Neurol Med Chir (Tokyo) 42, 140～142, 2002

Symptomatic Spinal Extramedullary Mass Lesion Secondary to Chronic Overdrainage of Ventricular Fluid

—Case Report—

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Abstract

A 69-year-old man presented with progressive nuchal pain and spastic gait 2 years after undergoing ventriculoperitoneal (VP) shunting for a pineal astrocytoma with obstructive hydrocephalus. The neurological manifestations were compatible with radiculomyelopathy caused by an upper cervical lesion. Magnetic resonance imaging showed an enhanced extramedullary mass lesion tightly constricting the upper cervical spinal cord. The pressure of the shunt system was 150 mmH_2O, and lumbar puncture revealed normal cerebrospinal fluid (CSF) pressure of 170 mmH_2O. After removal of the shunt system, the clinical symptoms and neuroradiological findings markedly improved. This symptomatic spinal mass lesion was thought to be formed secondary to chronic depletion of ventricular CSF through the VP shunt.

Key words: spinal mass lesion, meningeal enhancement, ventriculoperitoneal shunt, magnetic resonance imaging

Introduction

Magnetic resonance (MR) imaging has demonstrated cranial pachymeningeal enhancement and diffuse dural thickening in patients with intracranial hypotension,[3,5,7–9] and spinal dural enhancement in a few cases.[6,13] We describe a case of MR imaging evidence of a symptomatic localized mass lesion associated with a ventriculoperitoneal (VP) shunt. The spinal extramedullary mass lesion and clinical symptoms remarkably improved after removal of the shunt system.

Case Report

A 67-year-old man was admitted to our hospital in 1994 with a 2-week history of gait disturbance. MR imaging with contrast medium showed an enhanced mass in the pineal region with obstructive hydrocephalus (Fig. 1 *left column*). The histological diagnosis was pilocytic astrocytoma (grade II). Tumor progression occurred despite postoperative chemotherapy (4 cycles of a regimen of CBDCA-VP16) followed by radiotherapy (50 Gy), resulting in obstructive hydrocephalus. The patient underwent placement of a VP shunt with the programmable valve system set at 150 mmH_2O. The tumor was afterwards successfully palliated with chemotherapy using ACNU (Fig. 1 *right column*).

Two years after the VP shunt placement, he suffered progressive nuchal and shoulder pain, and spastic gait disturbance. Follow-up MR imaging showed a new enhanced extramedullary mass lesion tightly constricting the spinal cord at the upper cervical portion and cranial diffuse meningeal enhancement (Figs. 2 and 3 *left column*). The lesion was compatible with disseminated pineal glioma, primary spinal meningeal tumor, or other granulomatous disease involving the spinal dura. The shunt valve pressure was set at 150 mmH_2O. The cerebrospinal fluid (CSF) pressure on lumbar puncture was 170 mmH_2O. Although the ventricular size remained the same, we thought that the symptoms could be partly due to occult hydrocephalus caused by low drainage of the VP shunt. However, his condition worsened after reduction of the shunt

Received July 30, 2001; Accepted January 10, 2002
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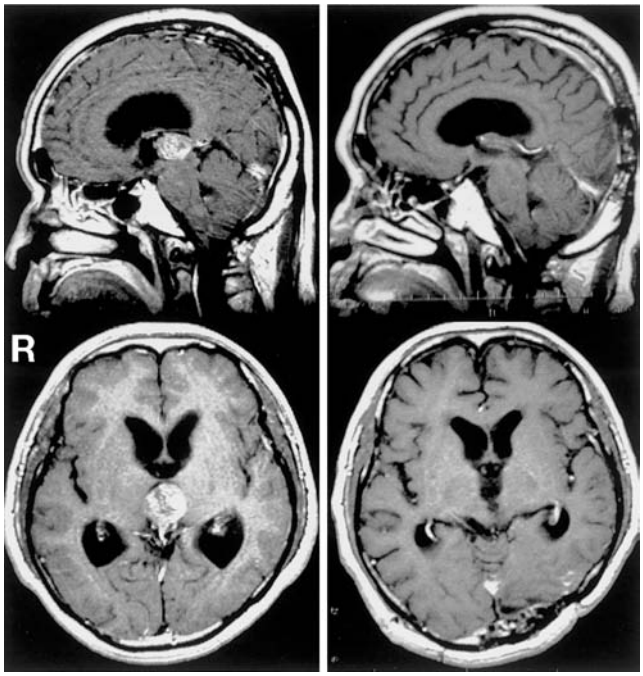

Fig. 1 **T_1-weighted magnetic resonance images with gadolinium on first admission (*left column*) showing an enhanced mass in the pineal region with obstructive hydrocephalus, and after radiotherapy and chemotherapy (*right column*) showing the tumor has disappeared.**

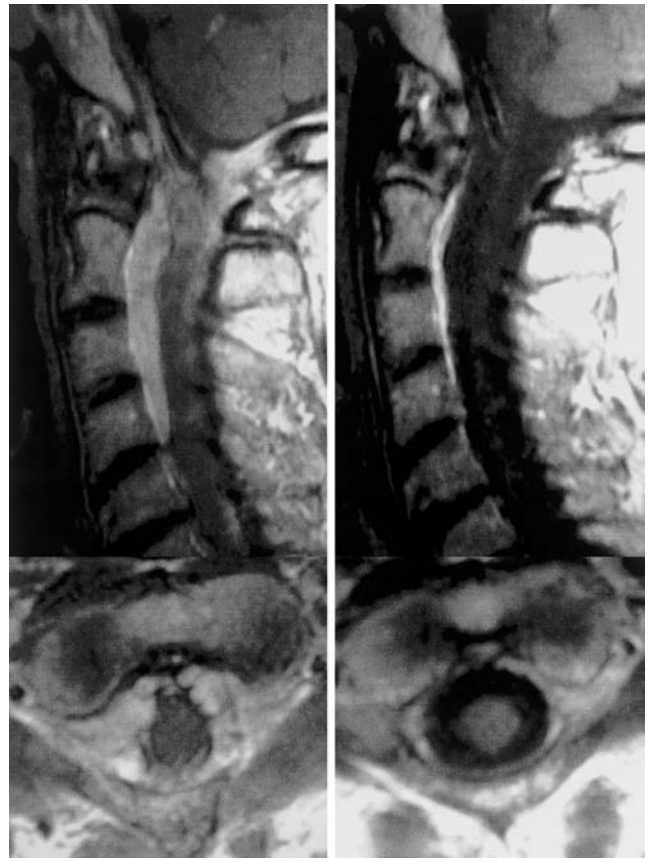

Fig. 2 **T_1-weighted magnetic resonance images with gadolinium 2 years after the ventriculoperitoneal shunt operation (*left column*) showing an extramedullary mass lesion constricting the spinal cord at the upper cervical portion, and one month after shunt removal (*right column*) showing the spinal mass lesion has disappeared. The axial images were obtained at the C-2 level.**

valve pressure from 150 to 120 mmH_2O. In contrast, his symptoms gradually improved after increasing the shunt valve pressure from 120 to 200 mmH_2O. The shunt system was removed and thereafter his nuchal pain and spastic gait remarkably improved. MR imaging after the shunt removal revealed marked reduction in the size of the extramedullary mass lesion (Fig. 2 *right column*). The cranial diffuse meningeal enhancement also disappeared (Fig. 3 *right column*).

Discussion

Spontaneous intracranial hypotension syndrome was first described in 1938.[10] Intracranial hypotension syndrome can be classified into five types according to the preceding event: 1) primary or spontaneous, 2) post-lumbar puncture, 3) post-head injury, 4) post-craniotomy, and 5) as a result of severe volume depletion.[1] VP shunt overdrainage can also cause intracranial hypotension syndrome.[2,4]

Intracranial hypotension syndrome is characterized by postural headache, low CSF pressure, and diffuse pachymeningeal enhancement on MR imaging.[11,12] MR imaging may also show subdural fluid collection and caudal displacement of the diencephalon and cerebellar tonsils, the so-called ''brain sagging.'' The pathogenic mechanisms of diffuse gadolinium enhancement of the meninges, formation of subdural fluid collection, and changes of CSF metabolism are still unclear. Biopsy of the meninges of patients with intracranial hypotension suggested that a thin subdural zone of fibroblasts and small thin-walled dilated blood vessels were the anatomical basis of pachymeningeal enhancement.[7]

Three cases of spontaneous intracranial hypotension with spinal dural enhancement were reported in 1998.[6] However, the present case involved symptomatic spinal mass lesion in association with intracranial hypotension syndrome.

In our case, the shunt valve pressure and the CSF

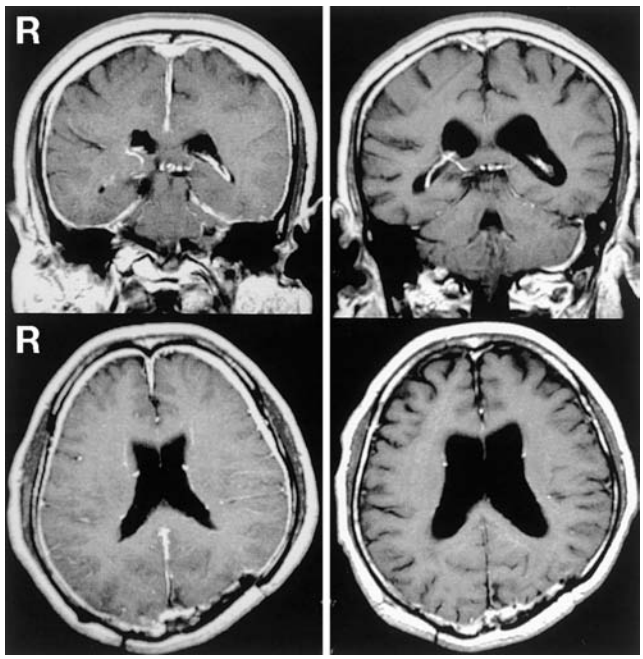

Fig. 3 **T_1-weighted magnetic resonance images with gadolinium 2 years after the ventriculoperitoneal shunt operation (*left column*) showing diffuse meningeal enhancement, and one month after shunt removal (*right column*) showing no meningeal enhancement.**

pressure on lumbar puncture may not have been compatible with the intracranial hypotension syndrome. However, the clinical symptoms and neuroradiological findings had remarkably improved after removal of the shunt system. Therefore, we speculate that the focal meningeal mass formation and the diffuse meningeal enhancement were caused by intermittent intracranial hypotension due to postural changes of CSF pressure. Focal meningeal thickening, which was caused by low CSF pressure, could have provoked radiculomyelopathy at the upper cervix.

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