REVIEW ARTICLE

Middle fossa arachnoid cyst: a review

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Arachnoid cysts are congenital cerebral spinal fluid (CSF) collections that most of times are incidentally diagnosed, but can cause symptoms of elevated intracranial pressure. In most cases, middle fossa arachnoid cysts (MFAC) have a benign natural history and remain asymptomatic, requiring only conservative treatment and surveillance. On the other hand, those patients who present or become clearly symptomatic, surgical intervention is mandatory. There are mainly two options of surgical treatment: cystoperitoneal shunt or cyst fenestration either microsurgery or endoscopic. This article aims to review the clinical presentation of MFAC, discuss the management of symptomatic and asymptomatic patients and review current surgical techniques.

Keyworks: Arachnoid cyst; Middle fossa arachnoid cyst; Neuroendoscopy; Intracranial hypertension

INTRODUCTION

Arachnoid cysts are congenital collections of cerebral spinal fluid (CSF) surrounded by arachnoid membranes, probably formed as a result of separation or duplication of those membranes during the embryonic period [1]. These cysts can arise elsewhere in the central nervous system, mostly in the arachnoid cisterns, being the middle fossa the most common location, accounting for 48-66% of all arachnoid cysts[2].

Most of the middle fossa arachnoid cysts (MFAC) are asymptomatic and incidentally diagnosed during other investigations, just a minority of them will expand and compress surrounding structures, becoming symptomatic[1]. The cyst can enlarge due to several mechanisms such as CSF secretion from the cyst wall through an ATPase enzyme, a ball-valve mechanism from the duplicated layer that entrap CSF and via an osmotic gradient caused by a higher protein concentration inside the cyst [1,3,4].

As stated before, most patients harboring MFAC will be asymptomatic and just few of them will suffer cyst expansion causing mass effect and consequent symptoms due to raised intracranial pressure, focal neurological deficits or obstructive hydrocephalus. Besides, cysts can rupture following minor trauma, causing subdural hygroma/hematoma. For those, surgical treatment is well accepted with good outcomes [1,4,5]. However, the symptomatic ones are the exception and most of pediatric neurosurgeons will face asymptomatic patients with large MFAC and apprehensive parents.

The optimal management of MFAC is intimately related to a solid knowledge about the natural history, clinical presentation, classification, and treatment options available. Therefore, the aim of this paper is to review these topics and discuss the optimal management of symptomatic and asymptomatic patients.

Natural history, clinical presentation and classification

Arachnoid cysts constitute about 1.4% of all intracranial space-occupying lesions, but only 5.3% are symptomatic. Cysts located at the suprasellar, quadrigeminal, cerebellopontine angle and ambient cisterns are more prone to be symptomatic, once they have the potential to obstruct the CSF pathway and cause hydrocephalus. On the other hand, MFAC are the majority of the asymptomatic ones, representing 47% of those[1].

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The decision-making process regarding treatment for a symptomatic cyst is most of times straightforward, mainly when the hydrocephalus is the cause of symptoms, being a surgical procedure advocated. On the other hand, the management of asymptomatic or even oligo-symptomatic patient harboring arachnoid cysts can be quite challenging, because the symptoms can be clinically vague and not related to the cyst, and the risks of surgery are not negligible. The pediatric neurosurgeon dealing with this situation must consider some important questions, such as “How often the cysts enlarge?”, “Is there a cut off age for enlargement risk?”, “What are the risks for cognitive development?”, "Is it a lethal disease?”, “What are the risks of a subdural hematoma?”, "Is there any relationship between arachnoid cyst and the seizure foci?”. We intend to answer all these questions based on the literature available about MFAC.

Currently, greater use neuroimaging methods had led to an increase in the number of incidentally diagnosed arachnoid cyst[1]. It is a common finding of an investigation of head trauma, seizures, developmental delay, or even headaches. The first concept to be kept in mind is that MFAC does not appear to be a lethal disease, even when related to a chronic subdural hematoma (CSDH). Nonetheless, we should consider the lack of evidence in literature as limitation to the best knowledge. Cress et al.[6] found no major neurological sequelae or mortalities in patients with arachnoid cysts and intracystic hemorrhage, adjacent subdural hygroma or subdural hematoma. In this study, larger arachnoid cyst (> 5cm) and recent head trauma were considered risk factors for arachnoid cyst rupture/hemorrhages[6].Wu et al. [7]performed a systematic review about CSDH and arachnoid cysts and showed a risk of 2.3% of developing CSDH with most patients (68%) presenting history of recent head trauma or sport-related injury. Almost 90% of patients of this review had MFAC and the rate of success treating the CSDH was 98.8%. Therefore, we can extrapolate these results and conclude that there is no clear benefit in prophylactic treatment of MFAC to avoid a CSDH, once it is a rare complication with good outcomes, and no formal sports restrictions are recommended for asymptomatic patients. Sports with great risk for head injury as boxing, martial arts, hockey, and football must be discussed individually with the patient and parents.

Another important situation to deal with is the presence of an asymptomatic patient with a large MFAC, and its risks of enlargement and cognitive decline. Samuel Hall et al.[1] studied the natural history of arachnoid cysts of 116 patients and reported that72.4% were asymptomatic. In 85.7% of these patients, the cyst remained stable, while in 10% the lesion reduced and in only one patient the arachnoid cyst grew. None of the asymptomatic patients have developed new symptoms during the study. Furthermore, their literature review showed cyst enlargement ranging from 2.5% to 10% and becoming symptomatic from 0.5% to 2.7%.Lee et al. [8] studied the growth of asymptomatic arachnoid cysts in the pediatric population with five years old or less (n = 86). The only factor related to the risks of growth was the age less than six months. In their study, none patient older than three years old showed enlargement in cyst size. Kim at al. [9] studied the neurocognitive profile of patients with arachnoid cysts before and after surgery and concluded that there is no significant improvement after the treatment. Schertz et al. [10] also studied the neurodevelopment in patients with large arachnoid cysts (included the Galassi II and III types) and did not find any significant difference between the surgical and the control group neither. These literature data allow us to conclude that asymptomatic arachnoid cysts have a low rate of cyst enlargement, being extremely rare for patients older than five years. Similar, the cyst volume and its treatment appear not to be related to any cognitive improvement, and reasonably not a formal indication for surgery.

Epilepsy image investigation can commonly find incidental arachnoid cysts, once temporal lobe epilepsy is one of the most common partial epilepsy syndromes and MFAC are the most common arachnoid cysts as well, they can coexist, but it is of paramount importance to understand if there is any relationship. Arroyo et al. [11]. retrospectively evaluated 867 patients with defined epilepsy syndrome and found 17 patients with arachnoid cysts, being 12 located in the middle fossa. However, only four patients (23.5%) had the focus adjacent to the cyst, allowing the conclusion that arachnoid cysts are often incidental findings. On this hand, Del Brutto et al. [12] compared two groups: patients with incidental arachnoid cysts and patients without any arachnoid cysts, founding an incidence of 4% of seizure/epilepsy in those harboring arachnoid cysts and 2.7% in those without arachnoid cysts. They conclude that there is a lack of association between arachnoid cysts and seizure/epilepsy. The coexistence of epilepsy and arachnoid cyst requires a diligent epileptic focus investigation by a multidisciplinary team before any indication of surgery.

Symptomatic cysts predominate in the pediatric population and usually the symptoms occur due to raised intracranial pressure (headaches, papilledema, nausea/vomiting, increased head circumference) or from mass effect (focal neurological deficits) [1]. MFAC do not commonly cause hydrocephalus and the main symptom reported is headache, becoming quite challenging to define the real relationship between cyst and the symptom in some cases [5]. In such situation, a very close follow-up with a pediatric neurologist and MRI looking for indirect signs of raised intracranial pressure (empty sella, optic sheet enlargement, Chiari I) is of paramount importance to not perform an unnecessary procedure. A team consensus will be necessary for the management success. As stated before, subdural hematoma is a rare presentation of MFAC and can

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be successfully managed surgically or even conservatively. The treatment of CSDH and arachnoid cyst is beyond the objectives of this paper and will not be discussed here.

The classic tomographic classification of MFAC, described byGalassi et al. [13], is still the main one in use nowadays. It divides the MFAC in three types: I, small, restricted to the anterior middle fossa, below the sphenoid ridge; II, superior extension along the Sylvian fissure and displacement of the temporal lobe; III, fills the whole middle cranial fossa and presents frontal and parietal lobes displacement (Figure 1). Type III lesions do not show communication with basal or sylvian cisterns, as demonstrated through cisternography [14]. Johnson et al. [4] proposed a modification in this classification, adding the type IV, when there is skull budging.

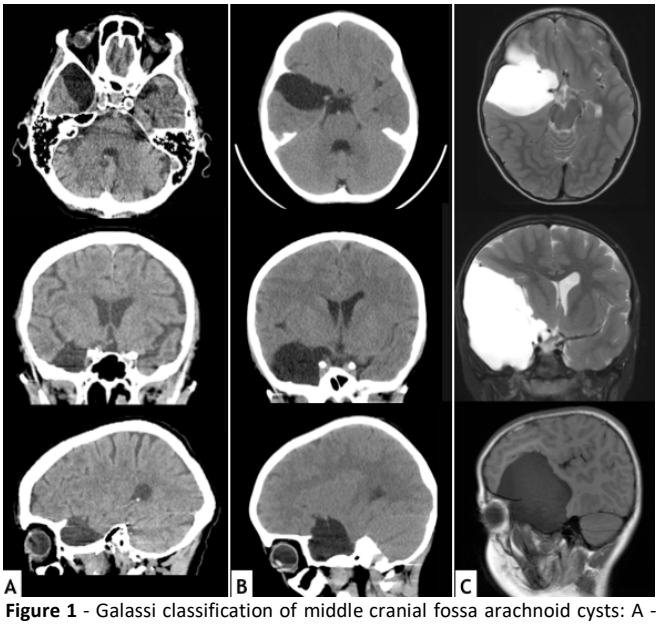

Figure 1 - Galassi classification of middle cranial fossa arachnoid cysts: A - Small cyst restricted to the anterior middle fossa and below the sphenoid ridge, classified as Galassi type I (Case courtesy of Dr. Bruno Di Muzio, Radiopaedia.org, rID: 31399); B - Cyst showing displacement of the temporal lobe and its superior extent along the Sylvian fissure, classified as Galassi type II; C - Large cyst fillingthe whole middle cranial fossa and displacing the frontal and parietal lobes, with midline shift. Classified as Galassi type III.

Treatment

Arachnoid cyst is a benign disease, in most of the cases asymptomatic, with low risk of complications and expansion after five years of life [8]. Therefore, the current tendency is to be conservative in asymptomatic patients [3]. Lesions Galassi type I do not show mass effect and do not need any intervention; lesions Galassi type II and III must be evaluated according to clinical presentation to offer the best therapeutic option [15].

Patients with unequivocal signs of elevated intracranial pressure, as hydrocephalus or papilledema, have clearly surgical indication. Those with symptoms attributed to the cyst, as macrocephalus or skull budging, are also eligible for surgical procedure. As stated before, patients presenting headaches, epilepsy/seizure, cognitive impairment need a very meticulous multidisciplinary evaluation before any surgical proposal [8–10,16].

The surgical options to treat arachnoid cysts are: cystoperitoneal shunt, microsurgical cyst fenestration, and endoscopic cyst fenestration [2,17].

Cystoperitoneal shunt presents good rates of cyst decompression in the literature, as Gangemi et al. [18] demonstrated remission in 96.8% of the cases with this technique. On the other hand, Chen et al. [19] performed a systematic review and meta-analysis about treatment of MFAC and showed a shunt malfunction around 40% and shunt dependency around 42% . It is already known that these shunting issues cause lifelong concerns to patients and their families, therefore, this procedure is rarely used as a first treatment currently [2].

Okano et al.[2] studied the outcome of 28 patients underwent craniotomy and fenestration of the cyst by microsurgical approach. They reported that 90.5% of patients related some symptom improvement after surgery, and the size of the cyst reduced in all cases (100%). Postoperative exams showed subdural effusion in 82.1% of the patients, all of them were asymptomatic and only one needed subdural peritoneal shunt . Chen et al. [19] reported rates of symptoms and cost reductions of 87%, total complications of 49%, short term complications 44% (mostly subdural effusions) and long-term complications of 3%. The microsurgical technique has the advantages of being a bi-manual technique with wrist movements allowing to pull and cut the arachnoid membranes, and to work in a dry and clean field. The clear disadvantage is the high rate of subdural effusions, besides that very few patients will present symptoms or even will need some treatment (Figure 2).

Neuroendoscopy became very popular and is the gold standard to treat some arachnoid cysts (intraventricular, suprasellar). But, considering MFAC, neuroendoscopy can be quite challenging and expertise and mastery are needed to perform it. Oertel et al. [20], in a retrospective analysis of 25 years, showed that endoscopy was feasible in 89 of 95 surgeries. 86.3% of the patients improved their symptoms and 65.3% presented radiological benefit. In only 4% of the cases, postoperative shunt was needed. However, when they analyze exclusively the MFACs patients, they found that pure endoscopy had to be abandoned in four cases (12.9%), because of bleeding and a lack of anatomical landmarks due to opaque membranes. Several complications were reported, as oculomotor and trochlear nerve palsies, vasospasm with hemiparesis, permanent diabetes insipidus. They abandoned the pure endoscopic technique for MFAC and changed to a microsurgical procedure. Di Rocco et al. [5] also reported the limits of endoscopic treatment, being anatomy the main limitation and bleeding another important cause to change the procedure. On the other

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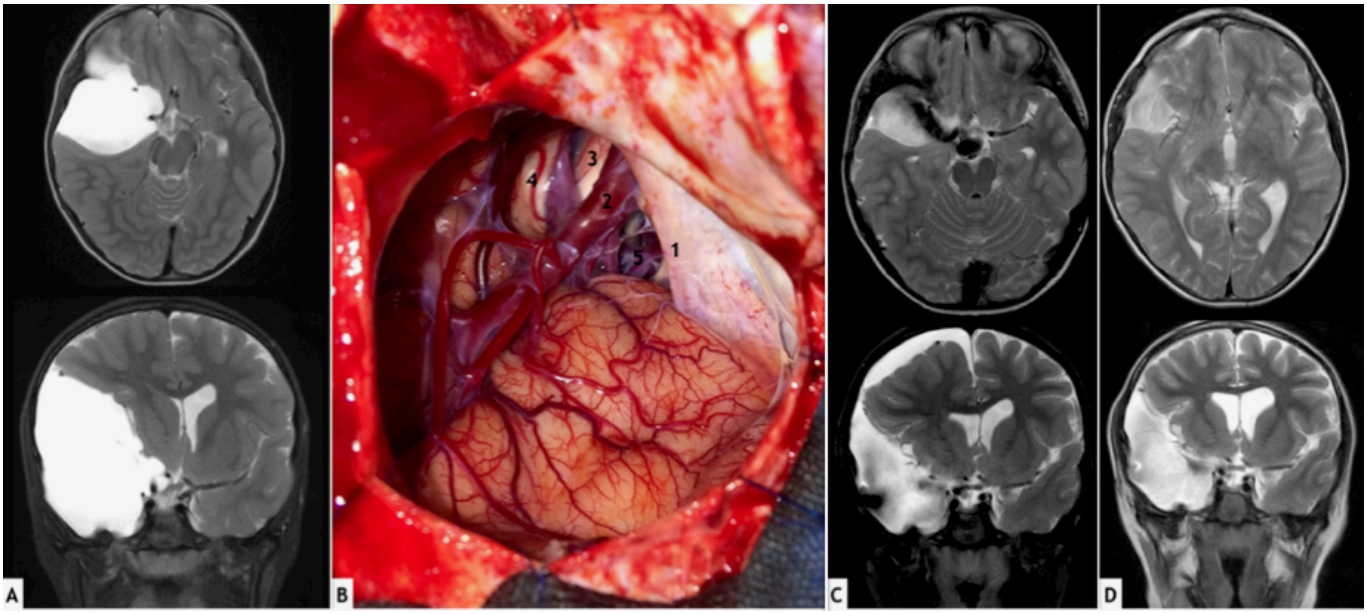

Figure 2 - Pre operative MRI image and surgical picture: A - Axial and coronal views of a large Galassi type III cyst; B - Surgical picture after right middle fossa arachnoid cyst fenestration through craniotomy and microsurgery (1 - Tentorium; 2 - Internal carotid artery; 3 - Optic nerve; 4 - Olfatory nerve; 5 - Large opening of the Liliquist membrane exposing the infra tentorial contents as both oculomotor nerves and basilar artery); C - One month post operative MRI showing good communication of the cyst with the basal cisterns, and subdural effusion on the coronal view; D - Six month post operative MRI showing **spontaneous resolution of the subdural effusion.**

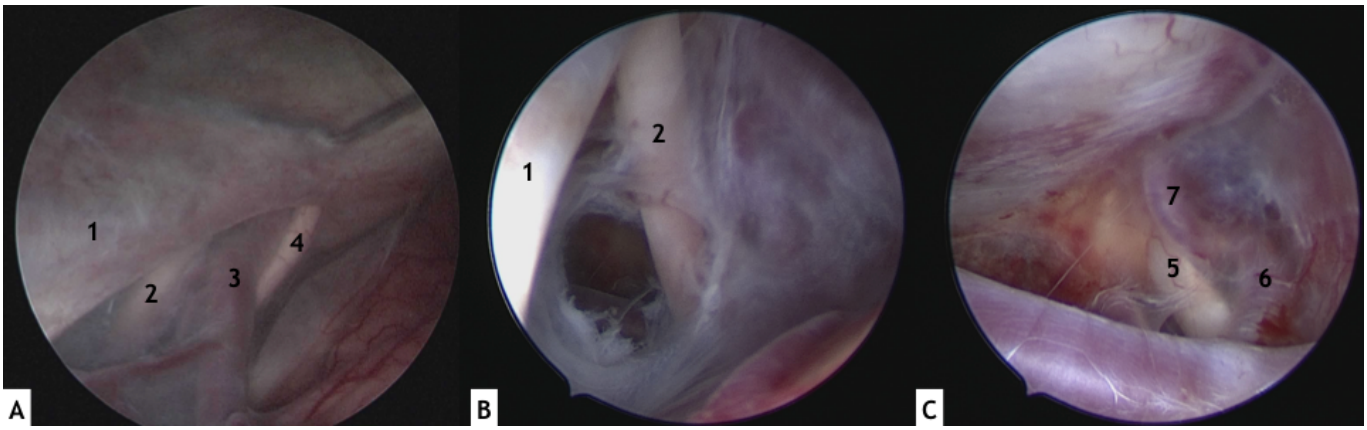

Figure 3 - Intra operative pictures of an endoscopic approach to treat a left middle fossa arachnoid cyst: A - Initial endoscopic view of the cyst and the surgical anatomic landmarks covered by arachnoid membrane; B - Opening of the Liliquist membrane between the left oculomotor nerve and tentorium; C - The endoscope passed through the fenestration showing the infra tentorial content, showing the right oculomotor nerve, right posterior cerebral and posterior communicating arteries. (1 - Tentorium; 2 - Left oculomotor nerve; 3 - Internal carotid artery; 4 - Optic nerve; 5 - Right oculomotor nerve; 6 - right posterior cerebral artery; 7 - Posterior communicating artery)

hand, Chen et al. [18] results showed that the endoscopic technique has a high rate of efficacy (90%) and the lowest rate of short-term complications (23%), and they believe that endoscopic technique may be the first choice to treat MFAC with other techniques reserved for failures (Figure 3).

CONCLUSION

MFAC presents a very benign natural history and solid concepts about cyst enlargement, symptoms, risks, and treatment options are fundamental for the best management. Asymptomatic patients must be closely followed, and no surgery offered. When symptoms are vague/not clear (headaches, epilepsy/seizure, cognitive impairment) a very diligent multidisciplinary evaluation is mandatory.

Surgery can be offered to patients that are clearly symptomatic, especially with signs of raised intracranial pressure and mass effect. Cyst fenestration seems to be the best option for surgical treatment, being the shunt an alternative if the fenestration fails. Microsurgical or endoscopic fenestration have similar rates of success and complications, and the neurosurgeon's expertise must be considered to choose one over the other. In both techniques, subdural effusion is the most common complication, being asymptomatic in most of cases.

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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

Consent to participate

The patient gave consent to use his information and images for research proposes.

Consent for publication

The patient gave consent to use his information and images for publication.

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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