Giant interhemispheric cysts associated with agenesis of the corpus callosum

KOREAKI MORI, M.D.

Department of Neurosurgery, Kochi Medical School, Nankoku City, Japan

Interhemispheric cysts, often associated with agenesis of the corpus callosum, are rare lesions demonstrating little uniformity of pathogenesis. Four large interhemispheric cystic lesions with several unique features are reviewed. Magnetic resonance imaging clearly showed agenesis of the corpus callosum and was useful in the diagnosis of interhemispheric cysts. The differential diagnosis of these lesions is discussed, along with therapeutic considerations.

Key Words • cyst, interhemispheric • corpus callosum • hydrocephalus • cyst, arachnoid • magnetic resonance imaging

Giant interhemispheric cysts are rare lesions which, until the advent of magnetic resonance (MR) imaging, were mainly diagnosed by computerized tomography (CT). Various types of cystic lesions occur in the interhemispheric area. Often they are associated with agenesis of the corpus callosum, but there is no uniformity in pathogenesis among previously reported cases. These cysts should be differentiated from one another, as the prognostic and therapeutic implications may differ.

Four large interhemispheric cystic lesions presenting at four different institutions prompted this report. Magnetic resonance imaging, which is currently the diagnostic tool of choice, clearly demonstrated agenesis of the corpus callosum and was useful in the differential diagnosis of these lesions.

Case Reports

Case 1

This 5-month-old boy was delivered normally at term after an uneventful pregnancy. At birth, he weighed 2950 gm and his head circumference was 34.7 cm. He was subsequently noted to have an enlarged head (97th percentile), which increased progressively thereafter despite otherwise normal development. His head circumference was 41.5 cm at the age of 2 months and 43 cm at 3 months. No history of head injury or high fever was obtained. He was admitted to Osaka City University Hospital in May, 1976.

Examination. The anterior fontanel measured 4.5 × 5.0 cm and the head circumference was 45 cm. Infantile reflexes such as the Moro and grasp reflexes were normal. Plain skull films showed suture separation, and pneumoencephalography revealed air filling of the left lateral ventricle. A structure similar to a cavum veli interpositi was demonstrated. A tap of the left lateral ventricle through the left corner of the anterior fontanel was attempted but the needle entered a cyst instead. The cystic fluid was replaced with air. On pathological examination, the fluid was xanthochromic, with a cell count of 3/µm, a protein content of 390 mg/dl, a sugar content of 92 mg/dl, and a chlorine level of 691 mg/dl. Carotid angiograms revealed lateral displacement of the left anterior cerebral artery.

Operation. A left frontoparietal craniotomy was performed in May, 1976. Immediately after dural reflection, a large cyst measuring approximately 7 × 5 cm was encountered, filled with xanthochromic fluid. The falx was normally developed and the cyst was located between both cerebral hemispheres. Below the falx, the cingulate gyrus of the right cerebral hemisphere could be seen, but the corpus callosum was obscured. Due to its size, the cyst's anatomical relationship to the surrounding structures could not be verified. It was resected as extensively as possible. Pathological examination of its wall resulted in a diagnosis of arachnoid cyst.

Postoperative Course. The postoperative course was complicated by meningitis, which resolved in about 1 month. Postoperative cisternography revealed marked reflux of the contrast medium into the right lateral ventricle and stasis for more than 24 hours. Postoperative pneumoventriculography showed an enlargement of the right lateral ventricle. A ventriculoperitoneal (VP)
Giant cysts with corpus callosum agenesis

![Image](image.png)

**Fig. 1.** Case 2. *Left:* Preoperative enhanced computerized tomography (CT) scan showing a huge frontoparietal midline cyst and enlargement of the lateral ventricles. The cyst wall is not enhanced by contrast medium. *Right:* Postoperative CT scan showing shrinkage of the cyst and a decrease in the size of the lateral ventricles.

shunt was inserted in October, 1976. Postoperative CT scans showed separation of the lateral ventricles with a collapsed interhemispheric cyst between them. The patient has developed relatively well with normal head size but with slight psychomotor retardation.

**Case 2**

This 21-month-old infant was found to have an enlarged ventricle at the 37th week of gestation. He was delivered by suction at 38 weeks of gestation. At birth, he weighed 3060 gm, his head circumference was 33.7 cm, and his Apgar score was 8. He developed normally until 1 year of age, when he was noted to have an enlarged head (97th percentile). Frontal bossing was noted at his 1-year examination and he was referred to the Tottori University Hospital. He was diagnosed as having an interhemispheric cyst associated with hydrocephalus and was referred to the Department of Neurosurgery.

**Examination.** The patient had no neurological abnormality and his developmental milestones were normal. Computerized tomography scans showed a huge midline cystic lesion and enlargement of the lateral ventricles (Fig. 1 *left*). Magnetic resonance imaging revealed a huge interhemispheric cyst associated with agenesis of the corpus callosum and separating the lateral ventricles. Metrizamide CT cisternography revealed no filling of the cyst or the left lateral ventricle. These findings suggested that the cyst had no communication with the subarachnoid space, and that the left foramen of Monro might be obstructed.

**Operation.** Because of progressive enlargement of the head and increase in cyst size, a craniotomy was performed. The cyst was opened and communications were made between it and the subarachnoid space and the lateral ventricles. The cyst and its wall were resected as extensively as possible. The pathological diagnosis was that of an arachnoid cyst.

![Image](image.png)

**Fig. 2.** Case 3: Preoperative computerized tomography scans, without *(left)* and with *(right)* contrast enhancement, showing a huge left frontoparietal parasagittal cyst associated with agenesis of the corpus callosum. The cyst wall is not enhanced by contrast medium.

**Postoperative Course.** The postoperative course was uneventful and the patient was discharged in good condition. However, ventricular enlargement was noted 2 months later and a VP shunt was placed. Thereafter, his course was uneventful. Follow-up CT scans showed shrinkage of the cyst and a decrease in the size of the lateral ventricles (Fig. 1 *right*).

**Case 3**

This 44-month-old girl was born at the 40th week of gestation by suction delivery. She was brought for evaluation of a generalized convulsive seizure that occurred during sleep. An electroencephalogram (EEG) showed frequent spikes and waves, predominantly in the right parietal region. She was placed on a course of anticonvulsant medication and was referred to the Takayama Red Cross Hospital because of abnormal CT scans.

**Examination.** On admission, the patient weighed 15 kg and her head circumference measured 52.4 cm (approximately 2 standard deviations above the mean for her age group). On physical examination all other characteristics were found to be within normal limits. Neurologically, no abnormality was found. Her developmental quotient was greater than 85. No evidence of increased intracranial pressure was present. Optic fundi were normal. A huge interhemispheric cystic lesion, slightly off midline to the left and associated with agenesis of the corpus callosum, was revealed by CT (Fig. 2); the cyst wall was not enhanced by an
injection of contrast medium. The cerebral hemispheres were separated laterally and the left lateral ventricle, especially the body and occipital horn, was markedly enlarged, probably due to obstruction of the left foramen of Monro. The cyst seemed to have no communication with the lateral ventricle or subarachnoid space, and a thin septum was seen within its cavity at the level of the foramen of Monro.

Clinical Course. The patient has been followed without surgical intervention. The results of follow-up MR imaging were similar to those of the CT scans obtained 2 months previously (Fig. 3).

Case 4

This 54-year-old man with an unremarkable history and normal psychomotor development consulted a neighborhood doctor, complaining of dull pain in the occipital and neck regions. He was noted to have an abnormal CT scan and was referred to the National Cardiovascular Center.

Examination. Neurological findings on admission were unremarkable. Computerized tomography revealed a large cystic lesion with the same density as cerebrospinal fluid in the right frontal region close to the midline. This was associated with dilatation of the body and occipital horn of the right lateral ventricle, and agenesis of the corpus callosum. The falx was present. The cyst was separated from the frontal horn of the right lateral ventricle by a thin septum. There was slight bulging and thinning of the frontal bone adjacent to the cyst. Asymmetry of the lateral ventricle, the right side being larger than the left, was thought to be due to obstruction of the right foramen of Monro by the cyst. Metrizamide CT cisternography demonstrated minimal filling of the ventricle and the cyst. Magnetic resonance images revealed findings similar to those of the CT scans. The sagittal images clearly demonstrated agenesis of the corpus callosum (Fig. 4 left). Gray matter could be seen between the lateral border of the cyst and the medial aspect of the right frontal lobe in the coronal images (Fig. 4 right). This finding suggested that the cyst was extra- rather than intra-axial. Right carotid angiograms revealed opacification of the right anterior cerebral artery in the anteroposterior view. The pericallosal artery ran an abnormal course and appeared to be under slight tension. In the lateral view, this artery was seen to course posterosuperiorly over the corpus callosum.

Clinical Course. These findings confirmed the presence of an extra-axial mass lesion between the medial aspect of the right frontal lobe and the falx, and agenesis of the corpus callosum. Continuous pressure moni-

Fig. 3. Case 3: Preoperative magnetic resonance images. T₁-weighted (left) and T₂-weighted (right), showing no enlargement of the left parasagittal cyst.

Fig. 4. Case 4. Left: Magnetic resonance T₁-weighted images, sagittal view, showing agenesis of the corpus callosum and a huge cyst in the frontal region. Right: T₁-weighted images, coronal view, showing a parasagittal right frontal cyst associated with agenesis of the corpus callosum. The third ventricle is displaced downward. Gray matter is present in the medial aspect of the right frontal lobe close to the lateral border of the cyst, suggesting that the cyst is extra-axial.
Giant cysts with corpus callosum agenesis

toring in the lumbar subarachnoid space showed no pressure waves. Chromosomal examination was non-contributory. The patient has been followed without operation, and has remained neurologically intact.

Discussion

Cyst Classification

Interhemispheric cysts are congenital and usually present symptoms during childhood, but they may sometimes be diagnosed in adults, as in Case 4. Various types of cystic lesions occur in the interhemispheric or parasagittal area and they are often associated with agenesis of the corpus callosum. Previ-ously reported cases of agenesis of the corpus callosum with a midline cyst in the supratentorial compartment have variously been referred to as midline porencephaly, agenesis of the corpus callosum with porencephalic cyst, diencephalic cyst, primary or secondary interhemispheric cyst, and dorsal cyst of lobar holoprosencephaly. Some cysts communicate freely with the ventricular system. The common radiographic features are agenesis of the corpus callosum, a midline location, and ventricular dilatation. Gross psychomotor retardation has been reported in many patients with such an anomaly.

Two morphologically distinct types of cysts occur in the interhemispheric area: extra-axial cysts associated with agenesis of the corpus callosum, and intra-axial cysts. Diagnosis of the cysts with agenesis of the corpus callosum has been made easier with CT and MR imaging. Extra-axial cysts associated with agenesis of the corpus callosum should be differentiated from such intra-axial midline cysts as porencephalic cyst, dorsal cysts of holoprosencephaly, and upward extension of the third ventricle in agenesis of the corpus callosum (Table I). Because of the broad spectrum of cerebral malformations accompanying a cystic cavity in the dorsal midline, they may be referred to as dorsal cyst malformations. The differential diagnosis of midline cysts is important from both the prognostic and therapeutic points of view. Interhemispheric arachnoid cysts have been reported to be clinically benign.

Intra-Axial Cysts

"Congenital midline porencephaly," a different clinical entity, is dysgenetic hydrocephalus associated with scalp anomalies and characterized by a symmetrical defect of the bilateral posterior mantle along the midline and alopecia or cephaloeole in the parietal midline. The midline cyst is considered to be a variant of the dorsal cyst of lobar holoprosencephaly and arises from primary dysgenesis of cerebral midline structures. It is not the result of hydrocephalus.

Holoprosencephaly consists of a monoventricule communicating with the dorsal cyst in the midpial region. The falx and corpus callosum are absent. The patient may have a facial anomaly such as cleft lip and palate. Hydrocephalus seen in holoprosencephaly is considered to be intractable.

Simple agenesis of the corpus callosum is easily recognized on sagittal MR images. Dorsal extension of the third ventricle in agenesis of the corpus callosum or "diencephalic cyst" has been demonstrated in cor-onal CT scans. Agenesis of the corpus callosum by itself does not present clinical symptoms; however, its association with various congenital anomalies is well known. Consequently, the presence of agenesis of the corpus callosum may suggest such related anomalies as Dandy-Walker syndrome, Aicardi syndrome, and Chiari II malformation. The clinical symptoms depend on the nature of the associated malformations.

| TABLE I |

**Differential diagnosis of interhemispheric or parasagittal cysts**

<table>
<thead>
<tr>
<th>Factor</th>
<th>Interhemispheric Cyst</th>
<th>Porencephalic Cyst</th>
<th>Holoprosencephaly</th>
<th>Agenesis of Corpus Callosum</th>
</tr>
</thead>
<tbody>
<tr>
<td>nature of cyst</td>
<td>extra-axial</td>
<td>intra-axial</td>
<td>dorsal</td>
<td>cystic dilatation of 3rd ventricle</td>
</tr>
<tr>
<td>location of cyst</td>
<td>midline or parasagittal</td>
<td>midline or parasagittal</td>
<td>midline</td>
<td>midline</td>
</tr>
<tr>
<td>size of cyst</td>
<td>large</td>
<td>variable</td>
<td>small</td>
<td>small</td>
</tr>
<tr>
<td>mass effect</td>
<td>+</td>
<td>-</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>communication with ventricle</td>
<td>-</td>
<td>+</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>corpus callosum</td>
<td>±</td>
<td>+</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>falx</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>hydrocephalus</td>
<td>+</td>
<td>-</td>
<td>±</td>
<td>±</td>
</tr>
<tr>
<td>asymmetry of lateral ventricle</td>
<td>separation, abnormal course</td>
<td>abnormal course</td>
<td>agenous</td>
<td>abnormal course</td>
</tr>
<tr>
<td>deep cerebral vein</td>
<td>abnormal course</td>
<td>abnormal course</td>
<td>dysgenesis</td>
<td>facial anomaly</td>
</tr>
<tr>
<td>frequently associated anoma-</td>
<td>gyral abnormality, neuronal</td>
<td>abnormal course</td>
<td>facial abnormality</td>
<td>facial abnormality</td>
</tr>
<tr>
<td>lies</td>
<td>heterotopia</td>
<td>nonspecific</td>
<td>facial abnormality</td>
<td>facial abnormality</td>
</tr>
<tr>
<td>onset of symptoms</td>
<td>early or late</td>
<td>early</td>
<td>early</td>
<td>early, if present</td>
</tr>
<tr>
<td>neurological deficits</td>
<td>minimal</td>
<td>severe</td>
<td>severe</td>
<td>subclinical or severe</td>
</tr>
</tbody>
</table>

* Abbreviations: + = feature present; - = feature absent; ± = presence or absence of feature equivocal.
FIG. 5. Magnetic resonance T1-weighted image, coronal view, showing a large right parasagittal intra-axial cyst in a 39-year-old. Mass effect is minimal and the corpus callosum is present.

Extra-Axial Cysts

Interhemispheric extra-axial cysts are subdivided by location into bilateral midline cysts (Case 2 in the present series) and unilateral parasagittal cysts (Cases 1, 3, and 4). Coronal MR imaging is especially useful in differentiating interhemispheric extra-axial cysts from intra-axial cysts (Fig. 5). These cysts have no communication with the subarachnoid space or lateral or third ventricle, as can be confirmed by CT cisternography or cystography. The third ventricle is displaced downward by the cyst and thus one or both foramina of Monro may be obstructed with resultant mono- or biventricular dilatation. Because of the location of the cyst, ventricular dilatation is asymmetrical and marked in the body and occipital horn; a paper-thin mantle is sandwiched between the cyst and the dilated body of the lateral ventricle (Fig. 2). The frontal horn is compressed and displaced laterally (Fig. 4 right).

Interhemispheric cysts are unusually large and non-communicating, and are often associated with agenesis of the corpus callosum (Fig. 6). Associated neurological deficits are minimal. Neuroradiological images mimic those of porencephalic cyst, holoprosencephaly, and agenesis of the corpus callosum. These anomalies should be differentiated clinically (Table 1).

Histopathological Considerations

In two of the four cases presented here, surgical fenestration of the cysts was performed and pathological examination led to a diagnosis of arachnoid cyst. As the interhemispheric or parasagittal area is a rather uncommon site for the occurrence of arachnoid cyst, the pathological findings may be variable. Agenesis of

---

FIG. 6. Schematic illustration showing coronal sections of interhemispheric or parasagittal cysts.
Giant cysts with corpus callosum agenesis

the corpus callosum and interhemispheric agenesis containing ependyma, choroid plexus, and glial cells have been reported. \(^{(1,9,22,29,34,38-40)}\) These have been histopathologically diagnosed as neuroepithelial or gliopendymal cysts. With or without agenesis of the corpus callosum, interhemispheric or parasagittal arachnoid cysts tend to carry a favorable prognosis \(^{(10,11,44)}\) compared with neuroepithelial or gliopendymal cysts.

Neuroepithelial cysts are often associated with polymicrogyria \(^{(16,48)}\) and nodule neuronal heterotopia. \(^{(3)}\) These associated anomalies might be helpful in preoperative differentiation of neuroepithelial or gliopendymal cysts from arachnoid cysts. \(^{(15)}\) Ependymal cysts may be intra-axial (intracerebral) or extra-axial (subarachnoid) in location. \(^{(12,15)}\)

**Therapeutic Considerations**

When the cyst is large but the mass effect is minimal and the patient is neurologically intact, surgical intervention is not indicated, although the patient should be re-examined periodically. There are two principal surgical treatments for arachnoid cysts: membraneotomy and cystoperitoneal shunt insertion. \(^{(12,14)}\) Neither method is clearly preferable. In most cases, direct surgery on the cyst has been performed in order to: 1) leave the patient shunt-independent; 2) obtain sufficient decompression and communication between the cyst and the subarachnoid space and/or the ventricle; or 3) facilitate inspection of the underlying brain. \(^{(1)}\) In patients without hydrocephalus, cyst fenestration is the treatment of choice. At present, we believe that performing a craniotomy solely for diagnostic purposes is not justifiable.

Cystoperitoneal shunt insertion, however, has the following advantages: 1) in the case of a huge cyst or one associated with marked hydrocephalus, a shunt avoids extreme brain shift due to sudden decompression; and 2) the shunting operation is less invasive and thus is preferable in aged patients. Insertion of a VP shunt followed by resection of the cyst wall may also give results, especially in the presence of hydrocephalus. \(^{(10)}\)

In the group of patients reported here, craniotomy with fenestration of the cyst was found to be inadequate and a shunt was required to treat the hydrocephalus. It is also now well documented in the literature that attempts at fenestration of arachnoid cysts are unsuccessful in the vast majority of cases and that shunting is required subsequently. \(^{(6,25)}\) Considering the complications of craniotomy, and the large number of patients who need shunts even after fenestration, shunting may be the preferred initial surgical treatment for interhemispheric arachnoid cysts.

**Acknowledgments**

The author thanks Drs. Shuro Nishimura, Tomokatsu Hori, Mitsuaki Takada, and Yasuhiko Yonekawa for reviewing the cases. Gratitude is also expressed to Dr. Patrick O. Eghwurudjakpor, a clinical fellow at the Department of Neuropsychology, Kochi Medical School, for review of the English text, and Miss Chisato Hiroi for preparing the manuscript.

**References**


Manuscript received January 15, 1991.
Accepted in final form July 15, 1991.
Address reprint requests to: Koreaki Mori, M.D., Department of Neurosurgery, Kochi Medical School, Kohasu, Okobo-cho, Nankoku City, Kochi 783, Japan.