Congenital unilateral hydrocephalus

Saburo Nakamura, Hideyasu Makiyama, Atsushi Miyagi, Takashi Tsubokawa, and Hiroya Ushinohama

Department of Neurological Surgery and Department of Pediatrics, Nihon University School of Medicine, 30-1 Oyaguchi Kamimachi, Itabashi-ku, Tokyo, 173 Japan

Abstract. An 848-g infant delivered after a 30-week gestation had been diagnosed by ultrasonic examination as having a unilateral ventriculomegaly from the 29th week of gestation. Computerized tomography (CT) demonstrated a greatly dilated right lateral ventricle with no apparent space-occupying lesion within the ventricle and no passage of the contrast medium (metrizamide) through the foramen of Monro. The child was diagnosed as having unilateral hydrocephalus due to congenital obstruction of the foramen and underwent a ventriculoperitoneal shunt operation on day 49 (37 weeks and 5 days of amended gestation age; 1,420 g body weight). The postoperative course was uneventful and the patient was discharged on day 126 with no marked developmental retardation. Angiography carried out at 8 months postpartum revealed displacement and hypoplasia of the deep cerebral veins. The pathogenesis and treatment of unilateral hydrocephalus are discussed.

The term unilateral hydrocephalus was defined by Dott [4] in such a way that it should be restricted to cases in which one lateral ventricle or part of it becomes dilated as a result of an obstruction affecting its cavity or outlet. The present paper describes a case of such unilateral hydrocephalus and discusses the possible mechanisms underlying the pathogenesis in this case.

Case report

An 848-g infant was delivered by Caesarean section after a 30-week gestation. The child has been diagnosed by ultrasonic examination as having a unilateral ventriculomegaly from the 29th week of gestation (Fig. 1). The Apgar score was 5 at 1 min. The infant had severe respiratory distress that required mechanical ventilation. The head circumference at birth was 25.5 cm and the anterior fontanelle appeared flat.

Neurological examination revealed no apparent deficits, and clinical examination of the blood and urine disclosed no abnormality. A computerized tomographic (CT) scan (Fig. 2A, B) demonstrated a greatly dilated, whole right lateral ventricle and a dislocated midline structure toward the contralateral side. However, no apparent space-occupying lesion within the ventricle was recognized. The III ventricle was slightly dislocated to the left side but contained no mass lesion.

Over the following 29 days, the infant's head circumference increased by 2 cm. The intraventricular pressure was estimated to be 160 mmHg on day 36. A CT scan (Fig. 3A) carried out after the intraventricular injection of metrizamide revealed neither passage of the contrast medium through the foramen of Monro to the III ventricle nor any mass lesion at the septum pellucidum. Subsequent CT scans (Fig. 3B) showed no passage of the contrast medium through the foramen but migration into the paraventricular cerebral parenchyma. Based on the above findings, the child was diagnosed as having unilateral hydrocephalus due to congenital obstruction of the foramen of Monro and underwent a ventriculoperitoneal shunt operation on day 49 (37 weeks and 5 days of amended gestation age; 1,420 g body weight).

The postoperative course was uneventful and the infant's development proceeded well. A CT scan (Fig. 4A, B) carried out on the 21st day after surgery showed remarkable diminution of the right lateral ventricle.
Fig. 2 A, B. CT scans on day 6, showing the greatly dilated right lateral ventricle and dislocated midline structure toward the contralateral side.

Fig. 3 A, B. Metrizamide CT scans. A CT scan at 10 min after injection, showing neither passage of the contrast medium through the interventricular foramen (Monro) to the III ventricle nor any mass lesion at the septum pellucidum. B CT scan at 24 h after injection, showing no passage through the foramen but migration into the paraventricular cerebral parenchyma.

Fig. 4 A, B. CT scans on the 21st day after ventriculoperitoneal shunt surgery, showing a marked diminution of the ventricular size. Obstruction of the right foramen of Monro is indicated by the arrowhead in A.

Fig. 5. Venogram at 8 months postpartum, showing backward and upward displacement of the venous angle (arrowhead) and hypoplasia of the septal, thalamostriate, and internal cerebral veins and great vein of Galen (arrow).

Fig. 6. CT scan at 9 months postpartum, showing the collapse of both lateral ventricles and no space-occupying lesion at the septum pellucidum or in the III ventricle.

lateral ventricular size and obstruction of the foramen of Monro, with no neoplastic growth (Fig. 4A). Neurological examination failed to reveal marked developmental retardation, and the patient was discharged on day 126.

Angiography (Fig. 5) carried out at 8 months postpartum revealed backward and upward displacement of the venous angle and hypoplasia of the thalamostriate and septal veins. The vein of Rosenthal was also hypoplastic. No abnormal vessels or blushing were identified at the venous angle. CT scans (Fig. 6) indicated the collapse of both lateral ventricles and revealed no space-occupying lesion at the septum pellucidum. At 1 year after surgery the patient appears quite normal without any neurological signs or symptoms.