Arnold-Chiari Malformation with Cyst of Third Ventricle

A Case Report*

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The mechanism of hydrocephalus with the Arnold-Chiari malformation has been explained by Russell and Donald as impaction of the outlets of the fourth ventricle below the foramen magnum, with prevention of reflux of cerebrospinal fluid over the cerebral hemispheres. Relative failure of mechanisms for absorption of cerebrospinal fluid may be an additional factor. Other specific causes of hydrocephalus, such as aqueductal stenosis or closure and atresia of the foramina of Magendie and Luschka have been reported in association with the Arnold-Chiari malformation.

Gardner et al. reported an "arachnoid" cyst of the cerebellum causing hydrocephalus by compression of the fourth ventricle. The patient also had basilar impression and other anomalies of the craniocervical junction. The authors believed the cyst was congenital and a manifestation of defective permeability of the roof of the embryonal fourth ventricle. Gardner proposed this same developmental defect as the underlying factor in the pathogenesis of Arnold-Chiari malformation, hydrocephalus, myelomeningocele and other associated anomalies.

A subtentorial, supracollicular cyst causing obstructive hydrocephalus was reported by Alexander, who differentiated this case of a noncommunicating cyst from others resulting from diverticula of ventricles dilated by long-standing hydrocephalus. The cyst could not be filled with air at ventriculography, and the concentration of protein in the fluid in the cyst was greater than in the ventricular fluid.

In this report we describe the unique occurrence of a posterior third ventricular cyst causing hydrocephalus in a patient having the Arnold-Chiari malformation and myelomeningocele. The cyst did not communicate with the lateral ventricles; it blocked both foramina of Monro and was in direct communication with the fourth ventricle.

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Case Report

UVII §517914,* A 2-month-old girl was referred to the University of Virginia Hospital because of a lumbosacral myelomeningocele. At birth she was noted to have partial paralysis and hypesthesia of the lower extremities, urinary dribbling and absence of the anal sphincter reflex. Circumferences of the head and chest were 34 and 35 cm, respectively, and the fontanelles were soft. The myelomeningocele ruptured on several occasions prior to her referral. In time it sealed spontaneously, and then more rapid enlargement of the head was noted. The baby had recurrent episodes of bronchopneumonia, probably resulting from her inability to swallow and frequent regurgitation and aspiration. She received tube feedings and intravenous fluids. Antibiotics were given for treatment of bronchopneumonia—Chloramphenicol on one occasion and Declomycin on another.

Examination. The child appeared extremely ill, and weighed 7 lbs., 10 oz., only 1 lb. more than her weight at birth. Temperature was normal despite severe respiratory distress characterized by inspiratory stridor and intercostal retraction. The cranial circumference was 39 cm.; the fontanelles were bulging and the sutures were wide. The veins of the scalp were engorged. Coarse rhonchi were audible bilaterally. No cardiac murmurs were heard. A S by 3 cm. soft, well-covered myelomeningocele was present in the lumbosacral region. Spontaneous movements of the legs were limited to flexion of the thighs. Hypesthesia of the lower extremities was noted. Tone of the anal sphincter was poor and the child dribbled feces and urine. Diagnoses included myelomeningocele, hydrocephalus, probable Arnold-Chiari malformation with paralyses of the lower cranial nerves, bronchopneumonia and malnutrition.

Course. Initial efforts were directed at treatment of the respiratory infection and malnutrition. A ventriculogram showed failure of communication between the right and left lateral ventricles; both were severely dilated and did not communicate with the third ventricle. The fourth ventricle could not be visualized. Fig. 1 illustrates these findings which were interpreted as consistent with a mass in the third ventricle. Daily ventricular aspirations through the coronal sutures were performed to relieve increased intracranial pressure. Pre-operative cultures of ventricular fluid did not yield organisms.

Operation. Exploration through a right frontal craniotomy revealed a cyst of the third ventricle occluding both foramina of Monro. The cyst was incised on
Fig. 1. Pre-operative ventriculogram (anteroposterior, brow-up view) shows greatly dilated lateral ventricles. The 3rd ventricle contains a mass, later recognized as a cyst, and did not fill from either lateral ventricle.

The child deteriorated during the next few weeks and died at the age of 5 months.

Macroscopic Findings. At necropsy the brain, spinal cord and myelomeningocele were removed en bloc posteriorly. Elongation of the medulla oblongata into the cervical spinal canal was seen, and the lower 4 cranial nerves coursed superiority from their origins. The upper cervical nerves also were directed rostrally. Fig. 3 shows the compressed cerebellum with a small tongue extending below the level of the foramen magnum. The impression of a 2×3 cm. mass is also shown extending posteriorly from the roof of the fourth ventricle, and superiority between the cerebellar hemispheres. This mass was a subtentorial diverticulum of a cyst in the posterior part of the third ventricle (2×3 cm.) The fourth ventricle was sealed by arachnoidal fibrosis but communicated freely with the third ventricular cyst and its subtentorial diverticulum through a patent aqueduct of Sylvius. The cavity thus outlined was walled off from the remaining ventricular system and subarachnoid space, creating a separate mechanism of obstructive hydrocephalus. Cystic fluid was supplied from the choroid plexi of part of the third and fourth ventricles. This cystic fluid was clear and xanthochromic. The relationship of the third ventricular part of the

Fig. 2. A later ventriculogram (anteroposterior, brow-up view) demonstrates enormously dilated lateral ventricles and the mass in the 3rd ventricle. A cyst communicates with the lateral ventricle, formed in part by a septum extending from the midline laterally and inferiorly.

...the right side and a communication made between the lateral ventricles through the septum pellucidum. An outflow tract was established from the lateral and third ventricles to the subarachnoid space by way of the operative approach, an anterior lateral ventriculostomy. The aqueduct of Sylvius and the outlet of the fourth ventricle were not investigated at this time.

Postoperatively, the child had decreased respiratory distress and diminished inspiratory stridor. The fontanelles remained soft for approximately 2 weeks, but then the child gradually regressed to her pre-operative status, presumably because of closure of the lateral ventriculostomy and recurrent hydrocephalus.

A second ventriculogram showed enormously dilated lateral ventricles communicating with each other, but again the third and fourth ventricles did not fill. Fig. 2 illustrates these findings as well as septa within the ventricular system.

2nd Operation. The posterior fossa was explored, and the presence of an Arnold-Chiari malformation was established. Adhesive arachnoidal fibrosis was also encountered over the fourth ventricle. Indigo carmine was injected into the lateral ventricle, but failed to appear in the fourth ventricle. A Torkildsen procedure was performed, but post-operatively the child failed to improve. Paralysis of the vocal cords was confirmed by laryngoscopy, and a tracheostomy was done. A revision of the ventriculocisternostomy was attempted; however, relief of hydrocephalus was not accomplished.