Disproportionately Large Communicating Fourth Ventricle Associated With Syringomyelia and Intradural Arachnoid Cyst in the Spinal Cord Successfully Treated With Additional Shunting

—Case Report—

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Abstract

A 44-year-old woman presented with a rare case of disproportionately large communicating fourth ventricle (DLCFV) associated with syringomyelia and intradural arachnoid cyst in the spinal cord. Ventriculoperitoneal shunt operation was performed for hydrocephalus after subarachnoid hemorrhage. She developed DLCFV, which was then associated with syringomyelia and spinal intradural arachnoid cyst. Shunting of the fourth ventricle improved DLCFV, and then the syringomyelia and arachnoid cyst. Although the aqueduct was patent, independent pressure control of the fourth ventricle and the other ventricles was necessary to improve the symptoms. Shunting of the fourth ventricle should be considered for patients with DLCFV when the symptoms persist despite adequate pressure control of the other ventricles.

Key words: disproportionately large communicating fourth ventricle, syringomyelia, intradural arachnoid cyst, ventriculoperitoneal shunt, isolated fourth ventricle

Introduction

Disproportionately large communicating fourth ventricle (DLCFV) was first proposed in 1980 as a subtype of hydrocephalus with dilation of all ventricles.11 A few cases of DLCFV have been reported but some were confused with the well-known clinical entity of isolated fourth ventricle (IFV). IFV is a condition with marked enlargement of the fourth ventricle due to obstruction of both the inlet and outlet of the fourth ventricle and mainly caused by shunt operation, whereas DLCFV is characterized by apparent patency of the aqueduct. Syringomyelia is characterized by the presence of abnormal fluid-filled cavities within the spinal cord. DLCFV is quite rare and DLCFV associated with syringomyelia is extremely rare.4,13 Spinal arachnoid cysts are relatively uncommon lesions that can be found in the intra- or extradural compartment of the spinal canal and only a few cases of syringomyelia associated with a spinal arachnoid cyst were reported.5,6,12 Here we report a case of DLCFV coexisting with syringomyelia and spinal arachnoid cyst, and discuss the mechanisms of these conditions.

Case Report

A 44-year-old woman underwent a ventriculoperitoneal shunt operation with a fixed middle pressure valve for hydrocephalus after subarachnoid hemorrhage due to ruptured right vertebral artery dissecting aneurysm. She presented with headache due to low cerebrospinal fluid (CSF) pressure, which was treated by pressure control with an adjustable valve. At this moment, the fourth ventricle was not dilated (Fig. 1A). After setting the valve pressure at 115 mmH2O, she suffered from nausea, vomiting, and gait disturbance, and computed tomography (CT) revealed dilation of all ventricles. By gradually decreasing the valve pressure to 45 mmH2O, the lateral and third ventricles became smaller, but the fourth ventricle remained dilated (Fig. 1B). Magnetic resonance (MR) imaging demonstrated patency of the aqueduct (Fig. 2) and the patient was diagnosed as DLCFV. Her nausea and vomiting decreased and gait became better, and she was discharged from the rehabilitation hospital 2 months later.

Four months later, the gait disturbance worsened again mainly due to truncal ataxia. CT revealed the whole ventricles were enlarged, especially the fourth ventricle, and her
consciousness level gradually deteriorated. Shunt malfunction was suspected and the shunt system was exchanged. No signs of CSF infection were noted. Her fourth ventricle was still dilated, although the other ventricles had remarkably decreased in size. T2-weighted MR imaging demonstrated an intradural arachnoid cyst from the C6 to T2 vertebral segments on the anterior aspect of the cord (Fig. 3A). The adjacent syringomyelia extending from the craniocervical junction through the C7 vertebral segment seemed to communicate with the fourth ventricle (Fig. 3A, B). CT ventriculography performed to check the shunt system also showed syringomyelia (Fig. 3C, D). Since her gait disturbance had improved, we gradually decreased the valve pressure to 60 mmH2O. CT revealed slit lateral ventricles with an enlarged fourth ventricle and a small subdural hematoma (Fig. 1C). The syringomyelia and arachnoid cyst were still present (Fig. 3E). Additional shunting of the fourth ventricle with an adjustable valve was performed and the valve pressures were independently controlled according to her neurological function and the size of the ventricles. The size of the fourth ventricle decreased (Fig. 1D) and the syringomyelia and spinal arachnoid cyst were also shrinking (Fig. 3F). Accordingly, her truncal ataxia was gradually relieved and she could walk, although her gait was slow and still wide-based, without a cane.

**Discussion**

Collision of CSF pulse waves causing a water hammer effect on the fourth ventricle is proposed to be the mechanism causing DLCFV. DLCFV is not well recognized or discriminated from IFV. Moreover, suspected transitional cases caused by functional obstruction of aqueduct have also been reported. Our case might be also a transitional type to IFV because additional shunting of the fourth ventricle was necessary.

Syringomyelia is generally believed to be related to disturbance of CSF flow. Classification of syringomyelia based on pathological findings and MR imaging includes communicating syringomyelia, noncommunicating syringomyelia, atrophic cavitations, and neoplastic cavitations. Our case was suggested to be communicating syringomyelia based on the findings of MR imaging. The observation that the size of syringomyelia changed according to the fourth ventricle size reinforced this hypothesis. Various mechanisms of syringomyelia have been proposed. The hydrodynamic theory is that obstruction...
Fig. 3  Sagittal (A) and axial (B) T2-weighted magnetic resonance images revealing intradural arachnoid cyst extending from the C6 to T2 vertebral segments on the anterior aspect of the cord and adjacent syringomyelia extending from the cerebrospinal junction through the C7 vertebral segment at a shunt pressure of 100 mmH2O. Computed tomography ventriculograms showing syringomyelia (C, D), and the intradural arachnoid cyst and syringomyelia still present at a shunt pressure of 60 mmH2O (E) but improved after shunting of the fourth ventricle (F).

The present very rare case of DLCFV coexisted with both syringomyelia and spinal arachnoid cyst. The syringomyelia and the arachnoid cyst decreased in size by shunting of the fourth ventricle, suggesting that the mechanism of the spinal arachnoid cyst formation in the present case was different from previous cases.

The present very rare case of DLCFV coexisted with both syringomyelia and spinal arachnoid cyst. The syringomyelia and the arachnoid cyst decreased in size and the symptoms were relieved by additional shunting of the fourth ventricle. Therefore, we considered that inadequately high pressure of the fourth ventricle might be the main cause of these conditions. However, the pathophysiology of these three diseases remains unclear and controversial, but we suspect these interesting conditions might interact in the presence of CSF flow disturbances. Although DLCFV, unlike the well-known IFV, is usually treated with adequate control of shunting pressure, additional shunting of the fourth ventricle should be considered when the symptoms persist despite adequate pressure control of the other ventricles.

References


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