Coexistent intraventricular abnormalities in periventricular giant arachnoid cysts

Clinical article

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Objective. Arachnoid cysts are congenital lesions that arise during development by splitting of the arachnoid membrane. Large cysts can be adjacent to CSF pathways causing a marked midline shift and hydrocephalus. The association between a large arachnoid cyst and hydrocephalus has been commonly described as being due to a mass effect, but these previous reports have not focused closely on any associated intraventricular abnormalities.

Methods. Seven patients who were previously treated with a cystoperitoneal shunt presented with shunt failure, hydrocephalus, and/or cyst expansion. All of these patients had giant arachnoid cysts extending to the periventricular region from the original site, which was the sylvian fissure in 4 patients, and the suprasellar cistern, quadrigeminal cistern, and interhemispheric fissure in 1 patient each. Endoscopic exploration of the ventricular system and cyst fenestration was then performed in all patients.

Results. The endoscopic findings were obstruction of the cerebral aqueduct by a membrane not related to the cyst in 5 patients, occlusion of the foramen of Monro in 6, septum pelucidum hypoplasia in 2, and occlusion of the cerebral aqueduct by a quadrigeminal arachnoid cyst in 1. Endoscopic procedures performed were septum pelucidum fenestration and/or foraminoplasty in 5 patients, aqueductoplasty in 2, endoscopic third ventriculostomy in 5, fenestration of the lamina terminalis in 1, and direct cystocisternostomy in 1. After the endoscopic procedure, signs and symptoms of increased intracranial pressure and hydrocephalus improved in all patients, with a reduction in size of the cyst and the ventricle.

Conclusions. Ventricular abnormalities contributing to hydrocephalus may be associated with arachnoid cysts. These abnormalities may more likely reflect a common origin than a casual relation. Foramen of Monro stenosis and cerebral aqueduct occlusion associated with an arachnoid cyst can be more frequent than has been previously believed. In cases of periventricular giant arachnoid cysts, endoscopic exploration is a good alternative for examining the ventricular system and identifying and treating CSF obstructions caused by and/or related to arachnoid cysts.

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Key Words • aqueductal stenosis • flexible endoscopy • giant arachnoid cyst • hydrocephalus • neuroendoscopy • shunt failure

Arachnoid cysts are congenital lesions that arise during development by splitting of the arachnoid membrane. These cysts may become symptomatic via mass effect on surrounding structures, spontaneous rupture or hemorrhage, or obstruction of the CSF outflow pathways. The incidence of arachnoid cysts is 5 per 1000 patients in autopsy series. Most arachnoid cysts that become symptomatic do so in early childhood. The presentation of this condition varies with the location of the cyst—they can be found in the middle fossa, suprasellar, interhemispheric, diffuse supratentorial, or infratentorial regions. Diffuse supratentorial and infratentorial arachnoid cysts may cause marked midline shift, increased ICP, and hydrocephalus. Some authors have postulated a correlation between large arachnoid cysts and hydrocephalus based on an underlying aberrancy in CSF dynamics and obstruction of CSF pathways.

Some invasive and minimally invasive procedures have been proposed to treat arachnoid cysts, such as CP shunt placement, open craniotomy, keyhole microsurgery, and endoscopic fenestration with a rigid endoscope.
Methods

Patient Population

Between 2000 and 2007, 7 patients ranging in age from 6 to 22 months with periventricular giant arachnoid cysts underwent endoscopic exploration using flexible endoscopy. There were 3 female and 4 male patients. There was no history of birth trauma, intraventricular hemorrhage, or meningitis in any of the patients. All patients had been treated previously and only with a CP shunt; all returned to the hospital with neurological signs and/or symptoms of shunt failure and cyst expansion (Table 1).

Preoperative CT was performed in all patients. An arachnoid cyst in the sylvian fissure (Fig. 1A–C and Fig. 2A–C) with extension into the ipsilateral lateral ventricle and third ventricle was found in 4 patients; a suprasellar arachnoid cyst (Fig. 2A–C) with extension into the third ventricle and both lateral ventricles was found in 1 patient; an arachnoid cyst in the quadrigeminal cistern extending into the third ventricle was found in 1 patient; and an arachnoid cyst in the interhemispheric fissure extending inferiorly to both lateral ventricles and the third ventricle was found in 1 patient. In all cases there was associated hydrocephalus.

Surgical Approach

This procedure was performed while the patient was in a state of general anesthesia. With the patient supine, the scalp was shaved around the right forehead and then suitably prepared and draped. A 2-cm longitudinal skin incision was made slightly anterior to the coronal suture; 3 cm from the midline, a bur hole was placed in the same site. In children who had an open anterior fontanel, the incision for the entry point of the endoscope was made at the lateral margin of the fontanel on the right side. A blunt ventricular needle was passed into the anterior horn of the right lateral ventricle, and then the needle was exchanged for the peel-away sheath, through which the flexible cerebral endoscope (Codman, Johnson & Johnson) was introduced into the ventricle.

Results

Neuroendoscopic Findings

Within the lateral ventricles, the foramen of Monro, fornix, choroid plexus, septum pellucidum, caudate, thalamus, thalamoistriate, and anterior septal veins were found. It was imperative to recognize most of these structures to orient ourselves. Some anatomical abnormalities were observed at this point, including septum pellucidum hypoplasia in 2 patients and malformation of the roof of the third ventricle in 1 patient. Through the foramen of Monro (Fig. 3A), which was partially closed by the mass effect of the cyst in 6 patients, we reached the third ventricle, which was partially collapsed and displaced by the cyst in all patients. After further navigation into the posterior portion we found the entry of the cerebral aqueduct occluded by an ependymal membrane not related to

<table>
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<th>Case No.</th>
<th>Age (mos)</th>
<th>Sex</th>
<th>Location of Cyst</th>
<th>Endoscopic Findings†</th>
<th>Endoscopic Procedures Performed‡</th>
<th>Outcome</th>
<th>FU Period (mos)</th>
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<td>asymptomatic</td>
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<td>2</td>
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<td>suprasellar cistern</td>
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<td>seizures</td>
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</table>

* All patients previously experienced CP shunt failure, and were shunt-free after the endoscopy procedure. Abbreviations: CN = cranial nerve; FM = foramen of Monro; FU = follow-up; LV = lateral ventricle; SP = septum pellucidum; TV = third ventricle.
† All patients had a deformed third ventricle in addition to the endoscopic findings listed.
‡ All patients underwent a cystoventriculostomy in addition to the procedures listed.

Shunt failure has been one of the main problems in patients treated with a CP shunt; it has been reported that at least 50% of patients will require shunt revision at some point. The use of endoscopic methods to create areas of communication between the cyst and the ventricular system, and/or the subarachnoid spaces, is an alternative to CP shunt treatment of periventricular arachnoid cysts using cerebral endoscopy.

TABLE 1: Summary of 7 patients with periventricular giant arachnoid cysts*
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endoscopic Procedures

Of the 6 patients with stenosis of the foramen of Monro, a septum pellucidum fenestration (Fig. 3B) was performed in 5 patients and a direct opening of the right foramen of Monro in 1 patient. These 2 procedures were performed to gain entrance to the third ventricle and to allow communication between both lateral ventricles.

Cerebral aqueduct occlusion was observed in 6 patients, and the entrance of the aqueduct was closed by a membrane in 5 of these patients (Fig. 3D). The occluding membrane was not related to or in continuation with the arachnoid cyst. In the other patient with aqueductal stenosis (Case 5), the arachnoid cyst deformed the posterior portion of the third ventricle and occluded the cerebral aqueduct. Aqueductoplasty was performed in 2 patients (Fig. 3E), and ETV was performed in the next 5. In 1 of the patients a fenestration of the region of the tuber cinereum could not be performed due to the aberrant anatomy of the third ventricle, and therefore a fenestration of the lamina terminalis (Fig. 3C) was performed.

Cysternoventriculostomy was performed in all patients; the cyst was in communication with the ipsilateral lateral ventricle in the 4 patients with an arachnoid cyst located in the sylvian fissure. In patients with suprasellar...
and interhemispheric cysts, the cyst was in communication with both lateral ventricles and the third ventricle; in the patient with the quadrigeminal cistern cyst, the cyst was in communication with the third ventricle. It was possible to produce communication between the arachnoid cyst and the subarachnoid basal cistern (cystocisternostomy) in 1 of the patients with an arachnoid cyst in the sylvian fissure.

The catheter of the shunt was observed directly (Fig. 3F) with the endoscope, which was covered and occluded by the cyst wall in all patients. During the procedure, the adhesions were detached with micromonopolar and micrograsping forceps. The endoscope and the protective sheath were then withdrawn, and a piece of Gelfoam (Pfizer) was placed over the dura. In cases in which the incision was made through the anterior fontanel, the incision was sutured tightly to avoid a postoperative fistula.

There were no deaths in this patient series or any kind of surgical complications, and no patient required another type of neurosurgical procedure. After the endoscopic procedure, signs and symptoms of increased ICP and shunt malfunction disappeared in all patients, and the ventricular system and the cyst were reduced in size and resumed a normal appearance (Fig. 1D–I and Fig. 2D–F). Two patients who continued to experience hemiparesis and seizures had had these symptoms since birth. A single patient experienced transient palsy of the cranial nerve VI after surgery (Table 1). Cerebral function was normalized in all patients. Follow-up CT was performed in all cases, showing that the arachnoid cyst and ventricles were reduced in size after the endoscopic procedures (Fig. 1D–I and Fig. 2D–F).

Discussion

Most arachnoid cysts remain static fluid-filled compartments throughout a patient’s life; some become enlarged, causing mass effect on adjacent neural structures. The majority of arachnoid cysts become symptomatic in early childhood. The exact mechanism by which arachnoid cysts expand is unknown. Some investigators believe that the enlargement of the cyst appears to be the result of CSF infiltration. Other investigators believe that arterial pulsations via one-way valve mechanisms at the communication site may be another possible mechanism of cyst growth, which has been confirmed during endoscopic procedures. In those reports a slit-valve mechanism in the arachnoid cyst next to the basilar artery was observed. Fluid may be secreted directly into the cyst by arachnoid cells or by ectopic choroidlike structures.

Indications for operative treatment are variable with arachnoid cysts and should take into account a patient’s age. There is no doubt that surgical treatment is necessary in patients with increased ICP and corresponding clinical symptoms. A demonstration of cyst growth and neural compression, even if asymptomatic, should be an indication for consideration for surgery, especially in children,
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allow the potentially normal development and function of the adjacent brain.

Controversy still exists about the best treatment for intracranial arachnoid cysts. Open surgery remains an acceptable treatment option for many neurosurgeons. Complete excision of the cyst membrane appears to be the logical treatment, but unfortunately, this is rarely performed because of the close anatomical relationship between the membrane and the underlying neural tissue. It has been reported that almost 70% of patients with a craniotomy and cyst fenestration will eventually require a CP shunt. In all our cases, the arachnoid cysts were related to the lateral or third ventricle, and therefore an open craniotomy and microsurgical fenestration would involve potential morbidity. Although shunt insertion is safer than open surgery, it can also sometimes lead to complications, such as obstruction, infection, unexpected hemorrhage, and life-long shunt dependence.1,10,18,25,27

Abnormalities Related to Arachnoid Cysts

Abnormalities associated with arachnoid cysts include cranial asymmetry, agenesis of the corpus callosum, deficient cerebellar lobulation, Chiari malformation Type I, arteriovenous malformation, posterior fossa cyst invaginated into the lateral ventricle, and aqueductal stenosis.17 The association of hydrocephalus with arachnoid cysts has been reported in as many as 15% of patients with cysts in the quadrigeminal region and in the supra- and/or retrocerebellar region. In a prior report,4 associated aqueductal obstruction was secondary to the cyst, similar to one of our cases. In the present series of giant arachnoid cysts, 5 of our patients had aqueductal stenosis caused by a thin ependymoglial membrane (Fig. 3D) that was not related to the arachnoid cyst. This finding was observed in 3 of the patients with arachnoid cysts in the sylvian fissure, in 1 patient with a cyst in the interhemispheric fissure, and in 1 patient with a cyst in the suprasellar region. To the best of our knowledge, associated aqueductal stenosis not caused by the cyst itself, which was diagnosed using direct endoscopic visualization, has not been previously described. Alterations in the CSF pathways have also been described in association with arachnoid cysts; whether the presence of aqueductal stenosis is the cause of the arachnoid cyst enlargement needs to be defined. Abnormalities elsewhere within the brain, such as agenesis of the corpus callosum, aqueductal stenosis, Chiari malformation Type I, and deficient cerebellar lobulation subjacent to the arachnoid cyst suggest a later insult in embryogenesis.17

Endoscopic Approach in Arachnoid Cysts

Neuroendoscopy, as part of minimally invasive neurological surgery, has been an acceptable alternative to extracranial shunt placement. The morbidity associated with chronic and multiple shunt revisions is eliminated with a successful endoscopic procedure. Endoscopic third ventriculostomy and aqueductoplasty are the most commonly used procedures in cerebral endoscopy. Alternative procedures include fenestration of the septum pellucidum, fenestration of the cyst wall in associated arachnoidal or ventricular cysts, and fenestration of the lamina terminalis (Fig. 3C) when performing an ETV is not possible.16

The progression of congenital arachnoid cysts has rarely been documented.19 In some cases the development of arachnoid cysts may be pathogenically related to impaired CSF dynamics associated with preexisting hydrocephalus, eventually producing life-threatening intracranial hypertension with a midline shift or displacement of the brainstem (Fig. 2A).
Endoscopic fenestration reduces complications involving shifts of the intracranial structures attributable to rapid decompression. In the present patient series we preferred to use a flexible endoscope in all cases because of the complex anatomical configuration and structural abnormalities associated with arachnoid cysts inside the ventricular system, including those that occlude the normal CSF pathways. Analog rigid endoscopes may have better picture quality, and for some neurosurgeons they provide better orientation. We observed deformities of the third ventricle, displacement of the septum pellucidum, aqueductal stenosis, and obstruction of the ventricular catheter by the arachnoid cyst wall. After the septum pellucidum fenestration was performed to equilibrate pressure between both lateral ventricles, multiple arachnoid cyst fenestrations were performed to allow communication between the cyst within the ventricular system and the basal subarachnoid space. It has been emphasized in the literature that wide fenestrations should be performed during endoscopic cystoventriculostomy and cystocisternostomy to reduce the rate of recurrence during long-term follow-up. Suprasellar arachnoid cysts conventionally communicated with the basal cisterns and the ventricles by ventriculocistocisternostomy. Because of the complexity of our cases, direct fenestration into the subarachnoid space was achieved in only 1 patient, but either additional ETV at the region of the tuber cinereum or fenestration of the lamina terminals achieved communication with the basal cisterns indirectly. Endoscopic third ventriculostomy either at the region of the tuber cinereum or lamina terminalis has been well described for the treatment of hydrocephalus associated with aqueductal stenosis. After the endoscopic procedure, the intraventricular catheter within the cyst was liberated and detached with removal of the shunt during the same neurosurgical procedure.

The reduction in the arachnoid cyst size after an endoscopic procedure is variable. Indications for further surgical interventions are given, not by the radiographic findings, but by the patient’s symptoms. Permanent and satisfactory clinical improvement has been reported to occur despite only a moderate or minimal reduction of cyst volumes. Especially in patients treated with endoscopy, it may take more time to observe a decrease in cyst size and volume, based on imaging (Fig. 1G–I).

Based on these findings and results, we believe that endoscopic procedures facilitate the discovery of additional intraventricular pathologies associated with arachnoid cysts. In addition, this procedure has the advantage of avoiding the known complications of a CP cyst and avoids a major craniotomy. Considering these factors, we suggest that an endoscopic approach be the initial treatment option for most arachnoid cysts, and definitely the first option for ventricle-related arachnoid cysts due to their related abnormalities that cannot be treated by only treating the cyst.

In patients with arachnoid cysts, coexistent hydrocephalus may be the result of either direct obstruction of CSF pathways due to the mass effect of the cyst or to associated anomalies such as aqueductal stenosis unrelated and distant to the cyst. With conventional CP shunt placement or a craniotomy with cyst wall excision, hydrocephalus in certain groups of patients will be adequately treated as the decompressed cyst no longer obstructs CSF pathways. However, those procedures would not address the hydrocephalus in the other group of patients who have associated abnormalities unrelated to the arachnoid cyst wall. As described in 6 patients in this report, there was associated aqueductal stenosis that would have required additional ventriculoperitoneal shunting to relieve the hydrocephalus. Use of a flexible neuroendoscope facilitates a thorough exploration of the ventricular system, thus treating these abnormalities during the same surgical procedure and eliminating additional shunt procedures.

**Conclusions**

In our experience, patients with periventricular arachnoid cysts not infrequently have some other alteration in the CSF pathways that contributes to hydrocephalus, such as stenosis of the foramen of Monro and cerebral aqueduct. Microfenestration with cyst wall excision or the insertion of a CP shunt do not provide enough opportunity to treat the hydrocephalus in patients with ventricle-related arachnoid cysts and have a relatively higher incidence of failure. Exploration using the flexible cerebral endoscope is a good treatment alternative in cases of ventricle-related giant arachnoid cysts, and the ventricular system can be explored. With the significant advances in endoscopic instrumentation and experience, most of the arachnoid cysts should be managed endoscopically.

**Disclaimer**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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