Spon
taneous intracranial hypotension is an important cause of headaches, particularly in young and middle-aged adults. Characteristically, the headache is orthostatic and similar to a post–lumbar puncture headache. Spontaneous spinal CSF leaks can be demonstrated in most patients. Whether CSF leaks at the level of the skull base can cause spontaneous intracranial hypotension remains a matter of controversy. The authors’ aim was to examine the frequency of skull base CSF leaks as the cause of spontaneous intracranial hypotension.

Methods

Demographic, clinical, and radiological data were collected from a consecutive group of patients evaluated for spontaneous intracranial hypotension during a 9-year period.

Results

Among 273 patients who met the diagnostic criteria for spontaneous intracranial hypotension and 42 who did not, not a single instance of CSF leak at the skull base was encountered. Clear nasal drainage was reported by 41 patients, but a diagnosis of CSF rhinorrhea could not be established. Four patients underwent exploratory surgery for presumed CSF rhinorrhea. In addition, the authors treated 3 patients who had a postoperative CSF leak at the skull base following the resection of a cerebellopontine angle tumor and developed orthostatic headaches; spinal imaging, however, demonstrated the presence of a spinal source of CSF leakage in all 3 patients.

Conclusions

There is no evidence for an association between spontaneous intracranial hypotension and CSF leaks at the level of the skull base. Moreover, the authors’ study suggests that a spinal source for CSF leakage should even be suspected in patients with orthostatic headaches who have a documented skull base CSF leak.

Key Words • cerebrospinal fluid leak • cerebrospinal fluid rhinorrhea • headache • intracranial hypotension

Methods

This study was approved by our institution’s institutional review board.

Patients evaluated for spontaneous intracranial hypotension at Cedars-Sinai Medical Center, Los Angeles, California, between January 1, 2001, and December 31, 2009, were identified. The diagnosis of spontaneous intracranial hypotension was made using previously established diagnostic criteria (Table 1). Demographic, clinical, and radiological data were retrieved from the patients’ medical records.

Results

During the 9-year study period, 315 patients were evaluated for spontaneous intracranial hypotension. Of those, 273 met the diagnostic criteria for spontaneous intracranial hypotension and 42 did not. No patient was found to have evidence for a CSF leak at the skull base as the cause of spontaneous intracranial hypotension.
Among the 273 patients meeting the diagnostic criteria, 33 (12%) reported clear nasal drainage associated with their orthostatic headaches. A relationship between headache intensity and occurrence of nasal drainage was noted by 14 of 25 patients in whom such a relationship was ascertained. Testing for β2-transferrin was performed in 11 patients and the results were normal in all. Twelve of the 33 patients reporting clear nasal discharge as well as 6 others who did not report any nasal discharge underwent radionuclide cisternography with nasal pledgets to evaluate for CSF rhinorrhea. Results were negative in 17 patients and borderline results were reported for 1 patient. In spite of normal findings on contrast CT cisternography, this latter patient as well as 3 of the patients with clear nasal drainage but normal pledget studies underwent exploratory endoscopic or microscopic sinus surgery. No evidence for a CSF leak was found at the time of surgery in these 4 patients. Eventually, imaging demonstrated a spinal CSF leak or the presence of spinal meningeal diverticula in all these 33 patients.

Among the 42 patients not meeting the diagnostic criteria, 8 (19%) reported clear nasal drainage associated with their orthostatic headaches. A relationship between headache intensity and occurrence of nasal drainage was noted by 3 of 5 patients in whom such a relationship was ascertained. Testing for β2-transferrin was performed in 3 patients and the results were normal in all. Four patients underwent radionuclide cisternography with nasal pledgets to evaluate for CSF rhinorrhea. Results were negative in 3 patients and mildly elevated counts were reported for 1 patient, who had a low opening pressure on serial lumbar punctures (5 cm H2O [normal 6.5–18.5 cm H2O]). In this latter patient, the results of β2-transferrin testing, brain MR imaging, invasive intracranial pressure monitoring, CT myelography, and contrast CT cisternography were all normal and the nasal drainage resolved without treatment while the orthostatic headaches continued.

We also evaluated 3 patients who suffered from orthostatic headaches following surgery for skull base tumors that appeared to be associated with postoperative skull base CSF leaks. Magnetic resonance imaging showed the typical features of intracranial hypotension in 2 of these patients, but further investigations demonstrated spinal CSF leaks as the cause of the orthostatic headaches in all 3 patients.

### Illustrative Cases

**Case 1**

This 59-year-old man underwent an uneventful translabyrinthine resection of a vestibular schwannoma. Two to 3 days later, he noted an occipital headache that was exclusively positional, occurring within seconds of assuming the upright position and relieved within 5–10 minutes of lying down. He also complained of a salty taste in the back of his throat. The headache progressed and 2 weeks later, CT examination showed bilateral subdural fluid collections and a marked increase of pneumocephalus compared with his immediate postoperative CT scan (Fig. 1). A diagnosis of postoperative skull base CSF leak was made and he underwent surgery to close the external ear canal and eustachian tube. His headache continued to progress and MR imaging showed the typical changes of spontaneous intracranial hypotension, consisting of bilateral subdural fluid collections, brain sagging, and pachymeningeal enhancement (Fig. 2). Magnetic resonance and CT myelography revealed a thoracic CSF leak (Fig. 3) with an opening pressure of 6 cm H2O. A lumbar epidural blood patch was placed with complete resolution of the positional headaches and MR imaging findings (Fig. 2).

**Case 2**

This 53-year-old woman underwent an uneventful retrosigmoid suboccipital craniotomy for resection of a cerebellopontine angle meningioma. Within 1 week, she noted an occipital headache associated with neck stiffness that was positional in nature, being relieved—albeit incompletely—within 30 minutes of lying down. Magnetic resonance imaging showed the typical changes of
spontaneous intracranial hypotension, consisting of brain sagging and pachymeningeal enhancement (Fig. 4). She also complained of a salty taste in the back of her throat. Otologic evaluation was suspicious for postoperative CSF rhinorrhea and otorrhea, and the patient eventually underwent 3 surgical procedures to eliminate the skull base CSF leak. However, her positional headaches and MR imaging findings of spontaneous intracranial hypotension persisted. Magnetic resonance and CT myelography demonstrated numerous large spinal meningeal diverticula with an upper thoracic CSF leak (Fig. 5). The opening pressure was 11 cm H₂O. A lumbar epidural blood patch was placed with temporary resolution of the positional headache and neck pain.

Case 3

This 46-year-old woman underwent an uneventful retrosigmoid suboccipital craniotomy for resection of a cerebellopontine angle meningioma. She complained of a salty taste in the back of her throat, and 4 months later she was successfully treated with a 1-week trial of lumbar CSF drainage. Three months later, she noted a generalized po-
sitional headache, occurring within 20 minutes to 2 hours of assuming the upright position and relieved within 1–5 minutes of lying down. The results of brain MR imaging were normal, except for the expected postoperative changes. Because of the positional nature of her headaches and the presumed skull base CSF leak, the patient underwent 2 further craniotomies to repair the putative CSF leak and 1 surgical procedure to close the external ear canal and eustachian tube. However, her headaches persisted while the salty taste resolved. A total of 6 epidural blood patches were placed, each resulting in good but only temporary relief of symptoms. Magnetic resonance and CT myelography revealed an L4–5 pseudomeningocele at the site of previous lumbar drain placement, a possible CSF leak associated with a small right T-12 meningeal diverticulum, and numerous large, mainly lumbosacral meningeal diverticula (Fig. 6), with an opening pressure of 1.5 cm H₂O. The patient underwent surgical repair of the pseudomeningocele and the right T-12 diverticulum with resolution of her positional headache.

**Discussion**

In this study of a large number of patients with spontaneous intracranial hypotension or suspected spontaneous intracranial hypotension, not a single instance of a CSF leak at the level of the base of the skull was detected. This calls into question the assertion that CSF rhinorrhea or otorrhea is associated with orthostatic headaches. In our study, a surprisingly large number of patients underwent evaluation for CSF rhinorrhea or even surgical treatment for presumed CSF rhinorrhea. This was due, at least in part, to the relatively common complaint of clear nasal drainage (in about one-eighth of our patients with spontaneous intracranial hypotension). The nasal drainage, however, was found not to be caused by CSF rhinorrhea. Possibly, activation of parasympathetic fibers due to headache via the trigeminal-autonomic reflex resulted in nasal hypersecretion, as has been noted not only in the rare trigeminal-autonomic cephalalgias, such as cluster headache, but also in common headache disorders, such as migraine. Nasal drainage mostly occurred when headache was more intense. This observation is consistent with activation of the trigeminal autonomic loop, which has been shown in clinic-based studies to be a pain intensity–driven reflex. In our study, 2 patients had borderline or mildly elevated counts on nasal pledgets when radionuclide cisternography was performed. However, it was unlikely that CSF rhinorrhea was the cause of their orthostatic headaches, because the results of contrast CT cisternography were normal and there was no temporal relationship between the occurrence of nasal drainage and the symptoms of spontaneous intracranial hypotension. Moreover, in one of these patients subsequent CT myelography revealed the presence of a spinal CSF leak. Equivocal or false-positive results from nasal pledgets are not uncommon.

The demonstration of spinal CSF leakage in 3 patients who developed orthostatic headaches following removal of skull base tumors complicated by postoperative skull base CSF leaks is perhaps the most obvious evidence against a CSF leak at the level of the skull base as a cause of orthostatic headaches. Two patients had the typical MR imaging findings of spontaneous intracranial hypotension and evidence for spinal CSF leaks on imaging. These patients can be considered to have postcraniotomy spontaneous intracranial hypotension, as has been described following a craniotomy for clipping of intracranial aneurysms. In the third patient it was likely that the postoperative orthostatic headaches were related to persistent CSF leakage from a lumbar drain site.

This study has several limitations. First, we used the diagnostic criteria for spontaneous intracranial hypotension that we developed several years ago (Table 1). Although no spinal imaging is required to meet these criteria, one criterion is the demonstration of a spinal CSF leak, possibly skewing the study population to those without skull base CSF leaks. Therefore, we also included pa-
Level of CSF leak and headaches

tients in this study who were evaluated by us for intracranial hypotension and did not meet these criteria. Second, thorough testing for skull base CSF leaks was not done for all patients in whom spontaneous intracranial hypotension was suspected, and it is possible that some skull base CSF leaks went undetected. This is unlikely, however, because the yield of testing for skull base CSF leaks in those patients with spontaneous intracranial hypotension who were most likely to have such a leak—that is, those with rhinorrhea—was nil. Third, selection bias was present in our study, as more than 70% of our patients with spontaneous intracranial hypotension are referred from outside southern California and it cannot be excluded that patients with intracranial hypotension due to skull base CSF leaks are routinely recognized and treated in the community rather than being referred to us. However, in our experience (W.I.S. and M.S.S.) with more than 200 patients with skull base leaks, none had the typical clinical and radiographic findings of intracranial hypotension. Also, in an extensive review of the literature we have not encountered such a case. Thus, in patients with skull base CSF leaks and headaches—a regular occurrence in neurosurgical practice—an alternative cause for the headache should be considered.

**Fig. 5.** Case 2. Magnetic resonance myelograms showing numerous meningeal diverticula of variable sizes involving the cervical (A), thoracic (B), and lumbosacral (C) spine.

**Fig. 6.** Case 3. A: An MR myelogram showing several lumbosacral meningeal diverticula. B and C: CT myelograms showing a small meningeal diverticulum arising from the right T-12 nerve root sleeve (B, arrow) and a pseudomeningocele at L4–5 (C, arrow).
Conclusions

Spontaneous intracranial hypotension is not caused by a CSF leak at the level of the skull base. Clear nasal discharge in patients with spontaneous intracranial hypotension can be considered a false localizing sign and may be a trigeminal nerve–based epiphenomenon. Moreover, a spinal source for CSF leakage should even be suspected in patients with orthostatic headaches who have a documented skull base CSF leak.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following: Conception and design: all authors. Acquisition of data: all authors. Analysis and interpretation of data: all authors. Drafting the article: all authors. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Schievink.

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