Intracranial Arachnoid Cysts in Childhood: Pathogenesis, Clinical Features, and Management

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Arachnoid cysts are mostly incidental congenital lesions. Although commonly asymptomatic, it may present with nonspecific symptoms, such as headache, epilepsy, and signs of high intracranial pressure. In cases when symptoms are evident, surgical treatment has more precise indication, but the modalities and criteria for treatment remain controversial in the literature. All this makes arachnoid cysts a challenging topic in neurosurgery, raising discussions about diagnosis, management, and prognosis. This review aimed to present, in a didactic and straightforward way, already established knowledge and more recent updates, clarifying key points to guide the management and therapeutic decision of intracranial arachnoid cysts.

Keywords: Clinical Features; Congenital Abnormality; Intracranial Arachnoid Cyst; Pathophysiology; Review Article

INTRODUCTION

Arachnoid cysts (AC) are mostly incidental congenital lesions. It was first reported in 1831 by Richard Bright and affects about 2.6% of children.[1] Although commonly asymptomatic, it may present with nonspecific symptoms, such as headache, epilepsy, and signs of high intracranial pressure, among others.[2]

ACSs do not have a defined etiology and rarely cause symptoms or complications. In cases when symptoms are evident, surgical treatment has a more precise indication, but the modalities and criteria for treatment remain controversial in the literature.[3,4] All this makes arachnoid cysts a challenging pathology in neurosurgery and raises discussions about diagnosis, management, and prognosis.

This article consists of a detailed review of the latest information on ACs to clarify key points and assist the neurosurgeon in the therapeutic decision.

EPIDEMIOLOGY

It is estimated that 1.4% of the adult population and 2.6% of children have an arachnoid cyst.[3,5] The percentage of asymptomatic patients ranges from 72.4% to 96.2%. The male:female ratio varies from 2.3:1 to 4:1. The most common symptom is a headache. [3,4,6]

The most frequent location is the middle fossa, followed by the posterior and anterior cranial fossa. Other common locations are convexity, retrocerebellar region, suprasellar cistern, quadrigeminal region, cerebellopontine cistern, and interhemispheric.[2,4,7]

PATHOPHYSIOLOGY

This is not a fully understood topic, but most authors agree that it is a congenital condition. The cyst originates from a failure in the embryological development of the arachnoid, by its division or duplication, leading to a trapping of cerebrospinal fluid.1 There are also reports of patients who developed arachnoid cysts after traumatic brain injury, with predominance in early childhood and clinical manifestation in the following 24 months. [8] Another possible mechanism is a defect of condensation of the mesenchyma or abnormal flow of the cerebrospinal fluid (CSF). [9]

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Cyst growth can occur due to the production of CSF by cells in its membrane, as well as by osmotic gradient and even by a valve mechanism of its membrane with CSF influx. [10] Cysts near the basilar artery seem to have this valve mechanism intensified by the pulsation of this vessel. [11] The ectopic choroidal plexus within the AC has also been reported in the literature. In addition, the higher expression of aquaporins on the cyst wall seems to be linked to its growth. [9]

NATURAL HISTORY OF THE DISEASE

Brain image is usually performed in head trauma, meningitis, and macrocephaly, leading to the incidental diagnosis of ACs. Therefore, most patients are initially asymptomatic, and, according to the literature and follow-up studies, almost no children develop symptoms after diagnosis.[4,6]

Even complaints such as headaches, seizures, and cognitive delay are nonspecific and do not necessarily have a direct link to the cyst. Thus, there is no pathognomonic or highly suggestive history of CAs.

It is estimated that 98% of diagnosed cysts remain stable and do not grow, while only 1.23% develop rupture and acute subdural hematoma. [4] Growth is more common in children under four years of age, although rare.[1] The larger the cyst, the greater the chances of developing symptoms, and, in situations of head trauma, cysts with a diameter greater than 5 cm are more likely to rupture.[12]

Any expansive intracranial lesion may cause some neurological symptoms, and, considering the variety of possible locations for arachnoid cysts, the symptoms are diverse. Depending on its location, growth, rupture, bleeding, or mass effect, the patient may develop more specific symptoms. [1] Clinical signs of intracranial hypertension, focal deficits such as hearing loss, visual alteration, nystagmus, facial paralysis, hemiparesis, and dysphasia are some reported symptoms, [1,2,6,7] with varied prevalence and incidence. Headache is the most reported symptom. In the study by Huang et al., [4] among 267 symptomatic pediatric patients, 37% reported headaches. Hall et al. found a similar number, with 43% of children complaining of headaches. Macrocephaly, nausea, and vomiting are also commonly reported.

The association between locations and some symptoms should be highlighted, such as the suprasellar region and endocrinopathies or hydrocephalus, the quadrigeminal region with obstructive hydrocephalus and Parinaud's syndrome, and the association between cerebellopontine angle and progressive hearing loss.[1]

Examples of uncommon symptoms reported in the literature are depression, neuropsychiatric syndromes, and cases in which symptoms improve after marsupialization of the cyst. [13] There are also reports of torticollis and gastroesophageal reflux related to a giant posterior fossa cyst. [14]

Finally, congenital clinical conditions such as dwarfism, autism, dystonia, and leukodystrophy have been reported together with the presence of ACs. [4] However, there are no reports that they cause ACs.

RADIOLOGICAL CHARACTERISTICS

Intracranial arachnoid cysts appear on computed tomography (CT) (Figure 1) and magnetic resonance imaging (MRI) (Figure 2 A and B) as simple, extra-axial, thin-walled, and well-circumscribed cysts. [7,15,16] They can displace and deform adjacent brain tissue, cause bone remodeling, and, classically, have no internal components or contrast uptake.[17] They are isodense to CSF on CT, and all MRI sequences, with no restriction on diffusion or any heterogeneous signal in FLAIR sequence. Rarely, bleeding or flow changes within the cyst may modify the appearance on MRI.[7,17,18] At cisternography, arachnoid cysts are classified as early filling, when filled by contrast in the first 24 hours, or late, when this occurs after this period.[19]

Figure 1 - CT scan, axial cut. Arachnoid cyst in the right middle fossa. CT, computed tomography.
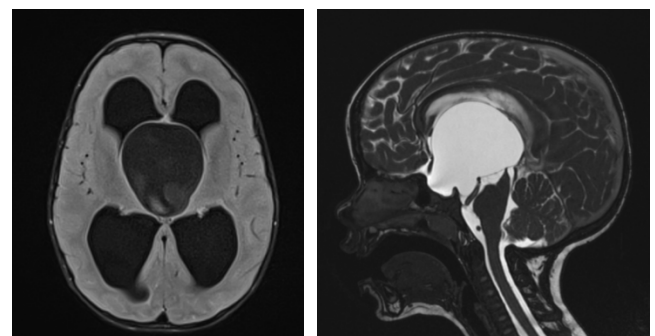
Figure 2- A, Brain MRI - T2 FLAIR axial cut. Cyst complete suppression. B, Brain MRI – Sagittal T2-weighted 3D-FIESTA acquisition. Cyst hyper signal, similar to CSF. MRI, magnetic resonance imaging. FLAIR, fluid attenuated

inversion recovery. FIESTA, Fast Imaging Employing Steady-state Acquisition. CSF, cerebrospinal fluid.

PATHOLOGICAL FINDINGS

Minimal changes in the development of the arachnoid membrane lead to its folding or duplication and the formation of cysts. Histopathology shows a cystic structure in which one of the walls consists of collagen fibrous tissue having its internal surface coated by arachnoid cells that are disposed side by side in layers of one or two cells, often forming clusters. [20] Rengachary et al.[15] described the following features as those that differentiate the arachnoid cyst from the normal arachnoid membrane: (1) duplication of the arachnoid membrane at the margin of the cyst; (2) a thick layer of collagen on the cyst wall; (3) the absence of trabecular processes traversing the interior of the cyst; (4) presence of hyperplastic arachnoid cells in the cyst wall.[1,15,21] Paler and stiffer cystic walls are usually found at older ages. Shaw et al.[22,23] associate the increase in cystic wall thickness due to the inflammatory process and minor bleeding. Long-term stress associated with pathological CSF flow may also contribute to cyst wall thickness.[23]

DIFFERENTIAL DIAGNOSIS

ACs have several possible differential diagnoses, with the epidermoid cysts (EC) being one of the most mentioned in the literature.[7,18,24–26] On CT, such pathologies can be difficult to differentiate, since both present with homogeneous and low-density content. Still, the CAs usually have rounded, thin and well-defined margins, unlike the irregularity seen in the periphery of the EC. Bone absorption adjacent to the cyst is also more frequent in ECs. [24] MRI greatly facilitates differential diagnosis in this case. The EC usually presents a lobulated configuration, which may involve cranial vessels and nerves, often with a variable degree of diffusion restriction. Finally, it is hyperintense or isointense in FLAIR due to its higher protein content.[1,17,24]

Numerous other congenital, infectious, tumoral, or vascular lesions may also be associated with intracranial cystic alterations: glial cysts, porencephalic cysts, [17,25] and intra-axial cystic neoplasms, such as pilocytic astrocytomas and hemangioblastomas. However, the former is generally surrounded by gliosis areas and is mostly intra-axial. Cystic neoplasms usually present with a well-delimited solid component adjacent to the cyst and contrast uptake.[1,17,26]

Neurocysticercosis is an important infectious cause, especially in endemic regions. However, in its usual presentation, such pathology usually occurs with several intracranial cysts with contrast enhancement, and signs of edema on FLAIR (depending on the stage of infection).[17,18]

Chronic subdural hematoma and hygroma may present different signal intensity, and their location in the subdural space is easily distinguishable on imaging. Generally, such lesions present evidence of previous hemorrhages observed on MRI and well-visualized contrast uptake in its membrane.[17,26,27] Retrocerebellar arachnoid cysts may still be confused with an enlarged cisterna magna. The position of the cerebellum falx contributes to the differential diagnosis, being displaced by the arachnoid cyst.[18](Table 1)

Table 1- Radiological differences (arachnoid x epidermoid cyst)

-	CT	MRI
Arachnoid Cyst	- Density like CSF. - If intra-cystic hemorrhage: moderately hyperintense in relation to the CSF.	- Isointense to CSF in T1 and T2. - Complete suppression to FLAIR. - No diffusion restriction in DWI.
Epidermoid Cyst	- In about 95% of the cases, the ECs are hypodense and present similarly to the CSF. - Calcifications are present in 10 -25%. - Intra-cystic hemorrhage is rare.	- Iso/hyperintense in relation to CSF both in T1 and T2. - Signal heterogeneity may be present. - no suppression or incomplete suppression to FLAIR. - Diffusion restriction in DWI.

Abbreviations: CT, computed tomography; MRI, magnetic resonance imaging; CSF, cerebrospinal fluid; FLAIR, fluid-attenuated inversion recovery; DWI, diffusion-weighted imaging; EC, epidermoid cyst.

CLASSIFICATION

ACs can be found distributed throughout the entire length of the central nervous system, being much more common in intracranial topography than in the spinal canal. The cysts of the middle fossa are responsible for most of all AC found.

Middle fossa cysts were classified by Galassi et al. [28] according to their dimensions, the mass effect exerted on adjacent structures communication with the subarachnoid space, and remain the most used nowadays.

Type I cysts are limited to the temporal pole, with small dimensions, biconvex shape, and free communication with surrounding subarachnoid spaces.

Type II cysts are the most frequent. They extend to the Sylvian fissure, have rectangular morphology, and have variable communication with the CSF flow pathways.

Type III cysts are the largest in size, occupying the entire middle fossa and reaching the whole length of the Sylvian fissure, and can occupy the anterior fossa, displace the frontal, parietal, and midline structures.[1,28] (Figure 3)

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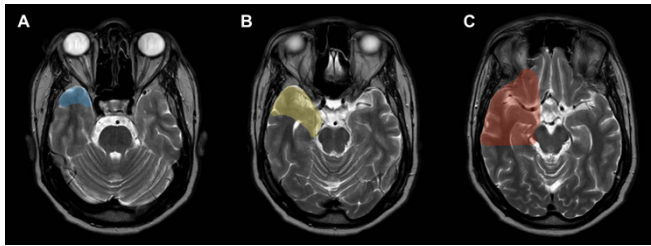
Figure 3- Galassi classification. A, Type I cysts, limited to the temporal pole, with small dimensions, biconvex shape, and free communication with surrounding subarachnoid spaces. B, Type II cysts (most frequent). Extension to the Sylvian fissure, rectangular morphology, and variable communication with the CSF flow pathways. C, Type III cysts. Largest in size, occupying the entire middle fossa and reaching the whole length of the Sylvian fissure. Can occupy the anterior fossa, displace the frontal, parietal, and midline structures. CSF, cerebrospinal fluid.

The second most frequent location is the posterior fossa, subdivided into (a) supra vermian, (b) retrocerebellar, (c) cerebellopontine angle, and (d) retroclival.

Other locations are suprasellar, ventricular, or convexity cysts.[1,3,4,19,20,29–32] The practical importance of location classification lies in the different symptomatological spectra and treatment possibilities, which will be discussed later. (Figure 4)
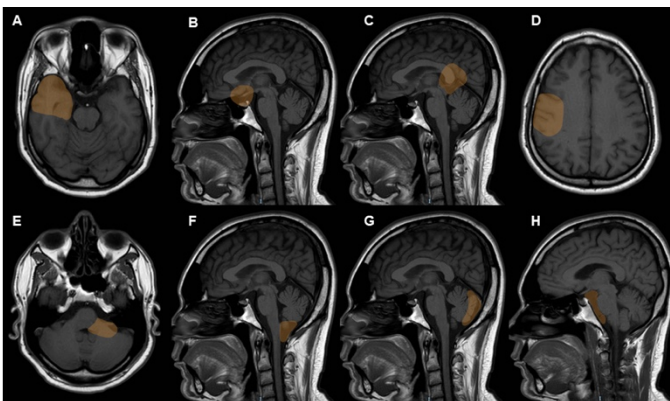
Figure 4- Main topographies of intracranial ACs. A, middle fossa. B, suprasellar. C, ventricular/paraventricular (quadrigeminal cistern). D, convexity. E, cerebellopontine angle. F, foramen magnum. G, retrocerebellar. H, retroclival. AC, arachnoid cyst

The ACs of the ventral midline (suprasellar and retro clival regions) cisterns have complex anatomy. They are divided into six types (three suprasellar and three retro clival) based on their anatomical origins.

Suprasellar cysts

André et al. [33] subclassified the suprasellar cysts according to their origin in relation to the arachnoid formations of the interpeduncular cistern.

The first category (Suprasellar arachnoid cysts (SAC) type 1 or SAC-1) comprises cysts of the chiasmatic cistern or diencephalic part of the Liliequist membrane. They are usually associated with hydrocephalus and signs of

intracranial hypertension and typically have a good prognosis when hydrocephalus is treated.

Cysts classified as SAC-2 result from enlargement of the interpeduncular cistern by defects associated with the mesencephalic part of the Liliequist membrane, with an elevation of its diencephalic part but without direct effect on the third ventricle. Usually, they do not require treatment.

Finally, SAC-3 cysts are an asymmetric enlargement of the interpeduncular cistern, involving other arachnoid membranes and cisterns, usually with lateral growth. Macrocrania and hydrocephalus are more common in these cases.
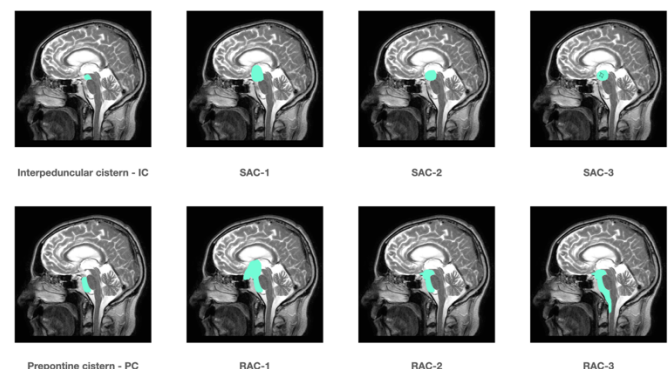
Retroclival cysts

Sarica et al.[34] subclassified the cysts from the arachnoid formations of the pre-pontine and pre-medullary cisterns.

Retroclival arachnoid cyst (RAC) type 1 (RAC-1), or suprasellar type, originates in the pre-pontine or pre-medullary cistern and have an upper extension beyond the diencephalic part of the Liliequist membrane, usually causing hydrocephalus.

RAC-2, or retroclival type, does not go beyond the diencephalic leaf of the Liliequist membrane and usually presents with cranial nerve deficits.

RAC-3, or retroclival type with spinal extension, are RAC-2 cysts with caudal growth to the pre-pontine cistern and whose symptoms are also related to the cranial nerves involved. (Figure 5)

Figure 5- Suprasellar and retroclival arachnoid cysts subclassification. SAC-1, cysts of the chiasmatic cistern or diencephalic part of the Liliequist membrane. SAC-2, enlargement of the interpeduncular cistern by defects associated with the mesencephalic part of the Liliequist membrane, with an elevation of its diencephalic part, but without direct effect on the third ventricle. SAC-3, asymmetric enlargement of the interpeduncular cistern, involving other arachnoid membranes and cisterns, usually with lateral growth. RAC-1, originate in the pre-pontine or pre-medullary cistern and with an upper extension beyond the diencephalic part of the Liliequist

membrane. RAC-2, does not go beyond the diencephalic leaf of the Liliequist membrane. RAC-3, caudal growth to the pre-pontine cistern. SAC, suprasellar archnoid cysts. RAC, retroclival arachnoid cysts.

The use of cisternography and magnetic resonance flow studies also classify arachnoid cysts as communicating (CIAC) if high contrast enhancement and CSF flow are evidenced and noncommunicating (NCIAC) if otherwise occurs.[35] The filling time by contrast, greater than or less than 24 h, also allows classifying them as cysts of early or late filling, which may influence the modality or indication of treatment.[19]

CLINICAL PRESENTATION

Widespread use of MRI and CT increased the number of arachnoid cysts diagnosed incidentally while investigating symptoms unrelated to them.[1,3,4,19,20,29–31] The evolution of these cysts can be highly variable, and cases have been documented in which an increase, decrease, or even disappearance of lesions during follow-up has been observed.[3,29,36] Al-Holou et al.[3] showed in their systematic review that most cases that underwent changes in their dimensions, a manifestation of symptoms, or need for surgical intervention were in patients younger than four years of age. Most cases have benign evolution, with no need for intervention.[29] The clinical characteristics of symptomatic arachnoid cysts will depend on the patient's age, cyst location, volume, and complications caused by another congenital abnormality. [4]

The most reported symptoms in the series were headache, nausea/vomiting, macrocrania, or other symptoms related to a progressive increase in intracranial pressure.[6,30,32,37] However, it is often difficult to correlate some nonspecific signs and symptoms with the finding of the arachnoid cyst on imaging.[3,37]

CLINICAL MANIFESTATIONS ACCORDING TO LOCATION

Middle fossa and sylvian fissure

The middle fossa or lateral fissure is the most frequent location of both incidental and symptomatic arachnoid cysts.[1,3,4,20,29–32,37] Nonspecific general symptoms, such as headache, nausea/vomiting, and visual changes, are often found in these cases. They may be associated with increased intracranial pressure caused by the expansive effect of the cyst.[30] Proptosis, contralateral paresis, seizures, developmental disorders, language disorders, delays in neuropsychomotor development, and attention deficits have been described in children with arachnoid cysts of the middle fossa,[3,20,31,38,39] with some studies reporting improvement in cognitive function after neurosurgical treatment.[38,40,41] On the other hand, other authors have also demonstrated no correlation between arachnoid cysts of the middle fossa, even higher volumes, and neurocognitive impairment in children.[31,42,43]

Posterior fossa

Second most frequent location, posterior fossa arachnoid cysts are more commonly associated with macrocrania and signs of increased intracranial pressure due to their mechanical role in the alteration of the CSF flow.[20,37]

Other authors also reported ataxia,[20,44] facial paresis,[45] hearing loss,[46] tinnitus, and vertigo.[47] Sarica et al.,[34] in their review on arachnoid cysts originated in the prepontine cistern, demonstrated higher frequency of headache in cysts with suprasellar extension (above the Liliequist membrane) and compression of the third ventricle, as well as higher frequency of cranial nerve palsies (VI, VII, and VIII) for cysts restricted to the prepontine cistern or with extension to the pre-medullary cistern.[34]

Despite reports demonstrating symptoms associated with cyst localization, a series of 23 patients with posterior fossa arachnoid cysts did not demonstrate expected symptoms for this location, such as cerebellar syndromes, vestibular or facial symptoms.[37]

Sellar Region

Sellar and suprasellar arachnoid cysts are common in children. When they are not incidental, they present with hydrocephalus as primary clinical manifestation, especially in cases of cranial extension of the cyst to the diencephalon, third ventricle, and obstruction of the interventricular foramens.[20,30,34,47] In his series of 34 cases, Ozek et al.[48] reported a high frequency of macrocrania and hydrocephalus, with about 26% of cases with visual or endocrine deficits and 18% with developmental delay. In a systematic analysis of 247 cases, Ma et al.[23] reported signs and symptoms of intracranial hypertension and developmental delay as more frequent preoperative symptoms in children under 4 years old. Endocrine and visual disorders are more relevant in the older group. The explanation for the reported signs and symptoms would be direct compression of the cyst on structures such as the pituitary stalk and optic chiasm. Bobble-head doll syndrome has also been reported at a lower frequency. It would be related to abnormal pressure on the wall of the third ventricle and dorsomedial thalamic nucleus. [48–50]

Ventricular cysts

The ventricular cavity is a relatively rare site for arachnoid cysts,[3] which usually originate from paraventricular arachnoid cisterns, such as the cistern of the velum interpositum and the quadrigeminal cistern.[51] Headache, vomiting, and macrocrania are the most reported symptoms from intraventricular cyst series, with reports of seizures, behavioral changes, and delay in neuropsychomotor development.[51–54] (Table 2)

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Table 2- Clinical manifestations according to location

Middle fossa	Posterior fossa
Headache	Macrocrania
Nausea/vomiting	ICH signs
Visual impairment	Hydrocephalus
Epilepsy	Ataxia
Language disorder	Facial palsy
Developmental delay	Vertigo/tinnitus
Sellar region	**Ventricular cysts**
Hydrocephalus	Headache
Macrocrania	Nausea/vomiting
Visual deficits	Macrocrania
Endocrine disorders	Hydrocephalus
Development delay	Seizures
Bobble-head doll syndrome	Development delays

Abbreviations: ICH, intracranial hypertension.

SPECIFIC CONDITIONS

Arachnoid cysts and headache

The association between headache and a cerebral arachnoid cyst is controversial among neurosurgeons and neurologists. Is there a typical headache pattern associated with cysts? Should the initial treatment be clinical or surgical?

In an interesting study, Eidlitz-Markus et al.[55] performed a retrospective analysis of the medical records of children who were taken to a tertiary center for headaches. Two hundred fifty patients presented signs of intracranial hypertension or change in headache pattern and were submitted to a brain MRI. Eleven patients (4.4%) showed an arachnoid cyst. Two children presented with headaches and signs of intracranial hypertension by cyst rupture. The other had the following clinical presentation: four with migraine pattern, two with tension headache, one with trigeminal neuralgia, one with recent-onset headache, and one with a mixed pattern of tension headache and migraine.

Edvardsson et al.[56] reported a cluster headache in combination with an arachnoid cyst with signs of mass effect. Although the clinical picture responded to triptans, the surgery was indicated with complete remission of the headache after surgery. The characteristics of headaches can vary, depending on the cyst's location.

Cherian et al.[57] argue that clinical treatment should always be tried primarily and the case followed strictly, and the surgical approach is indicated in patients that develop symptoms of rupture, increased intracranial pressure, or neurological deficits.

The risk inherent to the surgical procedure, the absence of proof of benefit after surgery, and the possibility of controlling symptoms with drug treatment lead to a consensus in the literature favoring the conservative management of cerebral arachnoid cysts without symptoms other than headache. A familial history of migraine can help in deciding on conservative approach.

Subdural hemorrhage associated with arachnoid cysts

Subdural hemorrhages are uncommon complications in arachnoid cysts of the middle fossa and rare in other sites.[20,30] Parsh et al.[58] found a 5-fold higher prevalence of arachnoid cysts of the middle fossa in patients with chronic subdural hemorrhage or hygroma. Some authors report a higher risk of bleeding in cysts of the middle fossa as a direct relationship with the anatomy of this region. The abundance of vessels in the lateral fissure and temporal pole, in addition to the weak mechanical protection offered by the low thickness of the squamous temporal bone, are related anatomical factors. Other authors suggest that this higher relationship would only be linked to the higher frequency of cysts in the middle fossa, reaching 60% of cases in some series.[59,60]

Traumatic brain injury is a well-accepted risk factor for subdural hemorrhage in patients with arachnoid cysts.[60,61] In addition to the recent history of trauma, Cress et al.[12] demonstrated that cysts larger than 5 cm are associated with a greater risk of bleeding or rupture.

Despite this higher prevalence, the risk of bleeding in patients with arachnoid cysts remains below 0.1%, and surgical treatment of arachnoid cysts may, by itself, lead to the formation of subdural collections and other morbidities, which leads us to the offer a conservative treatment in asymptomatic or oligosymptomatic cases.[3,58]

Arachnoid cyst and the practice of sports

Sport, whatever modality, is essential for intellectual development, stimulating rapid reasoning, competitiveness, preventing depression and anxiety, and acting in physical conditioning with improvement of musculoskeletal system and cardiorespiratory capacity. Despite all the benefits, it would be advisable to contraindicate sports activities, mainly contact activities, in patients with an arachnoid cyst? Physical stress can lead to cyst rupture? These are frequent questions in office environment made by parents of asymptomatic children with arachnoid cysts. Analyzing data in the literature that could answer these questions, we face some substantial limitations such as articles with a small number of investigated patients, papers only with case reports, absence of control groups or uniformity in the degree of symptoms severity, lack of prospective studies, publication bias by underestimated population.

A systematic literature review study shows a great diversity of management among specialists concerning sports activities since traumatic complications in AC patients are rare.[62] Although there are no guidelines for sport activities, children with AC may have an augmented risk of complications after traumatism. However, the physical and psychosocial benefits outweigh such risks.

In contrast, some series points out cysts are risk factors for developing hematoma after trauma. The health professional must make an individual assessment of each case, together with the child's family.[63,64]

Kertmen et al.[65] point out as main hypotheses for rupture of cysts and development of hematomas: (a) an increase in flow within the cyst in response to trauma leads to rupturing bridging veins and vessels in the cyst's wall; (b) lesser compliance of the cyst in relation to the brain during trauma, generating rupture of bridging veins.

In conclusion, prospective studies need to be carried out to define guidelines and better understand risks and benefits. AC carriers who practice sports with a high degree of physical contact, like martial arts, football, and boxing, should be investigated with imaging at the slightest symptoms such as headache. Activities with significant pressure differences in patients with arachnoid cysts, such as diving and parachute jumping, may increase the risk of rupture and hemorrhage in the AC.

CONCLUSIONS

Although widely studied for almost two centuries, ACs management is a subject of discussion until today. The specific patient characteristics, different morphologies, locations, and behavior of intracranial arachnoid cysts are confusing factors. To date, there is no consensus on its optimal management. The benefit of conservative or surgical treatment should be individualized in each case. This review sought to present, in a didactic and straightforward way, the knowledge already established and more recent updates, clarifying key points to assist the neurosurgeon in the therapeutic decision.

DISCLOSURES

Ethical approval

This study was performed in line with the principles of the Declaration of Helsinki. Approval was granted by the local Ethics Committee

Conflict of interest

The authors declare no conflicts of interest with respect to the content, authorship, and/or publication of this article.

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