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.

Case Report

UVII 517914. A 2-month-old girl was referred to the University of Virginia Hospital because of a lumbo-sacral 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 Decloymycin 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 retractions. The cranial circumference was 39 cm.; the fontanelles were bulging and the sutures were wide. The veins of the scalp were engorged. Coarse ronchi were audible bilaterally. No cardiac murmurs were heard. A 8 by 3 cm. soft, well-covered myelomeningocele was present in the lumbo-sacral 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 paralytic ileus 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

* Patient was referred by Dr. William E. Harman of Staunton, Virginia.
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 superiorly 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 superiorly between the cerebellar hemispheres. This mass was a subtentorial diverticulum of a cyst in the posterior part of the third ventricle. 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

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**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.

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**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.
Arnold-Chiari Malformation with Cyst

FIG. 8. Sagittal section through the pons, medulla, cerebellum and cervical part of the spinal cord, showing elongation of the medulla, downward displacement of a small tongue of cerebellar tissue, and the impression of a midline mass in the superior portion of the cerebellum. Arachnoidal fibrosis is evident around the lower end of the fourth ventricle.

cyst to the diverticulum, aqueduct of Sylvius, and fourth ventricle may be seen diagrammatically in Fig. 4.

The cerebral cortex was thin, especially anteriorly, over greatly dilated lateral ventricles. The gyral pattern was almost completely lost in the frontal poles. The ventricular lining was smooth. Horizontal sections through the right hemisphere (Fig. 5) revealed multiple cysts in the centrum semiovale. The left hemisphere, sectioned in the coronal plane, also showed multiloculation. The grey and white matter surrounding the posterior horns of the lateral ventricles was normal.

Microscopic Findings. The cysts in the ventricles and the cerebral hemispheres were cavities enclosed by mildly reactive glial tissue (Fig. 6) and were not lined with ependyma. Many protoplasmic astrocytes were seen in the surrounding tissue. An angiomatous malformation, containing some vessels with media muscularis, as well as large endothelial lined channels (Fig. 7), was found in the wall of the cyst of the posterior part of the third ventricle. The thin cerebral cortex contained only a few degenerated neurons and many plump astrocytes. The ependymal lining of the lateral ventricles was disrupted. Arachnoidal villi were normal. Hemosiderin-filled macrophages and focal fibrosis were identified in zones of surgical manipulation.

Foci of neuronal loss were seen in deep layers of the cerebellum. In addition, clumps of ependymal cells were identified in anomalous locations in the cerebellum. The leptomeninges of the entire posterior fossa were thick and contained fibrous and collagenous connective tissue.

FIG. 4. Median sagittal diagram of the third ventricle cyst (cross hatched) and its connections.

FIG. 5. Irregularly shaped cysts (arrows) can be seen in these horizontal sections through the right cerebral hemisphere.

FIG. 6. Photomicrograph of a portion of cerebral tissue between lateral ventricle (left) and a cyst (right). The wall of the cyst has undergone mild gliosis. Hematoxylin and eosin stain; X55.
channels are seen. Developed vessels as well as large endothelial-lined channels are seen. Hematoxylin and eosin stain; X55.

Discussion

The unique findings in this patient are interesting from several standpoints. A number of causes for the hydrocephalus were present. The cyst of the posterior part of the third ventricle not only obstructed both foramina of Monro, but also blocked the pathway from the anterior part of the third ventricle. Outflow from the cyst was impaired as evidenced by the presence of the subtentorial diverticulum and the convex appearance of the cyst in the ventriculogram. The aqueduct of Sylvius was patent; hence, either arachnoidal fibrosis of the foramina of Magendie and Luschka, or impaction of these openings below the foramen magnum could have prevented outflow from the cyst. The subtentorial diverticulum compressed the cerebellum and fourth ventricle, compounding the obstruction. Intracranial hypertension was relieved after the first surgical procedure, affording evidence that a defect in absorption of cerebrospinal fluid was not operative. Treatment of hydrocephalus in this patient necessitated establishing communications between the cyst of the third ventricle, the lateral ventricles and the anterior part of the third ventricle, before accomplishing any of the usual ventricular decompressing procedures.

The origin of the cysts in the ventricle and cerebral substance is uncertain; however, the presence of gliosis indicates a reactive process. Although the ventricular fluid was sterile pre-operatively, contamination might have occurred at the times of rupture of the myelomeningocele. Infections then could have been masked by antibiotic therapy. The smooth appearance of the ventricular lining precludes the possibility of massive ependymitis. Some reactive gliosis resulting from surgical intervention was undoubtedly superimposed.

The occurrence of the angiomatous malformation in the wall of the cyst of the posterior part of the third ventricle strongly implicated anomalous development as a cause of that cyst. Considering the cyst of the third ventricle as a developmental anomaly supports the concept that the Arnold-Chiari malformation is one of many manifestations of dysplasia of the central nervous system, and not caused by an underlying defect at one or the other end of the neural tube. This case is similar to a case of aqueductal atresia occurring with a lumbar meningocele \(^3\) in that both provide the same objection to the hydromyelic theory postulated by Gardner. \(^2\) With obstruction of outflow from the lateral and third ventricles, it is difficult to explain the pathogenesis of the myelomeningocele on the basis of transmission of pressure or pulse waves from the choroid plexi to the central canal of the spinal cord.

Summary

This is so far as we can determine the first reported case of an obtrusive posterior third ventricular cyst occurring in a patient having hydrocephalus, Arnold-Chiari malformation and myelomeningocele. The presence of an angiomatous malformation in the wall of the cyst suggests that both were of developmental origin. Consideration of the cyst as an anomaly of development strengthens the concept that the Arnold-Chiari malformation is only one manifestation of a range of dysplasias of the central nervous system, and not the result of a defect at one or the other end of the neural tube. In addition, the presence of the cyst offers evidence against the hydromyelic theory as an explanation of the cause of myelomeningocele. The factors contributing to the development and progression of hydrocephalus in this patient are discussed.

References

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