Unilateral hydrocephalus secondary to congenital atresia of the foramen of Monro

Case report

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Unilateral hydrocephalus is an uncommon entity which results from obstruction at the level of the foramen of Monro. It is usually brought about by tumors or inflammatory conditions. Congenital maldevelopment of the foramen of Monro is an often postulated, yet never proven, cause of unilateral ventricular enlargement. A case of unilateral hydrocephalus secondary to congenital atresia of the foramen of Monro is presented to document the occurrence of this condition.

KEY WORDS • foramen of Monro • herniation • unilateral hydrocephalus • ventriculoperitoneal shunt • ventriculoscopy

Unilateral hydrocephalus is a unique condition resulting from obstruction at one foramen of Monro. Neoplasms in the lateral or third ventricle and acute or chronic inflammatory conditions are the usual causes. While congenital atresia of the foramen of Monro is often mentioned in connection with unilateral hydrocephalus, there has not been a report documenting congenital obstruction at this level of the ventricular system.

A case is presented in which severe obstructive unilateral hydrocephalus developed in an infant secondary to congenital atresia of the foramen of Monro.

Case Report

This previously healthy 11-month-old baby boy presented to a local hospital with a 2-day history of fever, lethargy, and vomiting. The infant's delivery and neonatal development had been normal. The head circumference at birth was 35 cm (75th percentile). During the 3 months before admission, the infant's parents had noted a turning outward of the right eye which was attributed to strabismus. In the week prior to the child's acute illness, the parents had noticed drooping of the right eyelid. A diagnosis of meningitis was entertained at the local hospital, and a lumbar puncture was performed. Thirty minutes after the lumbar puncture, the child became apneic. The child was intubated and given mannitol, then transferred to Children's Hospital of Pittsburgh.

Examination. On arrival approximately 2 hours after the episode of apnea, the child was awake and alert, and had extubated himself. The occipitofrontal head circumference was 47 cm (95th percentile for his age). The right eye appeared proptotic and a partial right third-nerve palsy was evident. Hemorrhagic papilledema was present bilaterally. Plain skull radiographs demonstrated split cranial sutures and an enlarged right hemicranium. A computerized tomography (CT) scan showed massive dilatation of the right lateral ventricle. There was associated subfalcial herniation (Fig. 1). A contrast-enhanced CT scan did not show any parenchymal abnormalities.

FIG. 1. Computerized tomography scans showing massive dilatation of the right lateral ventricle with associated subfalcial herniation. No enhancing lesions are visible.
Operation. The infant was taken to the operating room where ventriculoscopy of the right lateral ventricle was performed. The septum pellucidum was intact. The location of the foramen of Monro was identified by tracing the thalamostriate vein and the septal vein to their point of convergence. The foramen was replaced, however, by an indentation covered by smooth, glistening tissue which resembled normal ependyma. No tumor masses or granulations were present. The choroid plexus appeared normal. A ventriculoperitoneal shunt was placed.

Cerebrospinal fluid (CSF) from the right lateral ventricle showed 8 red blood cells, no white blood cells, a protein content of 40 mg%, and a glucose level of 100 mg%. Lumbar CSF had no red blood cells, 2 lymphocytes, a protein level of 46 mg% and a glucose concentration of 90 mg%. Titers for toxoplasmosis, rubella, cytomegalovirus, and herpes (TORCH) were negative.

Postoperative Course. The infant’s recovery was uneventful in the postoperative period. The third-nerve palsy resolved completely within 5 days of shunting. At follow-up review 6 months after surgery, the child was neurologically and developmentally normal. A CT scan done at that time showed a significant decrease in the size of the shunted ventricle. The opposite lateral ventricle was normal in size and there were no contrast-enhancing abnormalities.

Discussion

Unilateral hydrocephalus with dilatation of one lateral ventricle is an infrequently reported occurrence. The concept of unilateral hydrocephalus was first put forth by Von Mohr in 1842. He reported two patients with unilateral ventricular dilatation and associated ipsilateral hemispheric atrophy. The foramen of Monro was patent in both cases. This led to confusion in the early literature, and many cases of hydrocephalus ex vacuo (atrophic shrinkage of adjacent brain substance with compensatory ventricular dilatation) were given the label of unilateral hydrocephalus or partial internal hydrocephalus.

In 1913, Thomas was able to produce experimental unilateral hydrocephalus by injection of Aleuronat into the lateral ventricle, causing dense scarring around the foramen of Monro. In 1917, Dandy also produced an experimental model of unilateral hydrocephalus by blocking one foramen of Monro with a piece of fascia.

Obstruction at the foramen of Monro is usually due to thalamic neoplasms, pedunculated intraventricular tumors, tuberculous masses, colloid cysts, ventriculitis, or nonspecific inflammatory conditions. The possibility that the foramen of Monro, which is 2 to 4 mm in diameter, might be congenitally atretic has been put forth by several authors. Ziegler (quoted by Dott) stated that in a “few” cases of unilateral hydrocephalus, the foramen of Monro was congenitally maldeveloped, but cited no references in support. Northfield believed that such an entity exists but was unable to cite any supporting cases. Russell stated that she found no convincing evidence of a case of congenital foramen of Monro atresia.

The case that is most suggestive of primary atresia of the foramen of Monro is that presented by Dott. He reported a 9-month-old infant with progressive head enlargement, irritability, left hemiparesis, and right homonymous hemianopsia. Unilateral left hydrocephalus was diagnosed by ventriculography. A transcortical operation was performed for fenestration of the septum pellucidum. Dott observed that the surface in the region of the foramen of Monro was smooth and could only be pinpointed by the convergence of the thalamostriate and septal veins. He also observed that the choroid plexus was quite atrophic. A biopsy from the region of the foramen of Monro yielded organized, fibrous scar tissue. Cerebrospinal fluid (CSF) from the enlarged ventricle contained 18 lymphocytes, and a protein clot formed on standing. The atrophic choroid plexus scar tissue in the region of the foramen of Monro and the CSF findings, however, would seem to point to post-inflammatory obstruction rather than congenital maldevelopment.

Although biopsies of the foramen of Monro were not taken in the case reported in this paper, the following evidence supports the diagnosis of congenital atresia of the foramen: 1) the smooth glistening ependyma-like surface covering the indentation at the site of the foramen; 2) the normal appearance of the choroid plexus; 3) the lack of inflammatory CSF findings; 4) the negative TORCH titers for inflammatory congenital disorders; 5) the large head circumference at birth consistent with a congenital rather than an acquired problem; and 6) the initial and follow-up CT scans with enhancement which showed no evidence of tumor.

Unilateral hydrocephalus is most likely to present as an expanding mass lesion. Focal neurological deficits occur as the obstructed ventricle compresses adjacent structures. Such deficits are often slowly progressive as CSF accumulates. However, an acute herniation syndrome similar to the one in this case may develop. The diagnosis of unilateral ventricular enlargement is readily made with CT scanning. Contrast-enhanced CT should demonstrate any tumor masses responsible for the obstruction. Metrizamide or air ventriculography may also be useful in this regard.

The procedure of choice for treating unilateral hydrocephalus regardless of the etiology is ventriculoperitoneal shunting. Successful shunting will allow for decompression of the affected ventricle with subsequent reversal of the associated symptoms. In those cases secondary to benign tumor masses, excision of the lesion may relieve this condition. Radiotherapy may be similarly effective for malignant lesions.

In the case of inflammatory obstruction of the foramen, many authors have advocated fenestration of the septum pellucidum. This generally involves a transcortical or transcallosal approach with an increased risk of morbidity. However, endoscopic fenestration of the
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septum has been accomplished. Fukushima, et al., described the use of a flexible ventriculofiberscope* for this purpose. The septum was perforated in six cases, two associated with tumor and four with inflammatory obstruction at the foramen of Monro. However, in all six cases, ventriculoatrial shunting was performed at the same time. The results of the endoscopic fenestrations were not described. The use of the rigid ventriculoscope3 for fenestration of the septum is not recommended. This would require blind manipulation in the region of the superior third ventricle, and the ability to create a reliable interventricular communication is questionable.

References

12. Von Mohr: Caspers Wochenschr 121:2-15, 1842

* Ventriculofiberscope manufactured by Olympus Optical Co., Tokyo, Japan.