Spontaneous third ventriculocisternostomy in an infant with obstructive hydrocephalus

Case report

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Spontaneous ventriculocisternostomy, the spontaneous communication between the ventricular system and the subarachnoid space, is rare. The authors report a case of an infant with obstructive hydrocephalus who developed a spontaneous third ventriculocisternostomy. The infant was initially evaluated for progressive ventriculomegaly and increasing head circumference (HC). During follow-up, the patient’s HC began to follow percentile lines and magnetic resonance (MR) imaging demonstrated a reduction of the hydrocephalus. Flow-sensitive phase-contrast cine MR images revealed cerebrospinal fluid (CSF) flow through the floor of the third ventricle between the tuber cinereum and the mammillary bodies connecting the ventricular system with the prepontine cistern. Although rare, clinicians should be cognizant of this phenomenon as it may eliminate the need for CSF diversion. (DOI: 10.3171/PED/2008/1/6/477)

KEY WORDS • cine magnetic resonance imaging • obstructive hydrocephalus • spontaneous ventriculocisternostomy • spontaneous ventriculostomy

Abbreviations used in this paper: CSF = cerebrospinal fluid; HC = head circumference; MR = magnetic resonance.

Spontaneous ventriculocisternostomy refers to the spontaneous communication between the ventricular system and the subarachnoid space. This phenomenon is associated with obstructive hydrocephalus and may result in the arrest or reduction of hydrocephalus. Well-documented cases of spontaneous ventriculocisternostomies are rare and have been infrequently reported in the English literature. In this report, we describe a case of an infant with obstructive hydrocephalus secondary to a tectal mass who developed a spontaneous third ventriculocisternostomy, as documented by flow-sensitive phase-contrast cine MR imaging, with subsequent improvement of her hydrocephalus.

Case Report

This female infant was referred to our clinic at the Prince of Wales Private Hospital at the age of 2 months for evaluation of hydrocephalus. She was born after 36 weeks’ gestation weighing 2475 g and with an HC of 32 cm. A prenatal ultrasonographic examination at 35 weeks and 5 days’ gestation had revealed borderline ventriculomegaly. The patient had been discharged home after an uncomplicated postnatal course. When the patient was 6 weeks of age, her HC was 40.1 cm. An ultrasonographic examination of her head demonstrated dilated third and lateral ventricles.

On initial neurosurgical examination, her fontanelle was full but not tense, the sutures were slightly split, and the scalp veins were slightly dilated. She did not have sunsetting eyes and was feeding well. The patient was thought to require a shunt. A brain MR imaging study was scheduled, and surgery was planned to follow review of the MR images. When the patient was reexamined at 3 months of age, after the MR imaging study had been performed, however, we found that she was still gaining weight and developing along normal milestones. Her HC was slightly above the 95th percentile and her fontanelle was full but soft and the sutures were no longer split. The MR images showed marked hydrocephalus (greater on the left side than the right) secondary to a tectal mass and subsequent aqueductal stenosis, and flow-sensitive phase-contrast cine MR images confirmed the absence of flow through the stenotic aqueduct (Fig. 1). In view of the patient’s clinical stability, the fact that her HC only diverged slightly from the 95th percentile, and her relatively young age, shunt placement was delayed and she was followed up closely with both neurological and imaging examinations.

When the patient was 4 months of age, her HC measured 46.5 cm. Although her HC was above the 95th percentile, she remained clinically stable with a full but soft fontanelle, and an ultrasonographic examination of her head demon-
strated stable ventriculomegaly. Consequently, a CSF diversion procedure was again delayed and she was monitored closely.

When she was 7 months of age, her HC was still above the 95th percentile (48.1 cm) but the disparity was lessening, and her fontanelle was open, soft, and sunken. Consequently, no surgical intervention was performed.

At the 13- and 20-month examinations, her HC measured 50.2 cm and 51 cm, respectively, both above the 95th percentile, but following along percentile lines. An MR imaging study performed when she was 19 months of age demonstrated some resolution of her ventriculomegaly. Sagittal T1-weighted images demonstrated a defect in the floor of the third ventricle between the tuber cinereum and the mammillary bodies, and flow-sensitive phase-contrast cine MR sequences revealed an abnormal CSF wave through the floor of the third ventricle between the tuber cinereum and the mammillary bodies connecting the ventricular system with the prepontine cistern (Fig. 2). There was no significant change in the size of the tectal mass, which most likely represented a tectal hamartoma.

At 44 months of age, the patient’s HC was 54 cm, MR imaging demonstrated further resolution of her ventriculomegaly (Fig. 3), and she was achieving age-appropriate developmental milestones.

Discussion

Spontaneous ventriculocisternostomy is a rare condition that results from the spontaneous rupture of the ventricular wall and pia mater with subsequent communication between the ventricular system and the subarachnoid space. In the setting of a competent CSF absorptive mechanism, this communication can bypass the CSF obstruction and result in symptomatic improvement and arrest or reduction of hydrocephalus. In this report, we describe a case involving an infant with obstructive hydrocephalus secondary to a tectal mass who developed a spontaneous third ventriculocisternostomy. This spontaneous third ventriculocisternostomy most likely occurred when the infant was between 3 months of age (when cine MR images demonstrated no CSF flow through the floor of the third ventricle) and 7 months of age (when the slope of the patient’s HC curve changed and began to follow percentile lines).

The first reported clinicopathological description of a spontaneous communication between the ventricular system and the subarachnoid space was reported by de Lange. In that report, the author described a child with hydrocephalus whose HC reached a maximum of 73 cm at 5.5 months of age and then gradually and spontaneously diminished in size. When the child was 22 months old, her HC measured 51 cm. She died of pneumonia the following month, and on postmortem examination she was found to have an aqueductal obstruction and highly distended lateral and third ventricles. Extending from each lateral ventricle were dorso-laterally directed channels, with the right channel reaching the surface of the brain, and serving as a conduit for CSF drainage into the subarachnoid space. Since this initial description, well-documented cases of spontaneous ventriculocisternostomies have been occasionally reported in the
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Most cases of spontaneous ventriculocisternostomy have been described at either autopsy or by ventriculography. Rovira et al. and Kim et al. have described patients with spontaneous ventriculocisternostomies with flow-sensitive phase-contrast cine MR imaging. Rovira et al. reported on 3 patients (17, 28, and 67 years old) with obstructive hydrocephalus due to aqueductal stenosis who each had a spontaneous ventriculocisternostomy through the floor of the third ventricle. Intracranial pressure monitoring was performed in 2 of the 3 patients and the mean pressure was <12 mm Hg; the third patient was completely symptom free. Kim et al. described a 59-year-old woman with a tectal glioma and chronic hydrocephalus who noted spontaneous relief of her headaches during follow-up. Phase-contrast MR imaging demonstrated a spontaneous ventriculocisternostomy through the floor of the third ventricle. To the best of our knowledge, the patient in our case is the youngest in whom a spontaneous ventriculocisternostomy has been confirmed by flow-sensitive, phase-contrast cine MR imaging.

Conclusions

In this report, we describe an infant with obstructive hydrocephalus secondary to a tectal mass who developed a spontaneous third ventriculocisternostomy with associated reduction in her hydrocephalus. Although spontaneous third ventriculocisternostomy is rare, clinicians should be aware of the phenomenon because it may, as in this particular case, obviate the need for a CSF diversion procedure.

References

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