Fourth ventriculoceles with extracranial extension

ROBERT A. FENSTERMAKER, M.D., UROS ROESSMANN, M.D., AND HAROLD L. REKATE, M.D.

Division of Neurosurgery, University Hospitals of Cleveland, and Division of Neuropathology, Institute of Pathology, Case Western Reserve University, Cleveland, Ohio

The radiographic features and long-term clinical outcome in three patients who presented at birth with a cystic suboccipital mass in direct communication with the fourth ventricle are reviewed. The pathological findings in a fourth infant who died are also discussed. All surviving infants were treated with cyst excision and diversion of cerebrospinal fluid. The prognosis in these children, followed from 6 to 20 years, surpasses that of the more common occipital encephalocele, for which this entity could be mistaken. The morphogenetic implications relative to more common congenital lesions in this location are discussed.

KEY WORDS: encephalocele, rhombic-roof ventriculocele, cerebellar vermis, Dandy-Walker malformation, fourth ventricle

THE prognosis for children with occipital encephaloceles is generally poor. The incidence of intellectual and physical disability, as well as death, is higher than 75%. Some have suggested lower figures, and have confirmed the role that hydrocephalus and brain within the sac contribute to the overall poor prognosis.10,11

This article describes a group of four patients, three of whom were brought to University Hospitals of Cleveland between 1963 and 1977. All were seen for low-occipital mass lesions initially believed to represent simple occipital encephaloceles. Subsequent radiological evaluation and surgery revealed a communication of the cystic mass with the fourth ventricle. The autopsy findings in a fourth child who died as a result of other anomalies are presented.

Case Reports

Case 1
This child was a 3560-gm product of a full-term uncomplicated pregnancy. At birth, he was noted to have a 4-cm mass located directly below the inion. Ventriculography demonstrated partial agenesis of the corpus callosum, hydrocephalus, and a rhombic-roof ventriculocele extending extracranially. The child was treated with cyst excision and ventricular shunting. He is now 20 years old, having graduated from high school with average grades. His full-scale intelligence quotient (IQ) is 111 with above-average verbal abilities but slow visual-motor skills.

Case 2
This 2480-gm baby girl was the product of a 37-week uncomplicated pregnancy. Following birth, the child was noted to have a 20-cm low-occipital mass covered by grossly normal integument. Ventriculography and pneumoencephalography revealed hydrocephalus with a high-riding tentorium as well as an absent fourth ventricular roof. She was treated by excision of the mass and cystoperitoneal shunting. The child is currently 10 years old, without neurological impairment, and has a full-scale IQ of 120.

Case 3
This 2980-gm male neonate was delivered at 33 weeks of gestation by Caesarean section. Pregnancy had been complicated by hydramnios and, at birth, a prolapsed cord was present. The Apgar scores were 5 at 1 minute and 5 at 5 minutes. A huge occipital mass was apparent (Fig. 1) and subsequent ventriculography showed elevation of the tentorium and communication of the fourth ventricle with the cystic mass (Fig. 2). Vertebral angiography revealed normal posterior cerebral arteries with no involvement of the occipital lobes. The suboccipital mass was supplied by a greatly dilated right anterior inferior cerebellar artery (Fig. 3). The child was treated with cyst excision and cystoperitoneal shunting. Currently, he is 6 years old and well, apart from ataxic gait and sensorineural hearing loss resulting in delayed language development.

Case 4
This 1200-gm boy was delivered at 32 weeks by Caesarean section for breech presentation. Apgar scores were 3 at 1 minute and 3 at 5 minutes. At birth, a leaking suboccipital mass was noted. The infant remained cyanotic despite respiratory support and died 2 hours after birth. At autopsy, polycystic kidneys, se-
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verely hypoplastic lungs, and biliary dysplasia were evident. The cerebellar vermis was hypoplastic and an enormously dilated fourth ventricle protruded through a cranial defect forming an 8-cm suboccipital mass. The cerebellar hemispheres were asymmetrical and the highly vascularized cyst was lined with a glial-ependymal membrane. The cerebrum, brain stem, and spinal cord were normal. Microscopic examination of the cerebellum revealed considerable dysgenesis with immature neuroepithelial cells surrounded by differentiating neurons.

Summary of Cases

All surviving children underwent ventriculography. Each study showed an elevation of the tentorium as in the Dandy-Walker malformation, with absence of the fourth ventricular roof. This latter fact was established in one child (Case 2) by direct puncture of the cyst wall just prior to surgery. The ventriculogram of one patient disclosed partial agenesis of the corpus callosum, a frequent concomitant finding in the Dandy-Walker malformation. One infant underwent vertebral angiography demonstrating lack of involvement of the cerebral hemispheres. Each patient then underwent cyst excision and posterior fossa exploration, followed by either cystoperitoneal or ventricular shunting.

As described above, the single non-survivor in our group of patients, in addition to his central nervous system malformation, suffered from Potter's syndrome, a uniformly fatal condition in itself. This child was not studied radiographically.

Discussion

Pathological descriptions of rhombic-roof ventriculoceles have appeared in the literature, both in association with and in the absence of an external defect. The most common of these is the Dandy-Walker syndrome in which vermal hypoplasia is believed by some to follow compression by a rhombic-roof ventriculocele.

Two recent examples of pathological analyses of fourth ventriculoceles with extracranial components are provided by Evrard and Caviness. They described an autopsy specimen in which a congenital suboccipital mass containing a cystic enlargement of the fourth ventricle was associated with hydrocephalus, occlusion of the fourth ventricular outflow apertures, and a midline diencephalic fusion. A later case reported by them describes the postmortem examination of an infant with a suboccipital mass containing both a rhombic-roof ventriculocele and a pulsion diverticulum of the occipital lobe herniating into the sac by way of a widened tentorial incisura.

Other cerebellar anomalies may involve primarily the hemispheres and, to a greater or lesser degree, the vermis as well. For example, the Arnold-Chiari Type III anomaly presents with a suboccipital or "cerebellar" encephalocele. In contrast to the Dandy-Walker malformation and to the cases described here, the Arnold-Chiari anomaly has a characteristically shallow pos-

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terior fossa and a low torcular. Some have cited the rare examples of suboccipital encephaloceles as a link between the Dandy-Walker malformation and the dysraphic states. More clinicopathological study of these less common conditions will be necessary to better define the relationship between these entities, if there is one.4,6,8,13,17

The children in this series all presented prior to the availability of computerized tomography (CT). Kojima, et al.,7 have reported two patients with fourth ventriculoceles and extracranial extension who were treated successfully, and have demonstrated the CT appearance of this condition. It seems reasonable to expect that, with the help of CT, diagnosis of fourth ventriculocoeles will become more straightforward.

There have been only occasional descriptions of children surviving with this condition and the issue of long-term outcome has, therefore, not been addressed. Frequently, it cannot be ascertained from these reports whether the suboccipital masses described were in communication with the fourth ventricle. Some may actually represent occipital encephaloceles or Dandy-Walker variants.5,9,14-16,19

Diverse theories have been advanced regarding the morphogenesis of these malformations. In the Dandy-Walker syndrome, atresia of the foramina of Luschka and Magendie has been implicated as the causative factor in formation of the fourth ventriculocoele. The finding of patency of these structures in some specimens, however, has been the chief objection to this theory. Friede3 stated that the most tenable theory concerning the morphogenesis of the anomaly is based on an initial arrest in development of the hindbrain, followed by "peristence of the anterior membranous area, ... non-opening of the cerebellar foramina and hindrance of migration of neuroblasts to the inferior olivary nuclei."

Normally, the straight sinus is near the vertex of the cranium early in development, and gradually migrates to its final low-occipital location.18 The persistence of a high-riding tentorium in the Dandy-Walker syndrome may reflect either an additional developmental arrest or a hindrance to migration by the expanding ventriculocoele itself. This and other factors help to place the initial event within the first 3 months of fetal development.

In patients with fourth ventriculoceles and extracranial extension, it is uncertain why there is an additional failure of the occipital bone with herniation of the ventriculocoele through the cranial defect. Such bone defects are known to occur with and without herniation of intracranial tissues.12 The absence of this component in all cases of Dandy-Walker is cause for speculation that the fourth ventriculocoele with extracranial extension may arise prior to achievement of integrity of the mesenchymal elements forming the occipital squama.5

The cases of fourth ventriculocoele presented here and by others add further to the spectrum of anomalies seen. When unassociated with other serious abnormal-

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Address reprint requests to: Harold L. Rekate, M.D., Division of Neurosurgery, University Hospitals of Cleveland, 2074 Abington Road, Cleveland, Ohio 44106.