Spontaneous third ventriculostomy: definition by endoscopy and cerebrospinal fluid dynamics

Case report

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Chronic obstructive hydrocephalus is known to cause ventricular diverticula and, rarely, spontaneous ventriculostomy. The authors present the case of a patient in whom a spontaneous third ventriculostomy was identified with long-standing hydrocephalus secondary to aqueductal stenosis. To their knowledge, this is the first report in which a spontaneous stoma in the floor of the third ventricle was evaluated using endoscopy and cerebrospinal fluid dynamics studies. Both studies confirmed that the spontaneous stoma is similar in structure and function to surgical third ventriculostomy. (DOI: 10.3171/2008.5.JNS08286)

Key Words • endoscopy • resistance to cerebrospinal fluid outflow • spontaneous third ventriculostomy

Operation. As the patient’s headache persisted, further evaluation was indicated. An endoscopic examination of the ventricular system showed absence of the septum pellucidum. The foramina of Monro appeared normal. The third ventricle was dilated, and a stoma was evident in its floor. This was situated midway between the mammillary bodies and the tuber cinereum, along the thinnest part of the floor, to the left of the midline (Fig. 1c). The edges of the stoma flapped gently, indicating bulk CSF flow. The flexible endoscope was advanced through the stoma, entering the subarachnoid space anterior to the basilar artery (Fig. 1d). A ventricular access device was inserted.

Postoperative Course. Six weeks postoperatively, 24-hour ICP monitoring was performed through the access device. A fluid-filled pressure transducer was zeroed to the level of the external auditory meatus. The ICP ranged from −9.4 to 10.3 mm Hg (mean −1.32 ± 2.97 mm Hg [SD]). A computerized constant flow ventricular infusion study was subsequently performed through the access device (Fig. 2), as described and validated by Czosnyka et al. The baseline pressure was 2.4 mm Hg. A plateau pressure of 8.0 mm Hg was obtained after infusion of 52 ml of normal saline at 0.5 ml/minute. Resistance to CSF outflow was calculated at 3.74 mm Hg/ml/minute, using

Abbreviations used in this paper: CSF = cerebrospinal fluid; ICP = intracranial pressure.
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customized software (ICM+, Cambridge University). The patient’s headaches subsequently resolved, and no further symptoms were reported on review at 6 months.

Discussion

Spontaneous ventriculostomy has been reported in a number of locations, including the atrium of the lateral ventricle,1,8,13,15 the suprapineal recess,2,15 and the floor of the third ventricle.5,11,14,18 It occurs in the context of chronic hydrocephalus, when chronic high-pressure CSF pulsation acts against thin regions of the ventricular wall and results in transmural penetration and rupture of the ependymal wall into the subarachnoid space. Ventricular diverticula share the same pathophysiology and represent local herniation of the ventricle; they occur more often than spontaneous stomas and have been reported in up to 25% of patients with severe and long-standing hydrocephalus.6,12,16 Of the reported ventricular diverticula, 62% originate from the inferomedial wall of the atrium, where the ventricular lumen is separated from the subarachnoid space by the thin white matter of the hippocampal alveus alone.7 Diverticular walls may be composed of ependyma, parenchyma, and pia and arachnoid layers, but occasionally only the pia and the arachnoid may be in continuity if the ependyma and the parenchyma are interrupted by progressive ventricular dilation.10 Persistence of elevated intraventricular pressure leads to rupture of the diverticulum with formation of a ventriculostomy. Both entities have been identified on pneumoencephalography4 or contrast ventriculography.2,7,8,10,11,19 Magnetic resonance imaging is useful in their diagnosis and accurate anatomical definition.14 Flow artifacts are evident on T2-weighted and FLAIR images; phase contrast cine MR imaging positively identifies CSF flow across the stoma. Occasionally loss of integrity of neural tissue or ventricular wall is visible on T1-weighted images.18

Eight cases of spontaneous ventriculostomy through the floor of the third ventricle have been described (Table 1).5,9,11,14,18 The median age at presentation was 34.5 years. In 2 patients, chronic hydrocephalus was related to aqueductal stenosis. The most prevalent symptom was long-standing intermittent headache. Symptoms resolved spontaneously in all but 1 patient in whom an opening pressure of 220 mm H2O on lumbar puncture prompted insertion of a ventriculoperitoneal shunt.18 The stoma was visual-

Fig. 1. a: Sagittal T1-weighted MR image showing the defect in the floor of the third ventricle (arrow). b: Phase-contrast cine MR image. The arrow points to flow through the floor of the third ventricle. c: Axial T2-weighted MR image showing enlargement of the lateral ventricles. d: Endoscopic view of the spontaneous stoma (arrow). The asterisks denote the mammillary bodies.

Fig. 2. Computerized ventricular infusion study showing a change in ICP (upper) and amplitude of the ICP wave (lower) during baseline (white area) and infusion (shaded area).
ized in 1 patient only at postmortem examination.\textsuperscript{11} The opening was situated in the weakest part of the floor of the third ventricle; it was 3 mm in diameter and was patent into the subarachnoid space. This is similar to the endoscopic description in our patient and appears identical to surgical endoscopic third ventriculostomy. Endoscopy has only been reported in one other case of ventriculostomy where hydrocephalus secondary to aqueduct stenosis was associated with a cystic lesion in the supracerebellar region.\textsuperscript{13} A flexible endoscope was advanced from the right lateral ventricle through an ostium in its medial wall behind the choroid plexus. The upper surface of the cerebellum and veins could be identified passing through the cyst. The presence of a membrane around the entire cyst could not be confirmed, suggesting that the diverticulum had ruptured and communicated directly with the subarachnoid space. This is similar to the endoscopic third ventricle; it was 3 mm in diameter and was patent into the subarachnoid space. This is similar to the endoscopic description in our patient and appears identical to surgical endoscopic third ventriculostomy. Endoscopy has only been reported in one other case of ventriculostomy where hydrocephalus secondary to aqueduct stenosis was associated with a cystic lesion in the supracerebellar region.\textsuperscript{13} A flexible endoscope was advanced from the right lateral ventricle through an ostium in its medial wall behind the choroid plexus. The upper surface of the cerebellum and veins could be identified passing through the cyst. The presence of a membrane around the entire cyst could not be confirmed, suggesting that the diverticulum had ruptured and communicated directly with the subarachnoid space. The size of the diverticulum and the ventricles was reduced after endoscopic third ventriculostomy.

Most cases of spontaneous ventriculostomy in other locations are associated with spontaneous resolution of symptoms.\textsuperscript{2,10} However, further surgical management has occasionally been necessary. One patient with a spontaneous ventriculostomy through the suprapineal recess demonstrated arrest of his hydrocephalus; however, his symptoms only regressed after a CSF shunt was inserted.\textsuperscript{17} This would imply that long-term close observation is indicated if the symptoms do not regress completely. Some authors have used continuous ICP monitoring to exclude persisting hydrocephalus.\textsuperscript{16} In our patient, the mean ICP over a 24-hour period was within normal limits. In view of our patient’s age and imaging findings, the resistance to CSF outflow before ventriculostomy was expected to be ~ 14 mm Hg/ml/min.\textsuperscript{3} Tisell et al.\textsuperscript{15} used a constant flow infusion test to evaluate the resistance to CSF outflow before and after endoscopic third ventriculostomy in 15 adults with aqueductal stenosis. The mean ± SD preoperative intraventricular resistance to CSF outflow was 17.1 ± 9.1 mm Hg/ml/min, and the mean postoperative value was 9.8 ± 4.6 mmHg/ml/min. Although we were unable to determine resistance to CSF outflow prior to spontaneous ventriculostomy in our patient, these values would suggest that our patient’s CSF absorption had been reduced to normal by the spontaneous ventriculostomy.

**Conclusions**

To our knowledge, this is the first description of a spontaneous ventriculostomy occurring at the floor of the third ventricle that has been evaluated by endoscopy and CSF dynamics studies. Both investigations confirm that the spontaneous stoma is structurally and functionally similar to surgical third ventriculostomy.

**Disclaimer**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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