Giant cyst of the septum pellucidum

Case report

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Symptomatic cysts of the septum pellucidum are rare. Up to 1969, only 17 published cases were considered to be symptomatic. In the case reported here, a 9-week-old girl was hospitalized because of disproportionate progressive enlargement of the head, lumbar myelomeningocele, and a history of generalized seizures. A giant septum pellucidum cyst was demonstrated on Conray ventriculography and later on computerized tomography (CT) scanning. Aqueductal stenosis was suspected. The cyst was excised at craniotomy. A ventriculoperitoneal shunt was required to treat the accompanying hydrocephalus.

KEY WORDS • septum pellucidum • “fifth ventricle” • cavum septi pellucidi

PATHOLOGICAL cysts of the cavum septi pellucidi have rarely been described in the literature. Up to 1969, only 17 of the published cases were considered to be symptomatic. A few others have been reported since. An additional case is reported here.

Case Report

This 9-week-old girl was admitted to our neurosurgery department, on February 4, 1980, with a history of generalized seizures, disproportionate enlargement of the head, and lumbar myelomeningocele. She was born on November 29, 1979, of a full-term first pregnancy. Delivery was normal. Respiration and crying were both immediate and spontaneous. Lumbar myelomeningocele and paraparesis were noticed at birth. During the 2nd month of her pregnancy, the mother had accidentally fallen from a second story level and required bed rest for 2 months to avoid a threatened abortion. During the 4th month of pregnancy, the mother suffered from influenza for 10 days. There was no hereditary history of congenital defects. The patient had a generalized seizure when 15 days old, and another about a week later. It was soon noticed that her head was enlarging abnormally.

Examination. The child was conscious on admission. Head circumference was 40.5 cm, and the anterior fontanel was full and prominent. Discrete collateral venous circulation was noticed on both sides of the forehead. The pupils and fundi were normal. A marked, predominantly distal paraparesis was evident; only weak thigh movements could be elicited. Both feet were held in a persistent equinovalgus attitude. Sensation was markedly impaired below L-3 bilaterally. Sucking and Moro reflexes were positive. Deep tendon reflexes were increased in the upper extremities but were absent in the lower limbs, where the muscle tone was also diminished. The anal reflex was weak. A 3.5 x 3.0 cm lumbar myelomeningocele was observed. It had an adequate cutaneous cover and could be transilluminated. Cranial suture separation and spina bifida of L-5 and S-1 were seen on plain x-ray films. Hemoglobin was 8.2 gm.

On February 13, 1980, after correction of the baby’s anemia, a percutaneous Conray ventriculogram was attempted under local anesthesia, and clear, colorless fluid with normal chemistry was obtained. Conray, 7 ml, was instilled, and a giant cyst of the septum pellucidum, measuring 5 x 4 x 6 cm, was demonstrated on x-ray films (Fig. 1). The cyst extended both sides of the midline in the frontal region. Communi-
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Fig. 1. Skull x-ray films, Towne view (left) and lateral view (right). A giant midline cyst of the septum pellucidum is seen, filled with Conray. No communication with the ventricular system is observed.

cation with the ventricular system was not accomplished with manipulation of the head, and the cyst remained unchanged on a film 24 hours later. Following percutaneous aspiration of the cyst 2 days later, the fontanel became sunken and the head measurement decreased to 38 cm. A CT scan on February 19 showed a midline frontal cyst filled with residual Conray (Fig. 2). The cyst obliterated both frontal horns, and the rest of the ventricular system, including the third ventricle, was enlarged and dorsally displaced. One week later, after aspiration of all the contrast material from the cyst, a repeat CT scan showed that the large cyst was isodense, and a distinctive "capsule" was observed (Fig. 3). In both studies, the fourth ventricle was small and in its normal position. Aqueductal stenosis was suggested in addition to the large septum pellucidum cyst. Both foramina of Monro were thought also to be obstructed by the cyst.

Operation. On March 4, 1980, the cyst was approached transcortically through a right frontotemporal craniotomy under general endotracheal anesthesia. A small amount of blue dye was injected into

Fig. 2. Computerized tomography showing a well demarcated anterior midline cyst filled with Conray. Both lateral ventricles are enlarged and displaced backward.
FIG. 3. Computerized tomography 1 week later, after aspiration of the cyst. The Conray has been removed. The isodense septum pellucidum cyst is clearly demonstrated, delimited by a thick membrane.

the cyst. The cavity collapsed when its translucent, shiny, thin wall was entered, and the contents escaped. Loose adherence of the membrane to the surrounding structures allowed almost complete excision of the cyst. Segmental thickening of the wall was noticed, particularly over the foramina of Monro, neither of which could be safely opened, so the third ventricle was entered from its anterior wall. Wide communication with both lateral ventricles was accomplished. Clear cerebrospinal fluid (CSF) was obtained from the lateral ventricles, whereas the cyst fluid had been blue-tinged. The choroid plexus of the right lateral ventricle was never visualized, but the plexus in the left lateral ventricle and in the third ventricle were clearly identified.

Postoperative Course. The postoperative course was uneventful. Microscopic examination of the cyst wall revealed “abundant gemistocytic glial cells in its inner surface.” The myelomeningocele was repaired on March 14, 1980, and the patient was discharged from the hospital 4 weeks later. Recurrent intracranial hypertension prompted her readmission on June 26, 1980. Further enlargement of the ventricular system was demonstrated on CT scanning. A ventriculoperitoneal shunt was inserted on July 8, 1980, and the patient was discharged 8 days later. The shunt has been revised three times since then. The child’s psychomotor development has been slow.

Discussion

Cavum septi pellucidi was described by Verga in 1851, and by Ferrario in 1855, but these abnormalities were known to Sylvius. Meyer, in 1930, and Pendergrass and Hodes, in 1935, described finding them on pneumoencephalography. Their appearance on CT has been discussed by Gonzalez, et al.

A cavity of the septum pellucidum has been reported on autopsy studies in 100% of premature brains. Its incidence in the adult brain has been a controversial matter, with data ranging from 6.5% to 85.0%; other reports have cited figures of 12%, 20%, and 60%. The septum may be solid, web-like in the center, or frankly cavitated (less than 25%). The septum pellucidum bridges the space between the corpus callosum and the fornix. Its cavum communicates in the embryo through the aqueduct septi with another larger caudal cavity: the cavum vergae. Its embryological development correlates with that of the corpus callosum and of the fornix. These latter structures are seen in the human fetus toward the 3rd month of intrauterine life (12th week). The cavum appears at the site of midline fusion of the hemispheric vesicles, where the sulcus medianus telencephalii medii originates. The cavum was formerly considered to be either a remnant of the interhemispheric fissure only interrupted by the corpus callosum, or a central pocket of the lamina terminalis disproportionately stretched by the rapid and expanding growth of the corpus callosum and the fornix. The septal cavities begin to close off, however, in a caudorostral direction at about the 6th month of intrauterine life. Complete obliteration is expected at full-term birth. Patients with persistent cava or cysts, symptomatic or not, of the septum pellucidum have been discussed in the literature. Isolated cysts of the cavum vergae have been reported. As Dandy pointed out, these cavities are improperly called “fifth and sixth ventricles,” for they do not...
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belong to the ventricular system nor have they the same embryological origin. Identical morphological and functional characteristics have been observed in the cellular lining of all these cavities. Thompso proposed to call them “cavum septi anterius” and “cavum septi posterius.” “Cavum” denotes a small incidental asymptomatic space; a larger symptomatic cavity is called a “cyst.” Identical septum pellucidum cysts have been described in two identical twin brothers, and in one of two dizygotic twin sisters. Brain tumors of the septum have been reported, and in two patients a colloid cyst of the cavum septi pellucidi has been surgically excised. Congenital absence of the septum has been observed with agenesis of the corpus callosum or arhinencephaly. Giant cysts of the septum associated with aqueductal stenosis and lumbar myelomeningocele have previously been reported in two patients. These cavities have been classified by Shaw and Alvord as: 1) incidental (asymptomatic), and 2) pathological (symptomatic), the latter being either simple or complicated.

Dandy first excised these lesions at craniotomy 50 years ago, and this procedure has reportedly been the preferred treatment; however, a “limited operation” was suggested in 1934 by Van Wagenen and Aird. Communication with the ventricular system by a ventricular needle being passed through a simple burr hole. In 1949, Miller used this latter procedure under radiographic control. Some cysts have been reported to rupture, establishing communication with the ventricles and amelioration of symptoms during or after pneumoventriculography studies. Expanding growth of the cyst may obliterate the foramina of Monro and the aqueduct, accounting for obstructive hydrocephalus and increased intracranial pressure. The origin of the fluid inside the cysts, however, has not been established. The mechanics of such obstruction have been radiographically documented “de façon formelle” by Aboulker, et al., with lipiodol as contrast material. Conray was used by Kansu and Bertan to fill a large “fifth ventricle” demonstrated earlier on CT in an 8-year-old boy with bitemporal hemianopsia.

Our patient is perhaps the youngest so far reported with a giant symptomatic (pathological) cyst of the septum pellucidum, and one of the few associated with aqueductal stenosis and lumbar myelomeningocele. The patient’s mother had an accidental fall early in pregnancy with a threatened abortion ensuing, and later she contracted an influenza-like illness. These two abdominal situations may have affected the child’s embryological development. At craniotomy, the foramina of Monro were seen to be obstructed by segmental thickening of the cyst membrane, and a choroid plexus of the right lateral ventricle was never visualized.

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