Communication of hydromyelic cavity with fourth ventricle shown by combined Pantopaque and air myelography

Case report

HECTOR E. JAMES, M.D., LUIS SCHUT, M.D., AND PATRICK P. PASQUARIELLO, M.D.

Division of Neurosurgery and Department of Medicine of the Children's Hospital of Philadelphia and the Hospital of the University of Pennsylvania, Philadelphia, Pennsylvania

✓ A 3-year-old child who had had respiratory and feeding difficulties at birth was admitted because of respiratory distress and stridor. Radiological evaluation with Pantopaque and air myelography revealed an Arnold-Chiari malformation associated with extensive hydromyelia and communication between the fourth ventricle and the hydromyelic cavity.

Key Words · Arnold-Chiari syndrome · hydromyelia · fourth ventricle · myelography

The Arnold-Chiari malformation and hydromyelia have received considerable attention in recent years because of Gardner's hypothesis regarding the etiology of hydromyelia and syringomyelia. This case supports the theory that impairment of normal cerebrospinal fluid (CSF) drainage from the fourth ventricle results in dilatation of the central canal of the spinal cord.

Case Report

This 3-year-old girl was admitted to The Children's Hospital of Philadelphia because of respiratory distress and stridor. There had been respiratory embarrassment at birth and subsequent difficulty with feedings. These problems resolved, but she had a persistent stridor until shortly before her admission when she suffered two apneic episodes. She presented with acute respiratory distress for which a tracheostomy was performed. Direct laryngoscopy revealed bilateral vocal cord paralysis.

Examination. The general physical and sensory examinations were normal. There was a diminished gag reflex on the right side, bilateral fasciculations of the tongue, and questionable weakness of the right extremities. Plain cervical spine films revealed a widened spinal canal in both the anteroposterior and lateral views with a "funnel-shape," the base directed cephalad. Skull films were initially reported as normal. A retrospective evaluation revealed findings suggestive of increased intracranial pressure. Pantopaque myelography performed via the lumbar subarachnoid route showed marked cervical and thoracic enlargement of the spinal cord. The contrast material was maneuvered into the fourth ventricle under fluoroscopy, passing by a clearly outlined channel (Fig. 1 left) into the hydromyelia cavity.
which extended from C-2 through T-12 (Fig. 1 right). Air myelography and pneumoencephalography were then performed with a total exchange of CSF; the study was performed with the patient first sitting and subsequently lying down. The fourth ventricle was well filled with air which also entered the hydromyelia cavity forming with the Pantopaque a double contrast outline; segmentation of the cavity was evident (Fig. 2).

Operation. A suboccipital craniectomy and a cervical laminectomy were performed. Downward displacement of the cerebellar tonsils was evident with upward direction of the cervical roots (Arnold-Chiari malformation Type 1). The cisterna magna was obliterated by adhesions, and the cervical spinal cord markedly distended.

Postoperative decompensated hydrocephalus developed and a ventriculoperitoneal shunt was inserted. Following this procedure the patient improved markedly, with strengthening of her voice and, several weeks later, diminished fasciculations of the tongue. Except for an episode of decompensated hydrocephalus due to malfunction of the shunt, she has done well now for 8 months postoperatively.

Discussion

The age of the patient at the onset of symptoms is unusual. Both in the Gardner and Goodall\textsuperscript{8} series of 17 patients and the Appelby,\textit{et al.}\textsuperscript{2} series of 11 patients, the youngest patients were aged 14 and 17 years respectively. Other published series do not reveal any clinical cases below the age of 10 years.\textsuperscript{4,10,12,13,14}

The evaluation of anomalies of the posterior fossa-cervical spine junction with positive contrast myelography has been well described by several authors.\textsuperscript{1-3,12} The use of
Communication between hydromyelic cavity and the 4th ventricle

Fig. 2. Under fluoroscopy and with tomography, air outlined the fourth ventricle, and a double contrast image inside the hydromyelia was then seen.

Air myelography for the same purpose was described in detail by Wickborn and Hanafee. Opaque myelography followed by pneumoencephalography to evaluate the size of the ventricles was recommended by Heinz, et al., for cases in which hydromyelia or syringomyelia was suspected.

Most authors have had difficulty in demonstrating the hydromyelia cavity by direct filling. Tjaden, et al., recommended positive contrast ventriculography to demonstrate the hydromyelia cavity in suspected patients. Filling of the cavity was reported previously by Debrung, et al., during an incidental ventriculogram. In reviewing their cases, Heinz, et al., recommended myelography prior to other studies, followed by pneumoencephalography when junctional anomalies were encountered. Assessment of ventricular size is important because hydrocephalus is commonly associated with Arnold-Chiari malformations. We like this approach because a more thorough evaluation of the occipital-cervical junction can be performed by myelography than with ventriculography.

The particular radiological features of this case are the clearly visualized communication between the fourth ventricle and the hydromyelia cavity (Fig. 1 left) not previously visualized to our knowledge, and the segmentation of the Pantopaque inside the hydromyelia cavity (Fig. 1 right). The latter had been described by Heinz, et al., though no explanation has been put forward as to its etiology. The radiological and surgical findings confirm Gardner's theory of hydromyelia formation. Recently Williams has modified the hypothesis, but the basic anatomical configuration is the same.

The therapy of the disease has recently been discussed and modified by Benini and Krayenbühl. They recommended a ventriculojugular shunt rather than decompression.
laminectomy, thereby relieving the caudally-directed pressure effect into the cervical spinal column. Our case subsequently required a shunt although an adequate decompression had been performed, and thus supports their theory.

References

Address reprint requests to: Luis Schut, M.D., Chief, Neurological Service, The Children's Hospital of Philadelphia, 1740 Bainbridge Street, Philadelphia, Pennsylvania 19146.