Intracranial arachnoid cysts: Current concepts and treatment alternatives

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Abstract

Arachnoid cysts are non-tumorous intra-arachnoid fluid collections that account for about 1% of all intracranial space-occupying lesions. In this article, we review the current concepts about these lesions and discuss the treatment alternatives. The aetiology of arachnoid cysts has been a controversial subject. They are regarded as developmental abnormality of the arachnoid, originating from a splitting or duplication of this membrane. The establishment of a single CSF space, by surgically communicating the cyst with the ventricular system or basal cisterns, appears to offer the best chance of a success in the treatment of arachnoid cysts. Long-term prognosis for patients with arachnoid cysts and well-preserved neurological conditions is good, even in the case of subtotal excision. Clinical follow-up and MRI allow earlier diagnosis of recurrence.

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Keywords: Arachnoid cyst; Computed tomography scan; Cysto-peritoneal shunt; Hydrocephalus; Magnetic resonance imaging; Neuroendoscopic treatment

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1. Introduction

Arachnoid cysts are non-tumorous intra-arachnoid fluid collections that account for about 1% of all intracranial space-occupying lesions [1,2]. These are the most common intracranial cysts [3] and accounts for approximately 1.5% of all intracranial masses treated in hospital [4,5]. In this article, we review the current concepts about these lesions and discuss the treatment alternatives.

2. Aetio-pathogenesis

The aetiology of arachnoid cysts has been a controversial subject and still remains unclear [6,7]. Arachnoid cysts usually arise within and expand the marigins of CSF cisterns rich in arachnoid (i.e. Sylvian fissures, suprasellar, quadrigeminal,
Arachnoid cysts consist of liquid formations surrounded by an arachnoideal sheet [6]. Arachnoid cysts can be congenital cysts (also called ‘true’ arachnoid cysts and, occur most common type) often or secondary arachnoid cysts. This should be differentiated these cysts from other types of cysts that result from CSF sequestration due to inflammatory or following traumatic processes, hemorrhage, chemical irritation, and tumors; these can be called secondary arachnoid cysts [3,6]. They differ from secondary (acquired) cysts by the structure of their walls, which contains arachnoid cells connected with unchanged arachnoid matter. In secondary cysts, which mostly occur after inflammation or brain trauma, the loculations of CSF are surrounded by arachnoid scarring. The intracranial arachnoid cysts are invariably intra-dural while the spinal cyst may be either intra- or extra-dural [10–13]. In congenital arachnoid cysts wall contains arachnoid cells connected with unchanged arachnoid matter and in secondary cysts, the loculations of CSF are surrounded by arachnoid scarring [11,12]. Congenital arachnoid cysts are maldevelopmental anomalies in which splitting or duplication of the primitive arachnoid membrane in the early embryonal life leads to collection of clear CSF-like fluid. In acquired cysts the intracystic fluid may be hemosiderin-stained or may contain inflammatory cells with a possible gradient [14]. The cyst is separated from the cortex and pia by subarachnoid space. This is small thin and transparent in places and contains several blood vessels that continue on the surface of the surrounding brain. Several theories have been proposed to explain the genesis of arachnoid cysts and include agenesis of part of the brain [7,12,15,16], a minor aberration in the development of the arachnoid [17], developmental defect in condensation of the mesenchyme or from abnormalities of CSF flow [18] (Table 1). Robinson in 1964 suggested that temporal cysts are caused by, which later filled with cerebrospinal fluid. He coined the phrase temporal agenesis syndrome. In 1971 he wrote this title should be abandoned and the syndrome takes its place amongst other arachnoid malformations. Many arguments provide evidence of the congenital genesis of primary arachnoid cysts: definitive prevalence in infancy and childhood, with most lesions diagnosed within the first two decades of life and exceptional reports of occurrences in siblings. Majority of ninety percent of arachnoid cysts (90%) are in some reports were found in supratentorial locations and 10% are located in were found in the posterior fossa. The most common supratentorial site is the middle

Table 1
Theories in the genesis of arachnoid cysts

<table>
<thead>
<tr>
<th>Theory</th>
<th>Supportive evidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Agenesis of part of the brain</td>
<td>• Definitive prevalence in infancy and childhood</td>
</tr>
<tr>
<td></td>
<td>• Most lesions diagnosed within the first two decades of life</td>
</tr>
<tr>
<td></td>
<td>• Exceptional reports of occurrences in siblings</td>
</tr>
<tr>
<td></td>
<td>• Patients may have additional malformations</td>
</tr>
<tr>
<td>Minor aberration in the development of the arachnoid that leads to splitting or duplication of the membrane</td>
<td>• Association of other developmental abnormalities of the brain, such as heterotopias</td>
</tr>
<tr>
<td>Defect in condensation of the mesenchyme or from abnormalities of CSF flow</td>
<td>• Splitting of the arachnoid membrane at the margin of the cyst</td>
</tr>
<tr>
<td></td>
<td>• Presence of thick layer of collagen in the cyst wall</td>
</tr>
<tr>
<td></td>
<td>• Absence of traversing trabecular processes within the cyst</td>
</tr>
<tr>
<td></td>
<td>• Presence of hyperplastic arachnoid cells in the cyst wall</td>
</tr>
</tbody>
</table>

Table 2
Theories to explain the increase in size of arachnoid cysts

| The ball valve hypothesis                                                                 | • An anatomic communication that acts functionally as a one-way valve between the cyst and the subarachnoid space |
|                                                                                       | • A slit-valve mechanism has been observed by means of cine-mode MRI preoperatively and confirmed during the endoscopic intervention |
|                                                                                       | • The presence of a ball valve has not been universally demonstrated                |
|                                                                                       | • This theory does not explain the reduction in size and the disappearance of arachnoid cysts that is sometimes observed |
|                                                                                       | • Not supported by evidence                                                          |
|                                                                                       | • Because the cystic content is similar in composition to CSF. No evidence for either a tight sealing of the extracellular spaces in the wall of the cyst |
|                                                                                       | • No evidence of the existence of an active transeellular fluid movement in some studies |
| Fluid production by the cells lining the walls of the cysts:                         | • Morphologic and ultrastructural evidence to support the secretory nature of the cyst wall |
|                                                                                       | • Clinical evidence of intracranial pressure elevation and expansion in some cases  |
|                                                                                       | • Ultrastructurally, the cyst lining demonstrates a similarity to subdural neurothelium and the neurothelial lining of arachnoid granulations in morphologic features |
|                                                                                       | • The argument against continuous secretion is that the cysts often remain static in size and sometimes disappear |

Liquid movements secondary to pulsations of the veins
cranial fossa (60%). Other sites include the quadrigeminal plate, sellar region, and convexity [19]. The majority of middle cranial fossa cysts (approximately 50%) are reported to be of moderate size [20]. Middle cranial fossa location may be explained by meningeal maldevelopment: the arachnoid coverings of the temporal and frontal lobes fail to merge when the Sylvian fissure is formed in early fetal life, thereby creating a non-communicating fluid compartment entirely surrounded by arachnoid membranes [21]. They occupy the anterior and middle portions of the temporal fossa and frequently displace the tip of the temporal lobe posteriorly, superiority and medially. 30% of middle cranial fossa cysts are large, occupying nearly the whole temporal fossa, sometimes extending to the frontal region and convexity [22]. Several mechanisms could account for the enlargement of these cysts and number of interesting theories have been proposed (Table 2) to explain the expansion of arachnoid cysts [17,23,24]. Some of them include the following: (1) the ball valve hypothesis [25], osmotic gradient between cystic content and cerebrospinal fluid [13], and fluid production by the cells lining the walls of the cysts [26,27].

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.

3. Clinical features

The clinical manifestations of arachnoid cysts are variable and often unspecific. In patients with arachnoid cysts neurological signs and symptoms reflect their size, anatomic distribution and their impact on CSF flow (Table 3). When small, arachnoid cysts are usually asymptomatic, but large supratentorial, suprasellar, and posterior fossa cysts may cause hydrocephalus. Large middle cranial fossa cysts may be associated with seizures, headaches, or, rarely, hemiparesis [5,28,29]. The most common presenting symptoms and signs are those of raised intracranial pressure, craniomegaly, and developmental delay. They are usually seen in cases of large supratentorial cysts, but may also be caused by smaller suprasellar or posterior fossa cysts associated with obstructive hydrocephalus. Hydrocephalus has been estimated to be present in 30–60% of patients with arachnoid cysts [6]. Other signs, such as craniomegaly, developmental delay, visual loss, and hypothalamic pituitary dysfunction have been reported [23,24]. One of the most important issue is 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 tenuous with symptoms such as attention-deficit disorder, aphasia, and migraine-like headaches [30].

4. Investigations

Diagnostic evaluation should include, not only the initial identification of intracranial arachnoid cysts but also the detection of mass effect, determination of the type of communication between cyst and subarachnoid space and recognition of the presence, location and severity of obstructive hydrocephalus and cisternal block [31]. Diagnostic tools for investigation of arachnoid cysts are computerized tomogram (CT) with contrast studies, magnetic resonance imaging (MRI), radioisotope scintigram and cine phase contrast MR imaging [32]. On CT, arachnoid cysts are observed as extra-axial cysts with the density of CSF, without any contrast enhancement. Adjacent calvarial remodeling is common, as is hypoplasia of the adjacent brain parenchyma, especially in middle cranial fossa arachnoid cysts [33]. The absence of signal from cortical bone and the ease of obtaining multiplanar views on MRI enable the margins of the cyst, its contents, and the full extent of the lesion to be easily defined. MRI signals are similar to CSF in T1- and T2-weighted imaging with no enhancement on gadolinium with normal signal from the contiguous [33]. Brookes et al. has demonstrated pulsatile movement of cerebrospinal fluid in the cyst on MRI [31].

Table 3

<table>
<thead>
<tr>
<th>Location</th>
<th>Clinical features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Supratentorial (90%)</td>
<td>Asymptomatic</td>
</tr>
<tr>
<td>Temporal (60%)</td>
<td>If large</td>
</tr>
<tr>
<td></td>
<td>Headaches</td>
</tr>
<tr>
<td></td>
<td>If the wall of the cyst is vascular</td>
</tr>
<tr>
<td></td>
<td>Can present with bleed subdurally or into the cyst</td>
</tr>
<tr>
<td>Suprasellar</td>
<td>Headaches</td>
</tr>
<tr>
<td></td>
<td>Vomiting</td>
</tr>
<tr>
<td></td>
<td>Hydrocephalus</td>
</tr>
<tr>
<td>Pineal arachnoid cysts</td>
<td>Obstructive hydrocephalus</td>
</tr>
<tr>
<td></td>
<td>Endocrinological disorders</td>
</tr>
<tr>
<td>Convexity</td>
<td>Focal neurological deficits</td>
</tr>
<tr>
<td></td>
<td>Seizures</td>
</tr>
<tr>
<td>Posterior fossa (infratentorial) (10%)</td>
<td>Headache</td>
</tr>
<tr>
<td></td>
<td>Vomiting</td>
</tr>
<tr>
<td></td>
<td>Cerebellar disorders</td>
</tr>
<tr>
<td></td>
<td>Usually associated with obstructive hydrocephalus</td>
</tr>
</tbody>
</table>

[23,24,26,27,30,31,32,33]
5. Differential diagnosis

Arachnoid cysts need to be differentiated from circumscribed areas of enlargement of compartments of the CNS that have CSF like signal intensity or density that sometimes poses a difficult diagnostic problem (Table 4) [33,34]. In some patients, a neuropathologic examination may be required to differentiate arachnoid cysts from other cystic lesions [35].

6. Natural course

The development and natural history of arachnoid cysts remain controversial as understanding of the natural history is a prerequisite to rationale therapy [36]. Most arachnoid cysts are static but occasionally involute and disappear over time [37]. A spontaneous disappearance of arachnoid cysts has been noted to be associated with head injury [37,38]. There is report where arachnoid cysts disappeared spontaneously without any major head and body trauma (only stress was usual home, school and sports activities) [39]. The basic mechanism of this natural cure is considered to be rupture of outer or inner membrane of arachnoid cyst which would facilitate absorption of its content [38,39]. In these cases it has been speculated that the cysts ruptured into cerebrospinal fluid circulation by the mechanical effects of some forced activities to the brain tissue and cyst, such as excessive breathing, coughing and sport activities. As absorption of the subdural fluid proceeded, cyst fluid leakage into the subdural space could be absorbed. In addition these factors may change the balance between intracystic and pericystic pressure and facilitate the rupturing of the cyst into subdural, subarachnoid and intraventricular spaces [37,39,40]. Yokoyama has shown such a communication scintigraphically [41]. This communication can also be visible at cine PC MR imaging in spontaneously regressed arachnoid cysts after head injury. It is still an open question whether spontaneous resolution of arachnoid cysts is caused by direct transport through the cyst wall or a valve-like mechanism through a communication [13,26].

7. Management

The large majority of cysts remain constant in size and a conservative management has been proposed for most of the patients [12,24,42,43]. Patients who do not demonstrate signs of increased intracranial pressure or focal neurological signs are also considered for conservative management as surgery may be associated with morbidity [42,43]. In these patients with close follow-up with computerized tomography (CT) and magnetic resonance imaging (MRI) is a treatment option [39]. Although there has been considerable controversy regarding the indications for the surgical treatment of asymptomatic arachnoid cysts, the literature seems to show some consensus that patients with symptomatic cysts causing seizures, hydrocephalus, increased intracranial pressure, neurological impairment and those complicated by intracystic or subdural hemorrhage should be treated [12,24,42–45]. Few authors believe that all arachnoid cysts exert a mass effect and even if asymptomatic, should be surgically managed to avoid potential risks of compression on surrounding brain structures, cyst rupture and intracystic or subdural hemorrhage [44,45]. Before considering surgery for intractable symptoms attributed to arachnoid cysts, their relationship should be explored in detail and objective criterion should be used [30].

8. Surgical approaches

The objective of surgical treatment for arachnoid cysts is to communieate the interior of the cyst with the anatomic corridors of the flow of CSF or the implementation of a shunt from the cyst or ventricular system to another cavities were the CSF could be absorb [43,46–49]. Operative techniques for the management of arachnoid cysts include microsurgical excision, cystperitoneal shunting, endoscopic ventriculocystostomy or ventriculocystocistencostomy, stereotactic cyst-ventricular shunting, and stereotactic intracavitary irradiation [50–55]. The choice of the most appropriate surgical approach is still debated and many operative procedures for the therapy of arachnoid cysts have been recommended and it remains controversial as to which is the best method (Table 5).

9. Complications

Treatment complications (subdural hematomas or hygromas) are strongly related to cyst location and size, with complications almost exclusively occurring in patients harboring Galassi types II and III cysts. The incidence of postoperative hygromas after open surgery with marsupialization is 6% [72]. Intraparenchymal hemorrhage in the underlying brain after decompression of the arachnoid cysts is an uncommon complication [73]. A case of brain stem hemorrhage after decompression of a Sylvian fissure arachnoid cyst has been reported [74]. Intracerebral hemorrhage after rapid decompression of chronic subdural hematomas is well known and hyperperfusion of underlying brain after surgical decompression has been documented [75,76]. Sgouros
Table 5
Operative procedures indications, advantages and disadvantages

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Objectives</th>
<th>Indications</th>
<th>Advantages</th>
<th>Disadvantage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Craniotomy with fenestration of the membranes [56–59]</td>
<td>• To resect the walls of cyst and communicate with the CSF flow</td>
<td>• Well circumscribed and is not associated with hydrocephalus or acute event</td>
<td>• Histopathological diagnosis possible</td>
<td>• Necessity to do a craniotomy</td>
</tr>
<tr>
<td>Endoscopic fenestration and marsupialization [25,60–64]</td>
<td>• Same</td>
<td>• Suprasellar arachnoid cysts</td>
<td>• Less aggressive, use a burr hole</td>
<td>• Needs more expertise</td>
</tr>
<tr>
<td>Shunt [65–71]</td>
<td>• Diversion of the CSF</td>
<td>• Middle cranial fossa cysts • Continuously raised intracranial pressure</td>
<td>• Simple • Less aggressive</td>
<td>• Multiloculated cysts • Any associated lesions</td>
</tr>
</tbody>
</table>

and Chapman have studied three children with middle fossa arachnoid cysts, presenting with nonspecific symptoms and otherwise well, before and after surgery with magnetic resonance and 99Tc-examethyolphyleineaminoxime single photon emission computerized tomography scans and shown that middle fossa arachnoid cysts may cause global impairment of brain function by interfering with its blood supply [77]. Although the pathophysiology of this complication is unclear, it might be due to re-perfusion injury, implying that there was raised intracranial pressure before cyst drainage. Other possible pathogenetic mechanisms of this complication are abrupt change in blood circulation, faulty autoregulation and brain decompression as the cause of superficial veins distortion [78]. If this is the case, more gradual decompression of huge Sylvian fissure arachnoid cysts using indirect surgical approaches with programmable shunts or more conservative procedures like simple tapping may theoretically decrease the incidence of such rare complications in similar cases [79].

10. Conclusions

Arachnoid cysts are benign congenital collections of cerebrospinal fluid, usually in areas that are rich in arachnoid. Symptoms depend on size and location. CT scan and MR imaging are diagnostic in the majority of cases, alleviating the need for histopathological confirmation. Small cysts with minimal symptoms should be treated conservatively with regular clinical and radiological follow-up examinations at 6-month to 1-year intervals. Large space-occupying cysts and those causing neurological impairment require surgical treatment. The definitive treatment for arachnoid cysts is surgery and the indications for surgery most likely are the presence of progressive hydrocephalus or intracranial hypertension. Though establishment of a single CSF space, by surgically communicating the cyst with the ventricular system or basal cisterns, appears to offer the chance of a success in the treatment of arachnoid cyst, but still a shunt is better and safer method in these patients.

References

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