Disproportionately Large Communicating Fourth Ventricle With Syringomyelia
—Case Report—

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Abstract

A 13-year-old boy presented with syringomyelia associated with disproportionately large communicating fourth ventricle (DLCFV) manifesting as symptoms attributable to hydrocephalus and characteristic posterior fossa symptoms. Magnetic resonance imaging demonstrated remarkable dilation of the fourth ventricle and syringomyelia. Ventriculoperitoneal shunting completely resolved all symptoms as well as the ventricular and spinal cord abnormalities. Pre- and postoperative cine magnetic resonance imaging revealed the change of cerebrospinal fluid flow signal in the area of the foramen magnum. We concluded that the syringomyelia could be described as enlargement of the central canal with DLCFV.

Key words: disproportionately large communicating fourth ventricle with syringomyelia, syringomyelia, Chiari malformation, ventriculoperitoneal shunt

Introduction

Disproportionately large communicating fourth ventricle (DLCFV) was first proposed in 1980 as a subtype of hydrocephalus with dilation of all ventricles, disproportionately enlarged but not isolated fourth ventricle, no history of ventriculoperitoneal (VP) shunt, and decrease in size of all ventricles after VP shunt. Two mechanisms have been proposed for the occurrence of DLCFV. Occlusion in or near the fourth ventricle outlet interacts with collision of cerebrospinal fluid (CSF) pulse waves against the obstruction to yield a “water hammer” effect on the fourth ventricle, or the brain stem parenchyma around the fourth ventricle is abnormally weak to CSF pressure. DLCFV is somewhat different from the well-known clinical entity of isolated or trapped fourth ventricle because of the apparent patency of the aqueductal canal. Here we describe a case of DLCFV associated with syringomyelia secondary to tonsillar herniation.

Case Report

A 13-year-old boy had complained of headache 2 years previously, and magnetic resonance (MR) imaging performed in another hospital demonstrated dilation of the fourth ventricle but normal other ventricles (Fig. 1). He began to suffer dizziness, headache, walking disturbance, and incontinence, and had nausea 4 weeks before admission. On admission, neurological examination showed nys-
Fig. 2 A: T₁-weighted magnetic resonance (MR) images on admission showing the remarkably dilated fourth ventricle, and mildly dilated other ventricles. B, C: Sagittal T₁-weighted (B) and T₂-weighted (C) MR images on admission showing tonsillar herniation (arrow), funnel-like enlargement of the entrance of central canal in the fourth ventricle, and huge syringomyelia.

Fig. 3 Preoperative cine magnetic resonance (MR) image (left) showing no cerebrospinal fluid (CSF) flow signal in the area of the foramen magnum (arrows), and a small CSF flow signal without pulsation in the area of the syrinx. Postoperative cine MR image (right) clearly showing CSF flow signal with pulsation in the area of the foramen magnum (arrows), but the CSF flow signal detected preoperatively in the area of the syrinx was absent.

Fig. 4 A: Postoperative T₁-weighted magnetic resonance (MR) images demonstrating remarkable resolution of the dilation of all ventricles. B: Postoperative sagittal T₁-weighted MR image demonstrating complete resolution of tonsillar herniation (arrow) and huge syringomyelia.

tagmus and truncal ataxia in addition to these symptoms. Routine biochemical analysis found no abnormalities. The main symptoms were attributable to hydrocephalus, but posterior fossa symptoms such as nystagmus and truncal ataxia were also present.

MR imaging demonstrated remarkable dilation of the fourth ventricle and mild dilation of the other ventricles (Fig. 2A). Moreover, MR imaging demonstrated tonsillar herniation, funnel-like enlargement of the entrance of central canal in the fourth ventricle, and syringomyelia extending from C-1 throughout the lumbar portion of spinal cord (Fig. 2B, C). Cine MR imaging revealed no CSF flow signal in the area of the foramen magnum and a small CSF flow signal without pulsation in the area of the syrinx (Fig. 3 left). Three days after admission, his consciousness deteriorated to drowsiness, and we decided to operate immediately.

Immediately after a routine VP shunt operation,
all symptoms were completely resolved. Postoperative MR imaging demonstrated that dilation of all ventricles, tonsillar herniation, and syringomyelia had disappeared (Fig. 4). Postoperative cine MR imaging revealed CSF flow signal with pulsation in the area of the foramen magnum. The CSF flow signal detected preoperatively in the area of the syrinx was absent (Fig. 3 right). Postoperative cisternography showed no obstruction in any ventricle (data not shown). The results of CSF cytological examination and biochemical studies were normal.

**Discussion**

The present case of DLCFV presented with disproportionate enlargement of the fourth ventricle with dilation of all other ventricles. VP shunt had not been performed before admission. Systemic examination, past history, and CSF cytological examination found no abnormalities. All ventricles were decreased in size after VP shunt, and postoperative cisternography revealed apparent patency of the aqueduct. These findings fully corresponded to the characteristics of DLCFV. Two years before admission, this patient presented with enlarged fourth ventricle without tonsillar herniation. As the fourth ventricle had been enlarged for 2 years, the posterior fossa gradually became tight, which led to tonsillar herniation. On admission, the fourth ventricle was remarkably enlarged with tonsillar herniation associated with syringomyelia.

Syringomyelia is known to occur with acquired lesions of the foramen magnum. In addition, syringomyelia is occasionally associated with posterior fossa tumors and tumor-induced tonsillar herniation. Various mechanisms of syringomyelia secondary to tonsillar herniation due to posterior fossa tumors and Chiari malformation have been proposed. Since congenital anomalies at the foramen magnum were first described in patients with syringomyelia associated with Chiari malformations, it has been widely accepted that obstruction of CSF flow at the outlet of the fourth ventricle exerts a “water hammer” effect on the central canal, which enlarges the central canal and leads to development of syringomyelia. Occasional blockage of CSF flow at the foramen magnum leads to pressure dissociation between the cranial cavity and the spinal canal through packing of the herniated cerebellar tonsils. The transfer of CSF from the cranial cavity to the central canal leads to the development of syringomyelia.

Acquired lesions at the level of foramen magnum can also occlude the free pulsatile movement of CSF (i.e., movement up and down the subarachnoid space across the foramen magnum) and cause the pulsatile pressure waves to force CSF into the cord through the perivascular and interstitial spaces. Moreover, communication between the fourth ventricle and the syrinx via a patent central canal at the obex is unnecessary for the development, maintenance, or progression of syringomyelia, and so the same mechanism may cause both syringomyelia associated with posterior fossa tumors and syringomyelia associated with Chiari malformation.

Syringomyelia can be classified into the communicating, non-communicating, and atrophic types. The communicating type is associated with hydrocephalus and shows enlargement of the central canal with direct communication with the fourth ventricle. The non-communicating type is described as enlargement of the central canal without direct communication with the fourth ventricle. The atrophic type is described as syrinx without communication with either the central canal or the fourth ventricle. In our case, preoperative MR imaging revealed syringomyelia with extension from C-1 throughout the lumbar portion of spinal cord. Preoperative cine MR imaging revealed no CSF flow signal in the area of foramen magnum, indicating that the outlet obstruction involved the fourth ventricle. After VP shunt, the huge syringomyelia had completely disappeared, and postoperative cine MR imaging revealed normal CSF flow signal with pulsation in the area of the foramen magnum. Although MR imaging did not confirm the patency of the central canal between the fourth ventricle and the syrinx in our case, postoperative MR imaging showed that the syringomyelia disappeared or shrank after VP shunt without direct procedures to the syrinx or foramen magnum decompression, which clearly indicated communicating syringomyelia.

We consider secondary tonsillar herniation by DLCFV could be main cause of syringomyelia. The syringomyelia in our case was probably caused by enlargement of the central canal and communication with the fourth ventricle. As the fourth ventricle enlarged, the posterior fossa became tight, which led to tonsillar herniation as seen in patients with posterior fossa tumor and Chiari malformation. Consequently, the CSF flow between the fourth ventricle and subarachnoid space around the foramen magnum was blocked, which led to the development of syringomyelia through the “water hammer” effect on the central canal.

**References**

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