Intracranial Arachnoid Cysts in Children: A Review of Pathogenesis, Clinical Features, and Management

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Arachnoid cysts are developmental anomalies that are most often diagnosed in childhood. They are often discovered as incidental findings found on imaging. Occasionally they may produce symptoms because of expansion or bleeding. There may be underlying maldevelopment of the cortex especially the temporal lobe. There is controversy regarding the role and the type of surgery indicated in its treatment. Recent descriptions of aphasia and attention-deficit disorders associated with these cysts indicate that we do not fully understand this entity. There is also no acceptable explanation for the male preponderance and increased incidence on the left side. The distribution, clinical features, treatment modalities, and some unusual syndromes associated with arachnoid cysts in children are discussed in this review. © 2002 by Elsevier Science Inc. All rights reserved.


Introduction

Arachnoid cysts are collections of fluid that develop within the arachnoid membrane because of splitting or duplication of the structure. In many instances the recognition of an arachnoid cyst is an incidental finding in symptomatic children examined for head injury or in infants with macrocrania. Arachnoid cysts are relatively rare. The reported incidence accounts for only 1% of intracranial space-occupying lesions. An apparent increase in frequency and a shift in age distribution toward the first years of life have been described in recent years, likely reflecting the widespread use of computed tomography (CT), magnetic resonance imaging (MRI), and ultrasound scan [1]. Arachnoid cysts are nearly always sporadic and single. Males are involved in more than two thirds of cases. The great majority of arachnoid cysts are now detected in the first two decades of life.

True arachnoid cysts are congenital. Secondary arachnoid cysts may result from postinflammatory accumulation of cerebrospinal fluid (CSF) in the subarachnoid space in patients with head injury, intracranial hemorrhage, or infection [2]. Occasionally they may become symptomatic because they enlarge and interfere with adjacent neural structures or CSF circulation. The signs that have been attributed to arachnoid cysts include cranial enlargement, epileptic seizures, hydrocephalus, and psychomotor retardation [3].

Why Do Arachnoid Cysts Expand?

Arachnoid cysts could be primary or possibly could be caused by an impairment of the CSF drainage generated by venous agenesis. Several mechanisms could account for the enlargement of these cysts: secretion by the cells forming the cyst walls, a unidirectional valve, or liquid movements secondary to pulsations of the veins [4]. A number of interesting theories have been proposed to explain the expansion of arachnoid cysts. Some of them include the following:

(1) The ball valve hypothesis: There may be an anatomic communication that acts functionally as a one-way valve between the cyst and the subarachnoid space. MRI studies have demonstrated this form of one-way pulsatile movement of CSF. A slit-valve mechanism...
has been observed by means of cine-mode MRI preoperatively and confirmed during the endoscopic intervention [5]. The presence of a ball valve has not been universally demonstrated, and this theory does not explain the reduction in size and the disappearance of arachnoid cysts that is sometimes observed.

(2) Osmotic gradient between cystic content and cerebrospinal fluid: This theory is not supported by evidence because the cystic content is similar in composition to CSF. No evidence was found for either a tight sealing of the extracellular spaces in the wall of the cyst nor for the existence of an active transcellular fluid movement in some studies [6]. Congenital arachnoid cysts are maldevelopmental anomalies and contain clear CSF-like fluid. In acquired cysts the intracystic fluid may be hemosiderin-stained or may contain inflammatory cells with a possible gradient [7].

(3) Fluid production by the cells lining the walls of the cysts: There is some morphologic and ultracytochemical evidence to support the secretory nature of the cyst wall. Clinical evidence has already suggested this phenomenon, including intracranial pressure elevation and expansion in some cases, and the observation that arachnoid cysts constitute closed compartments with a fluid content that cannot be derived from other CSF-containing spaces. Ultrastructurally, the cyst lining demonstrates a similarity to subdural neurothelium and the neurothelial lining of arachnoid granulations in morphologic features, such as intercellular clefts with sinusoid dilatations, desmosomal intercellular junctions (on which tonofilaments may be abutting), pinocytotic vesicles, multivesicular bodies, lysosomal structures, and the presence of a basal lamina. Some of these features, together with the presence of microvilli on the luminal surface, are consistent with fluid secretion [8]. Moreover, enzyme cytochemistry demonstrated (Na+ + K+)-adenosine triphosphatase in the plasma membranes lining the cavity, either directly (the apical membranes), or via the intercellular clefts (the basolateral membranes), and, with alkaline phosphatase occupying the opposite plasma membranes, this structural organization indicates fluid transport toward the lumen. The argument against continuous secretion is that the cysts often remain static in size and sometimes disappear, thus demonstrating that secretion is neither universal nor, likely, the only mechanism involved.

Computed tomography after metrizamide intrathecal has demonstrated partial penetration of the contrast even 8-12 hours after injection. Cine-mode MRI with a retrospective EKG-gated flow-sensitive fast imaging with steady-state precision sequence has demonstrated the certainty of communication between arachnoid cysts and neighboring CSF spaces with an accuracy of 90% [9]. The rate of filling of the cyst may have prognostic implications.

Ultrastructural and Morphologic Features

Arachnoid cysts most likely originate from a minor aberration in the development of the arachnoid that leads to splitting or duplication of the membrane. It has been postulated that the cyst develops from a defect in condensation of the mesenchyme or from abnormalities of CSF flow. The association of other developmental abnormalities of the brain, such as heterotopias, lend support to this developmental theory. Two hundred and eight reported cases of arachnoid cysts were analyzed by Rengachary and Watanabe [10], and they found that the structural features of the arachnoid cyst wall that distinguish it from the normal arachnoid membrane were as follows: (1) splitting of the arachnoid membrane at the margin of the cyst; (2) a thick layer of collagen in the cyst wall; (3) the absence of traversing trabecular processes within the cyst; and (4) the presence of hyperplastic arachnoid cells in the cyst wall.

Miyagami and Tsubokawa [11], in a study of five cases, reported that the structure of the arachnoid cyst wall was similar to that of the normal arachnoid membrane and that the inner surface of the arachnoid cyst wall was formed of one or several layers of arachnoid cells with slender processes, which contained large extracellular spaces but not microvilli. These cysts appear to be truly intra-arachnoid in location and appear to be formed by splitting or duplication of the arachnoid membrane [12]. Schachman and Friede [13], in a study of nine cases, found that the dominant phenomenon of the wall of the cyst was an absence of the normal trabeculation of the subarachnoid space; the trabecules being replaced by tightly packed collagen fibrils and a few scattered cells in between. They found no evidence for either a tight sealing of the extracellular spaces in the cyst's wall or for the existence of an active transcellular fluid movement.

Cysts in Special Locations and Their Clinical Significance

Sylvian Fissure/Middle Cranial Fossa

Nearly two thirds of pediatric arachnoid cysts are located in the sylvian fissure/middle cranial fossa. They may occasionally increase in volume, opening the fissure and exposing the middle cerebral artery. This exposure may result in compression and underdevelopment of the anterior superior surface of the temporal lobe. The origin of the cysts has been the subject of debate since they were first described. There is still controversy concerning whether they originate directly from the meninges adjacent to the temporal pole or whether partial agenesis of the temporal lobe favors secondary formation of the cyst [14]. The left hemisphere is more often affected [15], and headache is the most common presenting symptom. Proposals [16], contralateral motor weakness, and seizures [17,18] have been described. Mental impairment has been said to be associated in 10% of cases, with developmental delay.
more common in these patients. Macrocrania and asymmetric bulging of the skull may be present in infants. Therapy, when indicated, includes excision of cyst membrane and placement of cystoperitoneal shunts [19]. It is important to recognize that children with bitemporal arachnoid cysts may have glutaric aciduria type I (GAT1) and that even simple surgical procedures may be extremely harmful for such patients. All pediatric patients with bitemporal arachnoid cysts should therefore be screened for GAT1 before any surgical procedure takes place, especially if there is also macrocephaly, an acute encephalitis-like illness, or a dystonic, cerebral palsy-like condition [20]. Bitemporal arachnoid cysts are extremely rare, may occur unrelated to GAT1 [21], and have been reported in conditions such as neurofibromatosis [22].

Sellar Region Cysts

Cysts in the sellar regions are typically present in pediatric patients. Two types are described—suprasellar and intrasellar. Suprasellar cysts may cause obstructive hydrocephalus of the third ventricle at the level of the foramen of Monro. Some think the cyst and hydrocephalus are part of a common maldevelopment. Other symptoms described are visual impairment and endocrine dysfunction [23,24]. Enlarged head, growth retardation, developmental delay, and bitemporal hemianopia have all been described.

A bobble-head doll syndrome with 2- to 3-second anteroposterior involuntary movement of the head because of abnormal pressure on the third ventricle and dorsomedial thalamic nuclei has been described [25]. Surgical options include bypass shunting or resection.

Posterior Fossa Arachnoid Cysts

Arachnoid cysts of the posterior fossa are uncommon and must be differentiated from other cystic malformations of the posterior fossa, such as Dandy-Walker malformation. The foramen of Magendie is present in arachnoid cysts. Macrocrania and raised intracranial pressure are the most common signs. Cerebellar cysts demonstrate nystagmus and other cerebellar signs. Other rare manifestations reported include cervical spinal cord compression, which improved after posterior fossa cystoperitoneal shunting [26]. In one study of 12 patients with posterior fossa arachnoid cysts the most common presenting symptoms were gait disturbance and headache. Symptoms reversed with re-expansion of the cerebellum in 11 of 12 patients and one patient died [27]. Multiplanar MRI may provide sufficient evidence for the diagnosis of posterior fossa cysts, especially in cases of rotation or upward displacement of the cerebellar vermis. However, the cyst membrane and the communication of fluid between the cyst and the cistern cannot be demonstrated on MRI [28].

Fetal Diagnosis of Arachnoid Cysts

Because arachnoid cysts are congenital, it is possible to identify arachnoid cysts in the fetus. Widespread use of ultrasound has resulted in antenatal diagnosis of arachnoid cysts. It remains uncertain whether they anatomically similar and whether they behave the same as those detected postnatally. Some arachnoid cysts have disappeared before birth. Postnatally, many are asymptomatic and remain quiescent for years, although others expand and cause symptoms by compressing adjacent brain or expanding the overlying skull. The diagnosis of arachnoid cysts in the antenatal period is difficult and may be confused with other cystic structures [29,30].

Significance of Arachnoid Cysts in Specific Clinical Situations

Epilepsy

There are studies demonstrating seizure reduction after removal of arachnoid cysts [31]. It is unknown whether there is a relationship between epileptic seizures and intracranial arachnoid cysts without obvious intracranial pressure signs. Comparative studies of patients treated surgically and conservatively have sometimes failed to demonstrate any significant difference in outcome [32,33]. Intercital and ictal electroencephalographic changes may not correspond to the site of the arachnoid cyst, raising the question of whether it is only an incidental finding. For epilepsy surgery in general, congruence between the electroencephalogram and imaging is an important factor for predicting long-term prognosis [34].

Subdural Hematoma and Hygroma

Subdural hematomas and hygromas are infrequently encountered complications of arachnoid cysts of the middle cranial fossa and are particularly rare with cysts of other regions. Acute subdural hematomas and hygromas as a complication of arachnoid cysts has been widely reported in the literature [35]. Minor head trauma has been indicated to be a precipitating factor. Arachnoid cysts of the middle cranial fossa were found in 2.43% of patients with chronic subdural hematomas or hygromas in one report. This indicated a fivefold greater prevalence of arachnoid cysts compared with a control group [36].

Attention-Deficit–Hyperactivity Disorder and Speech and Developmental Delay

There have been reports demonstrating a relationship between the presence of temporal lobe arachnoid cysts and attention-deficit–hyperactivity disorder (ADHD) [37]. The cysts in the patients reported were situated in the left middle cranial fossa. The symptoms of ADHD improved with stimulants, although a causal association with the ADHD was considered plausible by the authors because of
the presence of learning and language disabilities in these children that might be explained by temporal lobe and sylvian region pathology. A possible mechanism may be the increase in the volume of the cyst compressing the anteroinferior surface of the temporal lobe. Mental impairment and developmental delay have been associated with large arachnoid cysts. The cysts and developmental delay may be part of a common developmental aberration. There is some evidence that improvements in cognitive function can be demonstrated postoperatively in temporal lobe arachnoid cysts [38]. These results indicate that arachnoid cysts in the left temporal fossa may impair cognition, that neuropsychologic tests are required to disclose such impairments, and that decompressive surgery may improve cognition. The increased incidence of arachnoid cysts in conditions such as Down syndrome [39], mucopolysaccharidosis [40], schizencephaly [41], and neurofibromatosis [42] may point toward a higher incidence in children with underlying abnormalities of the brain.

**Obstructive Hydrocephalus**

Obstructive hydrocephalus has been described in association with intracranial arachnoid cysts in children and is considered an indication for surgery. It has been proposed that the bobble-head doll syndrome is secondary to intermittent obstruction at the level of foramina of Monro [43,44]. There have also been reports of arachnoid cysts causing obstruction at the level of the fourth ventricle [45]. Infratentorial posterior fossa cysts presenting as hydrocephalus are also well known. These need to be differentiated from Dandy-Walker malformations. Intraventricular arachnoid cyst have been reported in the fourth [46] and lateral ventricles [47].

**Aphasia**

There have been reports of aphasia including Landau-Kleffner syndrome associated with left sylvian arachnoid cyst. Even in cases in which CT and MRI failed to reveal mass effect, positron emission tomography (PET) demonstrated hypometabolism in speech areas. Postoperative improvement in PET studies corresponded to improvement in vocabulary [48]. Studies using PET have been performed to determine whether left-hemisphere arachnoid cysts lead to reorganization of the language function. Group analysis demonstrated that patients revealed no more right-hemisphere activation than a matched control group. Several patients had clear language localizations in the left hemisphere during language comprehension; none of the patients demonstrated right-hemisphere activation. The authors concluded that left-hemisphere tissue must suffer considerable compromise before reorganization of language into the right hemisphere becomes necessary [49]. This observation is interesting and needs to be validated by additional studies.

**Are Arachnoid Cysts Responsible for Presenting Symptoms?**

Whether arachnoid cysts are responsible for presenting symptoms is a critical question that is often answered retrospectively, for example, in cases of removal of cyst leading to cessation of seizure. The most direct link is found in situations, such as obstructive hydrocephalus, in which the cyst is demonstrated to cause the obstruction and the symptoms are relieved after surgery. The cause-effect relationship is much more tenous with symptoms such as attention-deficit disorder, aphasia, and migraine-like headaches. Before considering surgery for intractable symptoms attributed to arachnoid cysts, this relationship should be explored in detail and objective criterion should be used. Examples for such an approach include electroencephalographic/imaging congruence in case of intractable epilepsy and impaired function demonstrated by PET in conditions such as aphasia. It should be remembered that associated neurologic disorders cannot always be attributed to the cyst. If surgery is being considered, a causal relationship between the symptom and the cyst should be plausible [50].

**Distribution of Arachnoid Cysts**

In one study of 67 children under 11 years of age (41 males and 26 females) with arachnoid cysts, 31 (42.2%) were supratentorial (interhemispheric, nine; temporal fossa, 10; convexity, five; sylvian fissure, three; suprasellar and/or retrostellar, four); 31 (46.2%) were infratentorial (supracerebellar and/or retrocerebellar, 22; foramen of Magendie, three; quadrigeminal cistern, five; pontocerebellar, one); and five (7.5%) were supratentorial and infratentorial. Macrocephaly was the presenting symptom in 48 cases (71.5%) [51]. In another study of 285 children with arachnoid cysts, 40% were found to be in the midline, with the most common site the sylvian fissure [52]. A third study of 34 cases of childhood intracranial arachnoid cysts revealed that two thirds (23) were located supratentorially, and the remaining 11 had an infratentorial localization. Of the first group, 15 were situated at the sylvian region, six were hemispherical, and two were suprasellar [53].

**Imaging**

On CT, arachnoid cysts are most often observed as extra-axial cysts with the density of CSF, which does not enhance. Adjacent calvarial remodeling is common, as is hypoplasia of the adjacent brain parenchyma, especially in middle cranial fossa arachnoid cysts. MRI signals are similar to CSF in T1- and T2-weighted imaging with no enhancement on gadolinium. Signal from the contiguous brain is of normal intensity. The absence of signal from cortical bone and the ease of obtaining multiplanar views enable the margins of the cyst, its contents, and the full extent of the lesion to be easily defined [54]. Brookes et al.
demonstrated pulsatile movement of cerebrospinal fluid in the cyst on MRI [55].

It has been demonstrated that diffusion-weighted MRI can be useful in distinguishing between arachnoid cysts and epidermoid tumors [56]. In most cases the CT differential diagnosis may include other cystic collections, such as craniopharyngioma, epidermoid, astrocytoma, and chronic subdural hematoma. However, on MRI the combination of extra-axial location, morphologic features, and signal intensity matching that of CSF allows one to make the diagnosis of an uncomplicated arachnoid cyst with confidence [57].

Ninety percent of arachnoid cysts in some reports were found in supratentorial locations and 10% were found in the posterior fossa. The most common supratentorial site is the middle cranial fossa (60%). Other sites include the quadrigeminal plate, sellar region, and convexity [58]. MRI may occasionally reveal a lesion that was isodense with the brain on CT [59]. Cisternography, metrizamide injection, and angiography are sometimes used for preoperative patient selection.

Treatment

Conservative management has been proposed for patients who do not demonstrate signs of increased intracranial pressure or focal neurologic signs because of the morbidity associated with the surgery. Surgery may also be indicated in cases with subdural hematoma and hydrocephalus and in carefully selected patients with epilepsy. The two most commonly used surgical procedures are cystoperitoneal shunting and cyst fenestration, and there is controversy on which is the better procedure [60,61]. Total cyst excision often is not possible, and most children do well with shunts. Surgery is said to have relieved symptoms such as intractable headache and epilepsy, although the role of surgery remains controversial and needs to be individualized. Prophylactic surgery has a few proponents but is generally not recommended.

Conclusion

Arachnoid cysts are benign congenital collections of CSF. Secondary arachnoid cysts may result from trauma or infection. In most cases, they are incidental findings, although they may become symptomatic because of an increase in size or hemorrhage [62]. Long-standing pressure effects may cause maldevelopment of the temporal lobe and may produce obstruction at the level of the third or fourth ventricle.

With widespread use of imaging, many arachnoid cysts are detected during the antenatal/neonatal period. Long-term follow-up has indicated that some are quiescent throughout life, some may remain dormant for many years before demonstrating signs, and some may even disappear [63]. Occasionally they may be multiple [64]. Symptoms depend on size and location. Seizures and headache are said to be the most common symptoms of middle cranial fossa cysts. There seems to be a definite increase in the incidence of subdural bleeds in these patients. Recent descriptions of aphasia, ADHD, and developmental delay are interesting and need additional confirmation.

Endocrine dysfunction and movement disorders have been described in association withellar cysts and with cysts in the third ventricular region, respectively.

The definitive treatment for arachnoid cysts is surgery; cystoperitoneal shunting is likely technically easier in children. Recent advances in neuroendoscopic surgery may alter this, and better results with fenestration have been reported recently [65]. The indications for surgery are not always clear-cut. The only absolute indications for surgery most likely are the presence of progressive hydrocephalus [66] or intracranial hypertension.

References

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