



Surgical indications in pediatric arachnoid cysts

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Received: 21 July 2022 / Accepted: 13 October 2022 / Published online: 26 October 2022
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Abstract

Introduction Arachnoid cysts are developmental lesions consisting of CSF collections within the subarachnoid space. There are many theories and hypotheses about their pathogenesis and histopathology and this may also explain the diversities seen in clinical behaviors of these cysts, their natural history and consequently their management where there is a great controversy about selecting patients for surgical intervention. The most common location in pediatrics is the Sylvian or middle cranial fossa and this made it gain more concern and greater debate about its management where its diagnosis is often accidentally or associated with nonspecific symptoms.

Aim Our aim in this article was to review the main surgical indications for pediatric arachnoid cysts in the literature.

Conclusion We concluded that the decision for surgical intervention for pediatric arachnoid cysts is not a simple one and highly debatable and should be taken cautiously especially with sylvian arachnoid cysts that may reach a large size without symptoms. However, there is a consensus that the occurrence of symptoms definitely correlated to the presence of these cysts or their rupture with consequent subdural hematoma or hygroma are indications for surgical intervention. Large cysts in locations compressing CSF pathways causing hydrocephalus are also candidates for surgical fenestration. The surgical gain from prophylactic surgery is questionable particularly when asymptomatic.

Keywords Arachnoid cysts · Intracystic hemorrhage · Fenestration · Sylvian fissure · Surgical indications

Introduction

Arachnoid cysts are developmental lesions consisting of CSF collections within the subarachnoid space. Many theories explained their formation as splitting of the arachnoid membrane during its formation [1, 2] or secondary to trauma [3, 4]. Genetic mechanisms for their development were also suggested [5–8]. Enlargement of these cysts, when it happens, was proposed to occur by different mechanisms as one-way valve mechanism for fluid entrance, osmosis, and active secretion by cells lining the cyst lumen [9–11]. The recent histopathological studies documented different wall morphology of these cysts that varies between arachnoid-like tissue, fibrous connective tissue, and mixture of various

elements as ciliated epithelium, glial, and neuronal components [12], and this goes in hands with the previous reports that reported cysts wall resembling arachnoid tissue or containing other elements as choroid plexus or respiratory-like epithelium [13–16]. These findings made the rising hypothesis that cyst morphology may be a relevant factor in cyst growth and may have an effect on cyst fluid composition and turnover [12, 17]. All these different facts about pathogenesis and histopathology may be the basis of the diversities seen in clinical behaviors of these cysts, their natural history, and consequently their management where there is a great controversy about selecting patients for surgical intervention [18, 19]. Because the most frequent site for arachnoid cysts is the sylvian fissure, those arachnoid cysts attain a great proportion of concern and debate among neurosurgeons not only in selecting the optimal surgical technique [20–22] but also in selecting patients with actual need for surgery [23]. Sylvian arachnoid cysts were classified according to their size into 3 grade by Galassi et al. [24]; however, it is not correlated to clinical presentations of these cysts that are mostly present by non-specific symptoms and signs, or they are discovered accidentally. Our aim in this article was to review the main surgical indications for pediatric arachnoid cysts in the literature.

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Symptomatic arachnoid cysts and their locations

There is no debate that surgery is indicated in cases of symptomatic arachnoid cysts where symptoms are generally due to focal pressure effects on surrounding neural structures or globally elevated intracranial pressure by the cyst itself or secondary to associated hydrocephalus [25, 26]. Although the location of the arachnoid cyst was not found as a significant predictor for surgery by some [27], arachnoid cysts in cerebellopontine angle, suprasellar region, ambient, and quadrigeminal cisterns were found to be correlated more with the presenting symptoms due to compression on surrounding neurovascular structures or compressing a narrow CSF pathway or foramina causing hydrocephalus, opposite to those located in middle cranial fossa where correlation to symptoms may be more challenging [28] although this is the most common location in pediatrics [29].

The clinical situation where the existing neurological symptoms and signs may be poorly correlated to the presence of arachnoid cyst is not uncommon, and this makes surgical outcomes unsatisfactory as was clearly evident in one study where surgical gain was obtained only in 28% of patients and the improvement rate in the cases that showed poor correlations between the presence of arachnoid cysts and clinical symptoms was very low [30]. The decision for surgical intervention must consider the expected surgical outcome, the fact that surgery is not guaranteed to relieve symptoms [25, 31–36], and the non-negligible incidence of complications associated with fenestration procedures [30, 37–42].

Quadrigeminal arachnoid cysts are indicated for surgical fenestrations when associated with macrocrania, headaches, vomiting, lethargy, papilledema, and impairment of upward gaze and other ocular disorders [43] which are caused mainly by hydrocephalus associating arachnoid cysts in this location as they compress or distort the cerebral aqueduct at an early stage.

Suprasellar arachnoid cysts should be managed by surgical fenestration when they are presenting with signs of increased intracranial tension, affection of the visual field or acuity, motor deficits, or head nodding [44, 45]. The occurrence of isolated endocrine disorders secondary to arachnoid cysts in this location should not be considered as an indication for surgery as many authors had observed the persistence of endocrine dysfunction despite successful surgeries [45, 46].

The decision to operate on a middle cranial fossa arachnoid cyst (sylvian fissure arachnoid cyst) is the most debatable one and should be taken cautiously to select the true symptomatic arachnoid cyst in this location. Surgical patients with cysts in this location are operated upon mostly for either headache or epilepsy [23]. However, the

correlation between these symptoms and the presence of the cyst remains questionable in most cases even with workup studies done by some investigators to identify surgical patients as prolonged intracranial pressure monitoring [23, 47] where results were not significant in gray zone cases or as studies on single photon emission computerized tomography scans that still has limited preliminary data [22, 48]. The same is true regarding epilepsy workup with EEG where results showed either poor correlation between seizures and cyst location, the presence of contralateral abnormalities, and about one-fourth of cases with epilepsy showed developmental cortical anomalies far from the cyst in video EEG studies [18, 49]. Some authors recommend treating sylvian fissure cysts only when associated with hydrocephalus and papilledema; otherwise, observation is recommended even with large cysts (Galassi type III) [50].

Rupture of arachnoid cysts

Rupture of the arachnoid cyst is one of the main indications for surgical intervention [27, 51, 52]. It may be traumatic, even after minor head trauma or may be spontaneous [51, 53]. Rupture of the arachnoid cyst was reported to be associated with intracystic hemorrhage, subdural hematoma either chronic or acute, and subdural hygroma [52, 54]. About 80–95% of cases in the literature had presented with headache, and nearly 40 to 50% had nausea and vomiting, and these symptoms are supposed, in most of cases, to be related to meningeal irritation rather than increased intracranial pressure which may be the case as well. Focal neurological deficits are relatively rare but possible [51]. The source of bleeding is hypothesized to be from the unsupported surrounding bridging veins traversing the arachnoid cyst wall that has less compliance than the normal brain [55]. Microsurgical fenestration of the cyst wall into the skull base cisterns in addition to evacuation of the associated hematoma or hygroma had a good outcome in the literature as regarding decompression of the brain and elimination of the risk of rupture recurrence [51], and it should be considered the choice of management as long as it is safe and feasible, keeping evacuation of the hematoma only in cases of high risk as very young age or too thick arachnoid basal cisterns although some prefer to limit surgical intervention to dealing with hemorrhage alone especially when those cases of arachnoid cysts were symptoms free before being ruptured. Others prefer to limit intervention to hematoma evacuation, and if symptoms persist, additional arachnoid cyst surgery should be done [56, 57]. Conservative management for ruptured arachnoid cyst was adopted in some reports with good clinical outcome, but long-term follow-up was lacking [58–60].

Size and cyst progression

Using the cyst size at diagnosis may be of value in making surgical decisions as suggested by some investigators [27] where a large size equal to or larger than 68 cm^3 was found to be a significant predictor for surgery. Significantly, large cysts are more likely to compress neurovascular structures and narrow CSF spaces as cerebral aqueduct and foramina of Monro, causing hydrocephalus. This is besides a larger size of the cysts is considered a potential risk factor for cyst rupture [51, 61]. Progression or expansion of the arachnoid cyst is unlikely to occur according to the natural history of these cysts [62]. But if it happened, this radiological progression should not be an absolute indication for surgical intervention that should be reserved only for rapidly enlarging symptomatic cases [63]. Serial imaging for progressive arachnoid cysts is the most wise management where most of these cases remain asymptomatic with favorable outcome [64]; besides, there are many reports on regression and spontaneous resolution [65–67]. Even in young age which is related significantly to cyst growth, observational management should be followed as this rapid growth is retarded over time till it reaches a plateau and no further progression was found in children older than 3 or 4 years [64, 68] at the time of diagnosis.

Indications based on work-up studies

Because active CSF production from the cyst wall is one of the possible mechanisms for cyst expansion that was proved recently [10], it may be important to identify non-communicating cysts that are at higher risk of progression in comparison to cysts communicating to the surrounding subarachnoid space or ventricles. For this purpose, studies utilizing CT cisternography and phase contrast cine flow MR sequences were performed to identify non-communicating cysts. In one study, 22 out of the 28 patients studied had incomplete communicating or non-communicating cysts [69], and in another one, the cine flow MR showed that 20 out of 28 patients had non-communicating cysts [70]. All patients with non-communicating cysts were operated upon by microsurgical fenestrations with postoperative reduction of size in all of them, while all other communicating cysts were followed up for more than 3 years without detection of any progression.

Prophylactic surgery

The risk of arachnoid cyst rupture associated with large cyst size or exposure to trauma had led to the rise of the question of prophylactic surgery for these cysts. The rationale for the

debate was the reported cases of spontaneous rupture, the reported cases of sports-related head injury associated with arachnoid cyst ruptures, besides the studies describing the arachnoid cyst as a risk for chronic subdural hematoma secondary to their rupture [71–74]. In a survey addressing the management of an incidental finding of a sylvian arachnoid cyst following head trauma, 17.7% of the participants were in favor of prophylactic surgery [22]. However, the value of preventive surgery was challenged by many authors, and a consensus was established that there is no practical role for prophylactic surgery for arachnoid cyst [18, 49]. This consensus was based on many facts; first, there are many reports about incidentally found arachnoid cysts in posttraumatic imaging, and none of these reported cases had showed cyst related hemorrhage or hygroma [75–77]. The second fact is that although hemorrhage in arachnoid cyst was occasionally reported as a complication of athletic participation for those patients, this hemorrhage was rare and almost had a good long-term outcomes that made neurosurgeons reluctant to justify prophylactic surgery or even sport restriction for those patients [78]. The same was found also for subdural hygroma complicating arachnoid cyst rupture where those patients had long-lasting spontaneous resolution of their symptoms with conservative management [79]. The third fact was about the risk of surgical treatment itself where the risk of postoperative subdural hematoma/hygroma was reported to be around 5% which is similar or even higher than the risk reported for spontaneous or traumatic cases [37]. It should be mentioned also in this context that the surgery might not guarantee the prevention of cyst rupture as was found in one study where 10% of a series of an endoscopically managed sylvian arachnoid cysts developed subdural hematoma or hygroma in the follow-up period in spite of the MR evidence of reduction of cyst volume in 50% of them [33].

Conclusion

The decision for surgical intervention for pediatric arachnoid cysts is not a simple one and highly debatable; however, there is a consensus that the occurrence of symptoms definitely correlated to the presence of these cysts or their rupture with consequent subdural hematoma or hygroma are indications for surgical intervention. Large cysts in locations compressing CSF pathways causing hydrocephalus are also candidates for surgical fenestration. The growing experience with the management of sylvian arachnoid cysts points to more conservative strategy without intervention even with large cysts unless there is an increase of intracranial pressure. The surgical gain from prophylactic surgery is questionable particularly when asymptomatic.

Author contribution The main idea of the article was postulated by Mohamed El Beltagy, while literature search was performed by Abdelrhman Enayet. Both authors shared analysis of the data in the literature. The draft was written by Abdelrhman Enayet, and the manuscript was critically revised and reviewed by Mohamed El Beltagy.

Availability of data and material Not applicable.

Declarations

Ethics approval and consent to participate Not applicable.

Consent for publication Not applicable.

Conflict of interest The authors have no competing interests to declare that are relevant to the content of this article.

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