (Gd-MRM) localize spinal CSF leaks (fig. 2). Standard CTM is best to localize suspected slow leaks, which remain confined over at most two vertebral levels between the time of fluoroscopically guided contrast instillation and CT acquisition.\(^\text{26}\) Dynamic CTM is best to localize suspected fast leaks, which would otherwise spread over many levels, obscuring the actual leakage site.\(^\text{26}\) In dynamic CTM, contrast is instilled incrementally via CT-guided lumbar puncture, followed by immediate scanning after each injection to localize the site of earliest contrast leakage. Since slow leaks tend to be associated with a small extra-arachnoid fluid collection, while fast leaks tend to be associated with a large extra-arachnoid fluid collection, spine MRI is used to choose between the standard and dynamic CTM techniques.\(^\text{26}\) Gd-MRM has higher sensitivity than CTM for small, slow leaks, but is more complex to perform.\(^\text{27,28}\) CTM and Gd-MRM both have higher sensitivity and spatial resolution than radionuclide cisternography.\(^\text{28,29}\) Radionuclide cisternography is increasingly considered obsolete.\(^\text{28}\) Despite their increased sensitivity, CTM and Gd-MRM fail to identify a CSF leak in close to half of subjects with suspected SIH.\(^\text{26,27}\) The diagnosis of SIH is based on a combination of clinical and imaging findings. The International Headache Society offered consensus.
criteria for diagnosis of SIH in 2004 (table 1). However, these criteria may be too restrictive.\textsuperscript{31,32}

**Conservative Treatment**

Although evidence is anecdotal, many patients respond to conservative measures alone, including bed rest, oral hydration, oral caffeine, and an abdominal binder. Other supportive measures have included oral corticosteroids, intravenous caffeine, and theophylline, but their effectiveness has been limited.\textsuperscript{5} Conservative measures may be less successful for SIH than for postdural puncture headache.\textsuperscript{33}

**Epidural Blood Patch**

An LEBP is the time-honored approach for the treatment of postdural puncture headache when conservative treatment fails.\textsuperscript{34} For patients with suspected SIH, based on clinical evidence and positive noninvasive MRI findings, a LEBP can be performed in the absence of invasive imaging studies such as CTM, gadolinium myelography, or radionuclide cisternography.\textsuperscript{7,23} Many patients with intracranial hypotension experience symptom relief within minutes after the LEBP.\textsuperscript{35,36} This early effect is likely explained by immediate changes in craniospinal CSF mechanics.\textsuperscript{35,36} Sustained relief presumably occurs due to tamponade and then sealing of the dural defect by the LEBP, followed by gradual restoration of CSF volume.\textsuperscript{35,37-40} The efficacy and time course of the latent phase depend on the spread of the autologous blood

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**Table 1. Diagnostic Criteria for Headache Attributed to Spontaneous (or Idiopathic) Low Fluid Pressure, from the International Headache Society**\textsuperscript{30}

<table>
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<th>A. Diffuse and/or dull headache that worsens within 15 min after sitting or standing, with at least one of the following and fulfilling criterion D:</th>
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<tr>
<td>1. Neck stiffness</td>
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<td>2. Tinnitus</td>
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<tr>
<td>3. Hypacusia</td>
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<td>4. Photophobia</td>
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<td>5. Nausea</td>
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**B. At least one of the following:**

1. Evidence of low fluid pressure on magnetic resonance scan (e.g., pachymeningeal enhancement)
2. Evidence of fluid leak on conventional myelography, tomographic myelography, or cisternography
3. Opening pressure <60 mm H\textsubscript{2}O in sitting position

**C. No history of dural puncture or other cause of fistula**

**D. Headache resolves within 72 h after epidural blood patching**
from the lumbar injection site to the site of CSF leakage.\textsuperscript{41–44} Reports of the extent of epidural spread after LeBP vary and may depend on the sensitivity of the imaging techniques used (e.g., MRI,\textsuperscript{42} CT,\textsuperscript{33} or Tc-99m-labeled erythrocytes).\textsuperscript{44} Generally, the extent of epidural spread increases with LeBP volume,\textsuperscript{44} is greater cranially than caudally, and shows considerable individual variability.\textsuperscript{42–44} When one LeBP is ineffective or only transiently effective, subsequent LeBPs may ultimately prove successful. Although efficacy rates of LeBP over 90% have been reported for postdural puncture headache,\textsuperscript{37,45} success rates for SIH range from 53 to 77\% after one or more LeBPs.\textsuperscript{41,46,47} Localization of leakage site by means of CTM or Gd-MRM permits targeted epidural blood patching (EBP). Indeed, targeted cervicothoracic EBP has succeeded in cases when LeBP has failed.\textsuperscript{41,48–50}

In a series of 42 patients with SIH, Ferrante \textit{et al.}\textsuperscript{7} reported complete recovery in all patients after one (90%), two (5%), or three (5%) blind lumbar EBPs using the following protocol: (1) maintain a 30-degree Trendelenburg position 1 h before, during, and 24 h after EBP; (2) premedicate with acetazolamide 250 mg 18 and 6 h before EBP; and (3) refrain from strenuous exercise for 2 weeks after the procedure. Prolonged Trendelenburg position is hypothesized to promote rostral spread of the EBP.\textsuperscript{5} Indeed, spine CT or MRI after the procedure, using iodinated or gadolinium contrast mixed with the autologous blood, showed epidural spread over many levels, from lumbar to cervical levels in 63\% of patients. Acetazolamide, a carbonic anhydrase inhibitor, decreases the rate of CSF production, and is hypothesized to decrease the rate of CSF leakage, promoting formation of a seal, though its benefit in this setting is unproven.\textsuperscript{7}

In some cases where EBP has failed, epidural fibrin glue patching has succeeded when used alone\textsuperscript{51} or mixed with autologous blood.\textsuperscript{52} Fibrin glue has several known risks, but serious complications are rare. These include viral infection and allergic reactions.\textsuperscript{53–55}
Surgery
Surgery is sometimes effective for the small minority of patients who remain refractory to conservative measures, one or more EBPs, and possibly one or more epidural fibrin glue patchings. Preoperative localization of the putative site(s) of CSF leakage by CTM or gadolinium myelography is essential.

In the eight previously reported cases in which an osteophytic spur, with or without superimposed disc herniation, pierced the dura and caused SIH, EBP successfully treated three and partially improved one; like the current case, surgery successfully treated three of the four other cases. Similar to the current case, surgery in these cases usually involves anterior discectomy, resection of the bone spur, and primary repair of the dural rent, sometimes accompanied by interbody fusion or placement of a lumbar drain.

Conclusion
Spontaneous intracranial hypotension is more common than generally realized and poses significant diagnostic and treatment challenges. SIH should be approached with a treatment algorithm similar to the familiar treatments for postdural puncture headache, including blood patch (fig. 4).

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Competing Interests
The authors declare no competing interests.

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