Successful neuroendoscopic third ventriculostomy for hydrocephalus and syringomyelia associated with fourth ventricle outlet obstruction

Case report

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The authors report the use of neuroendoscopic third ventriculostomy to treat successfully both hydrocephalus and syringomyelia associated with fourth ventricle outlet obstruction. A 27-year-old woman presented with dizziness, headache, and nausea. Magnetic resonance (MR) imaging demonstrated dilation of all ventricles, downward displacement of the third ventricular floor, obliteration of the retrocerebellar cerebrospinal fluid (CSF) space, funnel-like enlargement of the entrance of the central canal in the fourth ventricle, and syringomyelia involving mainly the cervical spinal cord. Cine-MR imaging indicated patency of the aqueduct and an absent CSF flow signal in the area of the cisterna magna, which indicated obstruction of the outlets of the fourth ventricle. Although results of radioisotope cisternography indicated failure of CSF absorption, neuroendoscopic third ventriculostomy completely resolved all symptoms as well as the ventricular and spinal cord abnormalities evident on MR images. Neuroendoscopic third ventriculostomy is an important option for treating hydrocephalus in patients with fourth ventricle outlet obstruction.

**KEY WORDS** • hydrocephalus • syringomyelia • Chiari malformation • neuroendoscopic surgery • third ventriculostomy

Fourth ventricle outlet obstruction is a rare syndrome that is associated with hydrocephalus and syringomyelia, which are frequently found in patients with Chiari malformation; such an obstruction is caused by chronic tonsillar herniation. Symptomatic hydrocephalus and syringomyelia have been treated by decompression of the posterior fossa, ventriculoperitoneal or Torkildsen shunt placement, and neuroendoscopic third ventriculostomy. We report on a patient in whom neuroendoscopic third ventriculostomy was used successfully to treat hydrocephalus and syringomyelia associated with fourth ventricle outlet obstruction.

**Case Report**

_History and Examination._ This 27-year-old woman began to experience dizziness, headache, and nausea 6 weeks before admission to the hospital. Admission neurological examination showed no abnormality despite these symptoms, and the results of routine chemical analysis of her blood were normal. Intracranial pressure measured by lumbar puncture was slightly elevated to 180 mm Hg.

**FIG. 1.** Noncontrast-enhanced CT scan obtained 2 years before hospital admission demonstrating normal ventricle size.

**Abbreviations used in this paper:** CSF = cerebrospinal fluid; CT = computerized tomography; MR = magnetic resonance.
MR images demonstrated dilation of all ventricles (bicaudate index 22.6%), downward deviation of the third ventricular floor, obliteration of the retrocerebellar CSF space, funnellike enlargement of the entrance of the central canal from the fourth ventricle, and syringomyelia involving mainly the cervical spinal cord (C, T₁-weighted image; D, T₂-weighted image). E: Cine-MR image demonstrating patency of the aqueduct and absence of a CSF flow signal in the area of the cisterna magna.

Operation and Postoperative Course. Neuroendoscopic third ventriculostomy was performed in the premammillary region. The postoperative period was uneventful and all symptoms resolved completely immediately after the operation. Postoperative MR imaging (Fig. 4A–D) demonstrated that dilation of all ventricles, downward deviation of the third ventricular floor, and syringomyelia had completely resolved (bicaudate index 14.4%). Postoperative cine-MR imaging (Fig. 4E) revealed a new CSF flow signal in the area of the third ventricular floor. Postoperative radioisotope cisternography indicated radioisotope reflux into the ventricles, probably through the fenestration of the third ventricular floor, but clearance of the radioisotope from the cistern still was delayed (Fig. 5).

Discussion

Milhorat, et al., recently reported clinical features in 364 patients with Chiari I malformation. According to their report, 65.4% of patients had syringomyelia and 6.9% had hydrocephalus. Both abnormalities are caused by chronic tonsillar herniation. The mechanisms precipitating adult onset of hydrocephalus associated with Chiari I malformation are unknown. Some authors have proposed that meningoencephalitis, subclinical inflammation, or even connective tissue proliferation without inflammatory changes could cause fourth ventricle obstruction. Our patient had no apparent episode of meningitis or other inflammation during the 2 years between admission and the earlier CT scan in which no ventricular dilation had been observed. Therefore, the cause of adult-onset hydrocephalus in this case remains unknown. The MR image was highly diagnostic in demonstrating the findings of hydrocephalus together with the fourth ventricle outlet ob-
Preoperative MR imaging indicated the presence of a Chiari I malformation; however, postoperative MR imaging demonstrated the cerebellar tonsil with no downward dislocation under the foramen magnum. Milhorat, et al.,17 have described the obliteration of retrocerebellar fluid space in patients with Chiari I malformation, which also was evident in this case; our patient may have had a Chiari I malformation.

In our case MR imaging revealed a notch in the fourth ventricular floor that indicated a funnellike enlargement of the entrance of the central spinal canal. Such a dilation, associated with syringomyelia extending from C-1 throughout the cervical portion of the spinal cord, together with its rapid resolution after third ventriculostomy, indicated that syringomyelia in our case represented a dilated central canal (hydromyelia).16 In demonstrating CSF dynamics, cine-MR imaging accurately depicted patency of the aqueduct and decreased CSF flow in the cisterna magna region; postoperatively it demonstrated a CSF flow signal in the area of the third ventricular floor that confirmed CSF flow through the ventriculostomy.6,17,22 These MR imaging findings were useful in choosing third ventriculostomy in our patient and evaluating its success.

Treatment of this uncommon form of fourth ventricular outlet obstruction is controversial. Exploration of posterior fossa structures and excision of the membrane obstructing ventricle outflow is often advocated.1,23 Lateral or fourth ventricle shunts for CSF diversion have also been placed.7,12,13,16 Neuroendoscopic third ventriculostomy has shown considerable promise in the management of noncommunicating hydrocephalus, with success rates as high as 80%.3,4,11,13,21 This type of ventriculostomy has been applied to hydrocephalus caused by fourth ventricle outlet obstruction1,3,13,19 and, in our case, it led to resolution of both hydrocephalus and syringomyelia associated with a Chiari I malformation. Neuroendoscopic third ventriculostomy has advantages over alternative procedures,20,14,15,24,26 and it should be considered for treating hydrocephalus in patients with Chiari I malformations.

Despite successful neuroendoscopic third ventriculostomy in our case, one abnormality remained in addition to the malformation itself: abnormal CSF absorption. Theoretically, third ventriculostomy is effective for obstructive hydrocephalus, but not for communicating hydrocephalus due to defective CSF absorption.3,4,11,13,21,25 We evaluated pre- and postoperative CSF flow studies in detail. Although MR imaging and cine-MR imaging demonstrated fourth ventricle outlet obstruction, both pre- and postoperative radioisotope cisternography showed nonfunction of CSF-absorbing structures in our case. Nonetheless, all symptoms resolved postoperatively, as well as the MR findings of hydrocephalus and syringomyelia.

Third ventriculostomy has shown limited effectiveness in patients in whom hydrocephalus is associated with spinal dysraphism, and even in some patients who have aqueductal stenosis.11,12,21,25 The lower success rate for third ventriculostomy in relieving hydrocephalus in these situations is related to structural immaturity of the subarachnoid space or the occurrence of subclinical adhesive inflammation in this space, which precludes adequate CSF absorption.2,13,23,25 However, several authors have stressed the importance of pulsatile CSF flow in causing symptoms associated with communicating hydrocephalus and have proposed third ventriculostomy in cases of normal-pressure hydrocephalus in which shunt placement has led to complications.3,10,18,19,28 Our case also confirms that third ventriculostomy is not contraindicated in hydrocephalus that includes a communicating component.

References


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