Syringomyelia in spontaneous intracranial hypotension

Case report

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The authors describe the presence of a syrinx in a patient with spontaneous intracranial hypotension. The likely pathophysiological basis of this hitherto unreported association is also presented. It is believed that chronic obstruction at the foramen magnum may be the most important factor for the development of syringes.

KEY WORDS • spontaneous intracranial hypotension • syringomyelia • magnetic resonance imaging

S PONTANEOUS intracranial hypotension is a rare but well recognized clinical entity caused by spontaneous extravasation of CSF from the spinal subarachnoid space. With widespread use of MR imaging, increasing numbers of these cases are being recognized. Cranial MR imaging findings in these patients have been extensively reported, whereas experience with spinal MR imaging findings has been limited, and the findings described in the literature include presence of epidural fluid collections and pachymeningeal enhancement following administration of contrast agents. We present a case of SIH in which spinal MR imaging demonstrated the presence of syringomyelia and epidural as well as extraspinal CSF collection. The presence of syringomyelia in a setting of SIH has not been reported previously.

Case Report

History and Examination. This 32-year-old woman presented with severe headaches that she had suffered for 4 years and numbness on the right side of her body that had lasted for 2 months. The headaches were positional and were aggravated by sitting or standing and relieved by lying down. She had no history of trauma or spinal intervention. Results of her neurological examination were remarkable for loss of sensations and deep tendon reflexes in the right arm.

Cranial MR Imaging. Multiplanar cranial MR imaging was performed using a 0.5-tesla magnet without intravenous administration of contrast agents. These tests revealed tonsillar herniation to the C-1 level, which was associated with effacement of basal cisterns and flattening of the pons against the clivus, and effacement of the pontomedullary sulcus. Sagging of the optic chiasm was seen, with protrusion of the pituitary gland into the suprasellar cistern. The midbrain was elongated anteroposteriorly, and descent of the canaliculus indicated descending transental herniation. Additionally, sagittal MR images revealed the presence of a syrinx in the visualized part of the cervical spinal cord (Fig. 1 left).

Spinal MR Imaging. Cranial imaging was followed by MR imaging of the cervical spine. Sagittal and axial images were acquired using T₁-weighted spin-echo and STIR sequences. Wide field-of-view images of the cervicodorsal spine were also obtained. On MR imaging of the spine the presence of a syrinx was confirmed in the cervical spinal cord extending into the dorsal region to the T-9 level (Fig. 1 center). Also seen was fluid collection in the epidural space from the foramen magnum to the C-2 level. The latter extended along the right C-2 nerve root into paraspinal space and the suboccipital region (Fig. 1 center and right). The fluid was isointense to CSF on both sequences and was especially observable on STIR images because the bright intensity of the fluid contrasted sharply with the dark signal from muscles and fat.

Initial Treatment. Because of the patient’s history of postural headaches and the imaging features of brain sagging and fluid collection in epidural and extraspinal space,
the diagnosis of tonsillar herniation with syrinx formation caused by spontaneous spinal CSF leakage and chronic intracranial hypotension was considered. The patient was maintained on conservative treatment for the next 2 months but failed to respond. Her headaches continued unabated and the weakness in her right arm progressed; weakness also developed in her right leg. She was admitted to another hospital because her condition had gradually deteriorated.

**Operation.** In view of the syrinx and rapid worsening of symptoms of myelopathy, foramen magnum decompression with excision of the C-1 arch was planned. At surgery, no definite dural tear was seen. Hence, after completion of the planned surgery, a large sheet of Gelfoam was placed over the dura extending from the posterior fossa to below C-1.

**Postoperative Course.** The patient noted marked relief of her headaches the day after surgery; complete relief occurred after a few days. Her strength on her right side has gradually improved, although it has not yet recovered completely.

**Follow-Up Review.** Repeated MR imaging of the head was performed 3 months after postsurgery and demonstrated resolution of features of brain sagging. The tonsils had ascended and returned to their normal shape. The chiasm appeared straight and the upward bulging of the pituitary was no longer seen. The basal cisterns were well visualized and the mammillopontine distance had increased. The pons had a normal shape and the pontomedullary sulcus was well visualized (Fig. 2 upper). These findings indicated resolution of acquired tonsillar herniation and intracranial hypotension.

Repeated MR imaging of the cervical spine demonstrated a surgically created pseudomeningocele in the suboccipital region. The syrinx had collapsed, and no fluid collection was now seen in the epidural or extraspinal space, indicating arrest of the CSF leak by the Gelfoam (Fig. 2 lower).

**Discussion**

Spontaneous intracranial hypotension is a syndrome of reduced intracranial volume and pressure that results from extravasation of CSF from the spinal subarachnoid space. Most cases are thought to resolve spontaneously. This condition may become chronic, however, and cause sagging of intracranial structures. The downward displacement of intracranial contents is most prominent in the posterior fossa. Typically, patients present with headaches, which worsen in an upright posture and are relieved by lying down. Occasionally, chronicity of the disease process may obscure the initial postural nature of the headache. Other symptoms that have been described in these patients are related to displacement and/or stretching of intracranial structures. These include hearing disturbances, tinnitus, vertigo, ataxia, visual deficits, and diplopia. Symptoms related to the spine are seldom seen in these patients and have been limited to neck pain and radiculopathy. Symptoms related to myelopathy, as seen in our case, have rarely been described. Theoretically, epidural fluid collections seen in these cases may also lead to myelopathy. Although we did not find any reports of such cases, there has been a report on a patient who had a positive Lhermitte sign related to the presence of cervical epidural CSF collection.

The cranial MR findings in SIH have been described in detail. There is characteristic sagging of intracranial structures in the absence of supratentorial disease. Tonsillar herniation, flattening of the pons against the clivus, obliteration of intracranial cisterns, sagging of the optic chiasm, descending transtentorial herniation, descent of the canaliculus, and evacuation of the pituitary gland through the diaphragma sella into the suprasellar cistern have been noted in almost all cases. Although tonsillar herniation is identical to that seen in Chiari I malformations, other associated findings of brain sagging and a history of postural headaches should eliminate this possibility. Indeed, it has been claimed that previous reports of patients with
Several authors have documented communication between such extradural fluid collections and the subarachnoid space on computerized tomography myelography, indicating that these collections represent extravasated CSF. Other tests like radionuclide cisternography or computerized tomography myelography can be performed to locate the site of the leak, especially if the collection extends over several vertebral levels.

An unusual finding in our case was the presence of syringomyelia; an association of syrinx with SIH has never been described. In fact, it has been asserted that the presence of a syrinx rules out the possibility of SIH. Our patient, however, had typical clinical and MR imaging features of SIH in addition to the presence of a syrinx. Although unreported in SIH, syringes have been reported as resulting from iatrogenic drainage of CSF from the lumbar subarachnoid space. Johnston, et al., described seven patients who developed tonsillar herniation and syringomyelia after placement of lumbar subarachnoid fluid shunts for various conditions. None of these patients had tonsillar herniation or syringomyelia before treatment. Although the authors mention only tonsillar herniation and syringomyelia as the imaging findings, the illustrations provided clearly show other features of brain sagging like flattening of the pons against the clivus, descent of the canalculus, obliteration of the cistern, bulging of the pituitary gland, and bowing of the chiasm. Thus, their cases had MR imaging features similar to those seen in cases of SIH. Indeed, tonsillar herniation caused by lumbar CSF diversion procedures is well described by several other authors and may produce symptoms due to intracranial hypotension.

We think that the rarity of syringes in cases of SIH is caused by the fact that most of these cases resolve either spontaneously or following treatment. Hence, the tonsillar herniation is never as longstanding as that seen following spinal drainage procedures. This hypothesis is supported by the fact that in cases described by Johnston, et al., syrinx was observed a few years after the placement of lumboperitoneal shunts. In our case, the symptoms related to intracranial hypotension were present for years before myelopathy developed. Such chronicity is the likely explanation for the development of a syrinx.

Most patients with SIH respond to conservative therapy, whereas others may require epidural blood patches, which can be targeted to the region of the leak as demonstrated on MR imaging. Only a few refractory cases require surgery. We chose to perform foramen magnum decompression and excision of posterior arch of C-1 along with placement of Gelfoam over the dura mater because our patient had shown rapid worsening of myelopathy. The postoperative MR image demonstrated successful treatment of the CSF leak, tonsillar herniation, SIH, and syringomyelia.

In conclusion, we present a case of SIH with syringomyelia. This case is instructive in that it shows another, hitherto unrecognized, clinicoradiological manifestation of SIH.

References


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