Cerebral dissection from syringomyelia demonstrated using cine magnetic resonance imaging

Case report

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A 16-year-old boy presented at the authors’ emergency department with a sudden deterioration of respiration. He had been paraparetic for 3 years and had become quadriplegic 2 days previously. Magnetic resonance images revealed a Chiari I malformation and a hydromyelic cavity extending from C-1 to T-11. Rostrally, a small cylindrically shaped lesion extended from the cervicomedullary junction to the left semi-oval center. The patient made a dramatic neurological recovery following sub-occipital craniectomy and upper cervical laminectomies with augmentation duraplasties followed by placement of a syringoperitoneal shunt.

KEY WORDS • cerebral dissection • syringomyelia • Chiari malformation • cine magnetic resonance imaging

SYRINGOBULBIA frequently appears as a bulbar extension of cervical syringomyelia. The terms “syringoecephaly” and “cerebral syrinx” have both been used to describe a cavity located rostral to the brainstem. According to the literature, extension of this cavity to a higher level is extremely rare. We could locate only five previous reports containing in vivo radiological and other imaging evidence of the syrinx extending rostral to the brainstem.

To our knowledge, our case is the first in which cine MR images visualized the continuity of CSF flow through the syringomyelia, syringobulbia, and syringocephalia. In this publication, we present an extreme example of altered CSF dynamics that led to the formation of a cerebral dissection and syringocephalia.

Case Report

This 16-year-old boy was referred from a local hospital because he experienced a sudden onset of dyspnea, dysarthria, and dysphagia.

Examination. On gross appearance, the patient was underweight and cyanotic. His four extremities were hypotrophic and, in addition, he had an oxycephaly-like skull anomaly resulting from a difficult labor during his birth. In the emergency department, the patient was drowsy and quadriplegic and exhibited a decerebrated posture. He perspired only on his right side and displayed hypesthesia in response to pain stimulus. The degree and level of his sensory disturbance, however, were unverifiable because of his drowsiness. In addition he had lost sphincter control. Whole-spine MR imaging revealed a longitudinal cystic lesion in the intramedullary portion of the spinal cord extending from the C-1 to T-11 level, which indicated syringomyelia (Fig. 1). Rostrally, an MR image of the brain revealed another cylindrically shaped cystic lesion extending from the left cerebellar peduncle and the medulla oblongata up to the posterior limb of the left internal capsule and semi-oval center. The maximum diameter of the lesion was found at the cervicomedullary junction, but the lesion was discontinuous with the hydromyelia at the C-1 level. On both T1 and T2-weighted MR images, the signal of the cystic fluid content was comparable to that of CSF, which indicated that the lesion in the brainstem was a syringobulbia (Fig. 1). The MR images also revealed a crowded craniovertebral junction, which was characterized as a Chiari I malformation, with tonsillar impaction through the foramen magnum; however, there was no associated hydrocephalus or signal change in the upper cervical cord. Because we initially suspected the lesion to be a lacunar infarct, we performed cerebral angiography, but the angiogram revealed no causative abnormality. To determine whether any communicating flow existed between the hydromyelia and the cerebral lesion, we performed cine MR imaging. However, it only demonstrated active pulsation in the spinal cord syrinx, with faint CSF flow in the ventral subarachnoid space and nearly absent dorsal CSF flow. There was no evidence of CSF flow communication between the hydromyelia and the cerebral lesion (Fig. 1). We hypothesized that the cause of the patient’s apnea and quadriplegia was brainstem swelling as-

Abbreviations used in this paper: CSF = cerebrospinal fluid; MR = magnetic resonance.
associated with the sudden development of the syringobulbia, rather than the syrinx itself. After injection of steroid medication, the patient’s respiratory distress and quadriplegic state improved slightly, but his condition again deteriorated and he suffered another attack of apnea 2 days later.

First Operation. On an emergency basis, we performed suboccipital craniectomy and C-1 total laminectomy with augmentation duraplasty 8 hours following the apnea attack. During the operation, severe adhesion was noted in the dorsal subarachnoid space and the CSF flow was nearly absent. Adhesiolysis was therefore performed to separate the dura and the inferior margin of the cerebellar tonsil.

Postoperative Findings and Course. Postoperatively, the patient’s condition markedly improved so that he was able to raise his arm over his shoulder. Cine MR imaging performed 10 days later revealed increased CSF flow, especially in the ventral subarachnoid space of the cervical spinal cord, although there was still faint CSF flow in the dorsal subarachnoid space and active flow of CSF from the cerebral aqueduct and fourth ventricle to the hydromyelia (Fig. 2). Surprisingly, it also revealed active upward flow of CSF to the dissected cavity-like lesion in the brainstem and reaching up to the internal capsule and semioval center, which indicated that the lesion was a cerebral syrinx formed by a dissecting upward jet flow from the hydromyelia (Fig. 2). Moreover, the fourth ventricle appeared to be mildly dilated, compared with findings on preoperative images, demonstrating the development of hydrocephalus caused by the active upward flow. Two weeks after the first operation, the patient again suffered from dyspnea; cervical MR images revealed both ventriculomegaly and an increase in the size of the syrinx in the brainstem and the upper cervical spinal cord, compared with findings on preoperative images, from which we inferred that there might be rapid refilling of the hydromyelic cavity from the hydrocephalic ventricular system.

Second Operation. One week later, we performed additional C-2 and C-3 total laminectomies with augmentation duraplasty and adhesiolysis. During the second operation, we found good pulsation of the dura at the site of the previous operation at the C-1 level, without visible pulsation distally on the C-2 and C-3 levels. Instead, we found a bulging of the dural sac caused by an enlarged syrinx.

Second Postoperative Findings and Course. Results of cervical MR imaging and cine MR imaging performed 2 weeks after the second operation revealed a decreased size of the patient’s hydromyelia in the cervicomedullary junction and an increased CSF flow in the dorsal subarachnoid space.
space in the upper cervical spinal cord. However, upward reflux flow of CSF from the hydromyelia to the cerebral syrinx persisted and no improvement of hydrocephalus was apparent. After the second operation, the patient’s condition again improved markedly and without deterioration. He was able to breathe regularly through his tracheal cannula and, at this point, we were able to transfer him from the intensive care unit to a general ward. His upper extremity motor power improved to Grade III and his pain sensation returned to almost normal. Nevertheless, he was still paraparetic and unable to void by his own volition. He was transferred to a rehabilitation ward 1 month after the second operation and was discharged 1 week later in a wheelchair.

One month after discharge, the patient returned to the emergency department with respiratory distress and worsened quadriparesis. An MR image of the brain revealed improved ventriculomegaly, with no change in the size of the cerebral syrinx and an increase in size of both the fourth ventricle and the syrinx at the cervicomedullary junction. The cerebellar tonsils were still herniated as a result of the severe adhesion noted in the dorsal subarachnoid space during the previous operations. Magnetic resonance images of the cervicothoracic spine revealed that the hydromyelia persisted with no change in size (Fig. 3).

**Third Operation.** Perceiving that the previous decompressive operations were insufficient to reduce the size of the syrinx, especially at the cervicomedullary junction, we performed T-5 and T-6 bilateral total laminectomies and inserted a syringoperitoneal shunt.

**Third Postoperative Findings and Course.** Magnetic resonance images of the cervicothoracic spine obtained 2 weeks later revealed a reduced hydromyelia along the whole length of the patient’s spinal cord (Fig. 3). Cine MR images demonstrated good caudal flow of CSF from the ventricular system to the central canal, and only a slight upward flow of CSF from the hydromyelia to the cerebral syrinx. The patient again displayed a marked recovery and, this time, he was able to walk with the aid of a walker or his parents. Six months after discharge, at his follow-up examination, no clinical deterioration was noted.

**Discussion**

The hydromyelic cavity associated with Chiari malformation infrequently extends into the brainstem and rarely extends into the cerebrum. Moreover, the exact pathogenesis of such a lesion is unclear. With regard to cavity formation in the cerebral syrinx, Jonesco-Sisesti described three possible positions for the cavity found in syringobulbia: 1) anterolaterally directed from the floor of the fourth ventricle external to the hypoglossal nucleus; 2) extending from the fourth ventricle along the median raphe; and 3) ventrally situated between the pyramid and the inferior olive (the rarest location). In our case, the syrinx extended from the C-1 to the T-11 level, and another cerebral syrinx extended from the cervicomedullary junction to the left semiolvent center. This latter syrinx was located from the fourth ventricle and aqueduct, as in with the rarest location described by Jonesco-Sisesti. It is probable that the formation of a cerebral syrinx begins with the formation of a faint communicating tract between the hydromyelic cavity and the cerebral syrinx. The tract may be formed by an unusual dissection of the normal brainstem in a location far from the fourth ventricle, or it may be due to an unusual diverticulum of the aqueduct and ventricular system, formed by an underlying malformation of the glioseptal structure of the cerebrum, and derived from a
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congenital factor or an acquired cause, such as birth trauma. The brainstem may then be easily and suddenly dissected by an increased upward flow pressure of CSF from the hydromyelic cavity to the ventricular system, which is caused by the cranial–spinal pressure dissociation associated with tissue impaction, such as that which occurs with a Chiari I malformation at the cervicomedullary junction. This tract may not be visualized on standard MR images due to compression of the tract by tissue impaction at the cervicomedullary junction. Likewise, in our case, both lesions were revealed to be separated at the cervicomedullary junction by standard sagittal MR imaging, but cine MR imaging demonstrated a communicating upward jet flow of CSF from the hydromyelia to the cerebral syrinx. The patient's relatively acute onset of dyspnea supports the idea that its cause may have been sudden brainstem dissection. The associated hydrocephalus may have developed from rapid refilling of the ventricular system by the increased returning flow of CSF through the decompressed cervical subarachnoid space to the intracranial portion after decompression, or from active upward flow of CSF from the hydromyelia to the ventricular system through the decompressed cervicomedullary junction.

The neuroimaging findings in this case also deserve comments. In the previous five reports of syringobulbia or syringomesencephaly the lesion was visualized using spinal cord syringography or sagittal views of MR imaging. However, the actual communication between the cerebral syrinx and the hydromyelic cavity, caused by upward jet flow of CSF, was verified by cine MR imaging in this case for the first time. The superb ability of cine MR imaging to demonstrate actual communicating flow of CSF undoubtedly will make it the method of choice in the examination of suspected syringobulbia or syringomesencephaly, and it may also play a significant role in defining the mechanism of the formation of syringobulbia.

Because the cerebral syrinx was demonstrated to be a cranial extension of the syringomyelia by using cine MR imaging, we expected that extensive decompression and adhesiolysis would be effective in regaining active subarachnoid CSF flow in the ventral and dorsal subarachnoid spaces at the cervicomedullary junction and in reducing the size of the cerebral syrinx. Such a procedure may also prevent the occurrence of a shunt-associated complication, such as a shunt malfunction or a myelotomy-related neurological deficit. When the patient’s condition again deteriorated, instead of inserting a syringo-subarachnoid shunt we performed additional C-2 and C-3 laminectomies and duraplasty to decompress the still obliterated CSF flow in the dorsal subarachnoid space at the cervicomedullary junction. However, after the second operation, cine MR images revealed an increased upward jet flow from the syringomyelia to the cerebral syrinx as well as progression of the patient’s hydrocephalus. This indicated that the previous two decompressive operations were not sufficient to restore the altered CSF flow dynamics at the cervicomedullary junction or to reduce the size of the hydromyelia. Even though the suboccipital craniectomy and several levels of upper cervical laminectomy with augmenting duraplasty is recommended as a treatment of choice for young patients with hydromyelia associated with the Chiari malformation, sometimes a shunt placement procedure is also indicated to decompress the syrinx cavity itself.

References

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