

Isolated Third and Fourth Ventricles Associated With Neurosarcoidosis Successfully Treated by Neuroendoscopy

—Case Report—

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Abstract

A 48-year-old woman presented with a unique case of isolated third and fourth ventricles associated with neurosarcoidosis, manifesting as upward gaze palsy. She had a past history of surgery for intracranial sarcoid granuloma 10 years previously and placement of a ventriculoperitoneal shunt 3 years later. Magnetic resonance imaging revealed isolated third and fourth ventricles. Endoscopic plasty of the right foramen of Monro, fenestration of the septum pellucidum, and revision of the ventriculoperitoneal shunt were performed. Postoperatively the size of the third and fourth ventricles was reduced, and the upward gaze palsy resolved. Endoscopic treatment is less invasive and effective for the treatment of multiple septum formation in the ventricular system.

Key words: neurosarcoidosis, isolated third and fourth ventricles, neuroendoscopy

Introduction

Hydrocephalus with multiple septations and compartments has been difficult and sometimes impossible to treat with the usual technique of shunt placement. Various radical approaches to restore the intraventricular circulation of the cerebrospinal fluid (CSF), such as fenestration of the septum pellucidum or intraventricular septation, and foraminal plasty, have been used in patients with hydrocephalus associated with intraventricular septum formation.

We treated a patient with isolated third and fourth ventricles associated with neurosarcoidosis by endoscopic fenestration of the isolated compartments.

Case Report

A 48-year-old woman complained of upward gaze palsy, and consulted our hospital. She had undergone surgery for sarcoid granuloma in the right frontal lobe 10 years previously, and a ventriculoperitoneal shunt was inserted 3 years after the first operation. On admission, she had upward gaze

palsy, but no other neurological deficits. Chest radiography disclosed bilateral hilar lymphadenitis compatible with sarcoidosis. Computed tomography revealed isolated dilation of the third and fourth ventricles, and slit-like lateral ventricle. Magnetic resonance imaging also revealed isolated dilation of the third and fourth ventricles with a patent aqueduct (Fig. 1). No abnormal enhanced lesions were demonstrated. We assumed that this isolation of the third and fourth ventricles was due to obstruction of the bilateral foramina of Monro and outlets of the fourth ventricle associated with neurosarcoidosis.

Endoscopic fenestration of the isolated compartments was planned. A standard coronal burr-hole was made. A peelaway sheath (14.0 F; Codman, Inc., Raynham, Mass., U.S.A.) was passed into the anterior horn of the right lateral ventricle. A flexible fiberoptic ventriculoscope (14.0 F; Codman, Inc.) was then inserted into the ventricle through the peelaway sheath. The right foramen of Monro was occluded with membranous tissue resembling a check valve (Fig. 2 *left, center*). Therefore, blunt plasty of the right foramen of Monro was performed using the endoscope. The endoscope was then introduced into the third ventricle. The aqueduct was found to be adequately patent into the fourth ventricle. The endoscope was carefully passed

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through the aqueduct. The outlets of the fourth ventricle were found to be occluded. Then, fenestration of the septum pellucidum between the anterior and posterior septal veins was performed with low-current monopolar coagulation and an attempt was made to simplify the ventricular system. The left foramen of Monro was occluded as expected (Fig. 2 right). The ventriculoperitoneal shunt was revised through the right anterior horn after the endoscopic procedures.

Postoperatively, symmetrical reduction in the size of the ventricular system was observed (Fig. 3) and her upward gaze palsy gradually resolved.

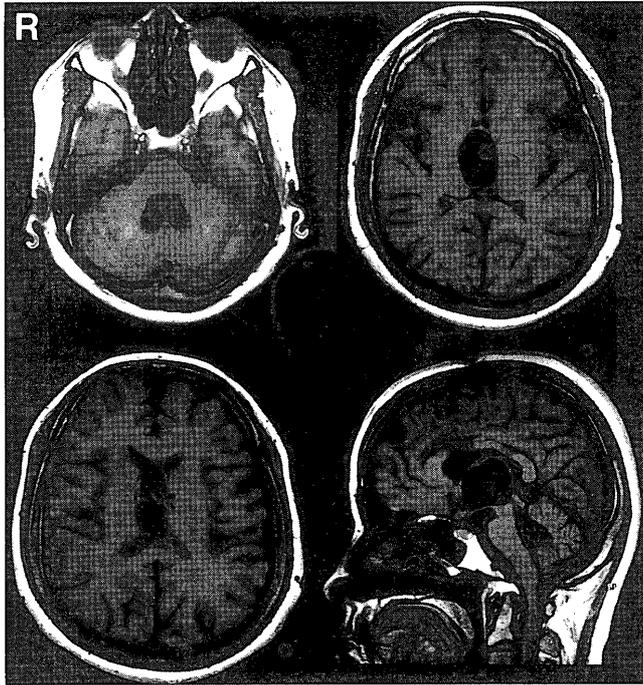


Fig. 1 Preoperative magnetic resonance images showing isolated dilation of the third and fourth ventricles.

Discussion

Sarcoidosis is a common, chronic granulomatous disease, but the etiology remains uncertain. Neurosarcoidosis occurs in 5–15% of cases of sarcoidosis.⁸⁾ Neurosarcoidosis can be divided into that of the central nervous system and that of the peripheral nervous system. Meningitis, granulomas, disturbance of the hypothalamus and pituitary gland, and hydrocephalus are the main manifestations of neurosarcoidosis in the central nervous system.^{1,12)} Hydrocephalus was involved in 6% of patients with neurosarcoidosis in a prospective study.¹⁾ Both obstructive and communicating hydrocephalus occur with neurosarcoidosis, and the main cause of hydrocephalus is granulomatous meningitis.^{2,13)}

In the present case, the ventriculoperitoneal shunt was effective after the shunt operation. However, judging from the operative findings, both shunt malfunction occurred and the ventricular system

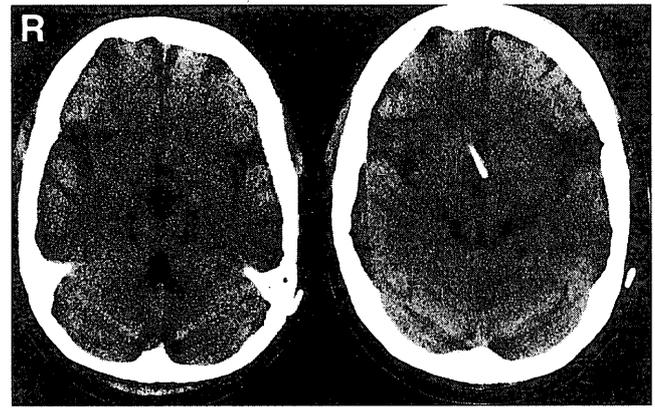


Fig. 3 Postoperative computed tomography scans showing symmetrical reduction in the size of the ventricular system.

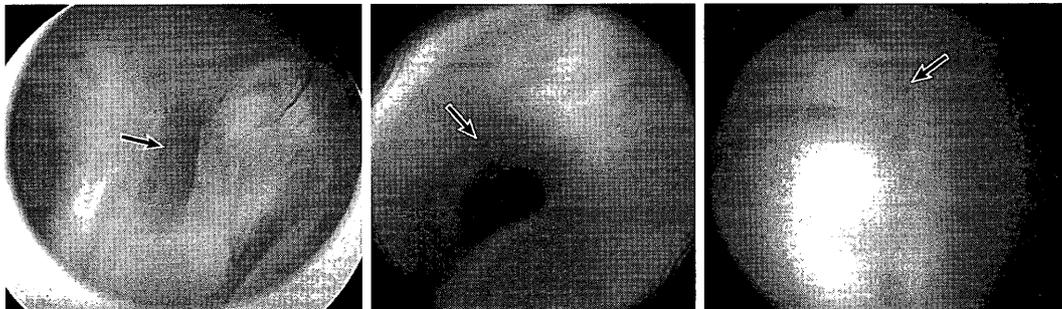


Fig. 2 Endoscopic views demonstrating the occluded right foramen of Monro resembling a check valve (arrows; left, closed; center, open) and the occluded left foramen of Monro (arrow, right).

developed complicated and isolated compartments. The right foramen of Monro was incompletely obstructed by a check-valve mechanism, the left foramen of Monro was completely blocked, and the outlets of the fourth ventricle were also occluded. CSF produced in the left lateral ventricle was drained by the inserted shunt, but the CSF produced in the right lateral ventricle passed one way through the right foramen of Monro into the third ventricle due to the check-valve mechanism. Obstruction of the right foramen of Monro was incomplete, so isolation of the third and fourth ventricles might occur. The dilated third ventricle had compressed the superior colliculus and caused the upward gaze palsy. After the endoscopic procedures, the upward gaze palsy gradually resolved, accompanied by reduction in the size of the isolated third and fourth ventricles.

Secondary hydrocephalus following meningitis, ventriculitis, or hemorrhage may be complicated by septum formation in the ventricular system, resulting in complex manifestations of CSF circulation disturbance.^{3,7,11)} Treatment of hydrocephalus with multiple septations and compartments has been difficult or impossible with normal shunt placement. Although loculated hydrocephalus is often treated by placement of an additional shunt, multiple shunt placement is complicated and often results in shunt malfunction. Recent advances in neuroendoscopic procedures now allow various techniques, such as third ventriculostomy, fenestration of the septum pellucidum or intraventricular septation, aqueductal plasty, and plasty of the foramen of Monro.^{4-6,9,10,14)}

Endoscopic procedures in our case of hydrocephalus associated with neurosarcoidosis obtained good results. Endoscopic treatment is less invasive and effective for the treatment of multiple septum formation in the ventricular system.

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