Benign aqueductal cyst causing bilateral internuclear ophthalmoplegia after external ventricular drainage

Case report

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The introduction of magnetic resonance (MR) imaging to the field of neuroimaging has allowed detection of various lesions that cause aqueductal stenosis. The authors report the case of a 3-year-old boy in whom a benign ventricular cyst developed in the aqueduct. The patient became drowsy after having complained of headache and vomiting; MR imaging revealed mild triventricular dilation and a normal-sized fourth ventricle. Repeated MR imaging performed 1 week later revealed an aqueductal cyst that had markedly enlarged during the intervening period. An external ventricular drainage system was installed, but recovery of consciousness in the child was unsatisfactory and a new bilateral internuclear ophthalmoplegia developed. Fenestration of the cyst wall and placement of a ventriculocisternostomy in the third ventricle were performed simultaneously by using a flexible neuroendoscope. By 2 weeks postsurgery, the patient’s neurological symptoms had completely resolved. This case illustrates that simple rerouting of ventricular cerebrospinal fluid (CSF) can aggravate the symptoms of this rare lesion by causing severe compression of periaqueductal structures by a cyst that maintains a high intracystic pressure. Endoscopic surgery was an excellent choice of treatment to achieve both cyst fenestration and normalization of intracranial CSF pressure by creating a ventriculocisternostomy.

KEY WORDS • aqueductal cyst • internuclear ophthalmoplegia • neuroendoscope

INTERNUCLEAR ophthalmoplegia is caused by various diseases that can selectively affect the MLF. Frequently encountered pathological entities causing INO in adults include brainstem infarctions and traumatic brainstem injuries, whereas in younger adults multiple sclerosis is the primary cause. Although uncommon, children can also present with INO as an early sign of brainstem glioma.

In this report, we present the case of a 3-year-old boy who had obstructive hydrocephalus caused by a benign ventricular cyst developing in the aqueduct. Following installation of an external ventricular drainage system, the patient experienced bilateral INO, which was treated successfully with the aid of a flexible neuroendoscope.

Case Report

History. This 3-year-old boy experienced consciousness disturbance following a bout of progressive headache and vomiting and was admitted to our institution. On admission, he was drowsy, but did not display any focal neurological deficit.

Abbreviations used in this paper: CSF = cerebrospinal fluid; CT = computerized tomography; INO = internuclear ophthalmoplegia; MLF = medial longitudinal fasciculus; MR = magnetic resonance.

Examination and Initial Treatment. Computerized tomography scanning of the brain revealed mild triventricular dilation, although the fourth ventricle was normal in size. Sagittal MR imaging demonstrated a slightly elevated mesencephalic tectum (Fig. 1), and the results of a dynamic flow study of cine MR imaging suggested that there was decreased flow of CSF. The child’s consciousness

Fig. 1. Sagittal T1-weighted MR image revealing mild dilation of the third ventricle and a patent aqueduct.
Benign aqueductal cyst

Improved gradually in response to glycerol, which was administered during the first several days. On an MR image obtained 1 week later, however, the patient’s aqueduct and the rostral portion of his fourth ventricle exhibited marked dilation (Fig. 2). No abnormally enhancing lesions were observed after administration of gadolinium–diethylene-triamine pentaacetic acid. The patient’s consciousness slowly deteriorated after this period, and an external ventricular drainage system was installed. After installation, his consciousness improved slightly, but he experienced a new bilateral disturbance of eye adduction on lateral gazes, which was accompanied by monocular nystagmus in the abducting eye. Computerized tomography ventriculography performed through the drainage tube demonstrated a defect filling with contrast medium in the dilated aqueduct; retention of the contrast medium lasted longer than 24 hours (Fig. 3). These findings indicated that there was a cystic lesion located in the aqueduct that caused CSF flow obstruction as well as compression of the periaqueductal brainstem region. We therefore selected endoscopic surgery to open the cyst and decompress the periaqueductal region.

Operation. A 4-mm flexible neuroendoscope (model NEU4L; Machida-Endoscope, Tokyo, Japan) was introduced through a right frontal burr hole into the third ventricle. Using the neuroendoscope, we could clearly view the semitransparent cystic lesion. The cyst appeared to have originated from the pineal recess and herniated into the aqueduct (Fig. 4). The cyst was opened using a biopsy forceps and a clear CSF-like fluid was released. A standard ventriculocisternostomy was placed in the third ventricle and the ventricular drainage tube was removed. A small piece of the cyst wall was resected during surgery and found to contain fibrous tissues but no tumor cells or glial tissue.

Postoperative Course. The patient’s consciousness became clear the day following surgery and the bilateral INO completely resolved by 14 days postsurgery. An MR image obtained 3 weeks postsurgery demonstrated ventricles that were normal in both size and shape, closure of the aqueduct, and patent CSF flow through the ventriculocisternostomy. The patient was discharged from the hospital without neurological deficit and he has remained this way. An MR image obtained 2 years postsurgery did not demonstrate ventricular dilation or cyst recurrence (Fig. 5).

Discussion

Because MR imaging enables detailed anatomical depiction of periaqueductal regions, nonneoplastic space-occupying lesions have increasingly been identified as causes of aqueductal occlusion. Ventricular cysts are one such lesion, but they rarely occur within the aqueduct itself. Although the natural history of these CSF-containing cysts still remains unclear, they occasionally display progressive expansion and become symptomatic. Among various hypotheses proposed to explain cyst growth, a one-way valve mechanism has been gaining greater popularity. In this model, a cyst receives more pressure from surrounding structures as it grows larger, which, in turn, causes more efficient closure of the
CSF communication, thus functioning as a valve. In the present case, we also speculated that the cyst became tightly fixed in the aqueduct and enlarged by the one-way valve mechanism. One possible explanation for the acute development of INO after ventricular drainage in our case would be that a sudden decrease in ventricular pressure induced a tighter contact of the cyst wall with periaqueductal structures, which caused compression of MLF fibers.

A few cases of bilateral INO associated with hydrocephalus have been reported in patients with Arnold-Chiari malformations. Magnetic resonance imaging in these patients did not demonstrate definite abnormal intensity changes, and the mechanism causing this specific symptom in those patients has remained unclear. Because the placement of ventriculoperitoneal shunts universally resolved their patients' symptoms, the authors of these studies speculated that direct compression by a distended ventricular system might have caused dysfunction of MLF fibers. In patients with aqueductal stenosis, rapid inversion of transtentorial pressure occasionally induces more severe functional impairment of the periaqueductal gray matter by increasing neuronal distortion. Similarly, the rapid inversion of the transtentorial pressure gradient may have contributed to the delayed recovery of consciousness and the new ophthalmological manifestations in our case as well.

Knowledge of such peculiar pathophysiological characteristics of the aqueductal cyst is essential for appropriate surgical treatment. Opening of the cyst should be performed to alleviate direct compression by the cyst, in addition to taking measures to decompress the ventricles at the same time. For this purpose, as shown in the present case, endoscopic surgery would be the best method because it allows the surgeon to perform two procedures—cyst opening and placement of a third ventriculocisternostomy—simultaneously with minimum intervention.

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