Review Article

Arachnoid cysts of the quadrigeminal cistern: Proposal of a therapeutic algorithm based on a systematic review of literature

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Introduction: Although quadrigeminal arachnoid cysts (QACs) are infrequent malformations of the arachnoid space, which affect the quadrigeminal cistern, they have important clinical consequences, such as hydrocephalus and compressive symptoms. Moreover, there is much controversy on its surgical management; thus, it is important to determine the current state of the art regarding the best treatment modality.

Objective: This study aimed to examine the effectiveness of different surgical approaches and revisit the anatomy of the quadrigeminal cistern.

Methods: A systematic review of the literature was conducted considering studies published in the last 20 years regarding the natural course and treatment of QACs. Only English papers were selected, and case reports were excluded from the analysis.

Results: We identified 286 articles, of which nine were selected for the qualitative analysis. Clinical presentations of QACs included mental disorientation, bladder incontinence, dysphagia, quadriplegia, ataxia, and nocturnal headaches, which were dependent on the cyst size, and a spontaneous resolution was not described. A microsurgical technique via a supracerebellar infratentorial approach and fenestration to the third ventricle and ventricular endoscopic third ventriculostomy were the main techniques, which were performed depending on the anatomical features and presence of hydrocephalus. Shunt placement was associated with high rates of complications and recurrence.

Conclusion: Despite the lack of solid evidence on the treatment of QACs, mainly due to their rarity, an endoscopic approach with ventricle cystostomy has been indicated as the more effective surgical technique.

Keywords: Quadrigeminal arachnoid cysts; Arachnoid cysts; Neuroendoscopy; Quadrigeminal cistern

INTRODUCTION

Arachnoid cysts are uncommon malformations of the subarachnoid space, in which the cerebrospinal fluid (CSF) is arrested into the splitting arachnoid membrane, forming a cyst that could become enlarged, isolated from the rest of the subarachnoid space and ventricles, and compressed the surrounding structures [1,2]. Additionally, other hypotheses have been put forward to explain its origin, such as the ectopic production of the CSF by the cyst wall, a slit-valve mechanism, and the osmotic gradient between the CSF and the cyst fluid [3].

Such malformations could virtually affect any part of the arachnoid space and account for 1% of all expansive intracranial injuries. Moreover, these cysts could most commonly affect the Sylvian cistern, followed by the cerebellopontine angle, and the quadrigeminal cistern is the third most common site, which is rarely affected, and accounts for 10% of all arachnoid cysts in a major case series [4] or ≤1% in other studies [5,6]. Quadrigeminal arachnoid cysts (QACs) are often associated with other congenital diseases such as holoprosencephaly, Chiari type II malformation, and encephaloceles, or are exceedingly rarely expressed in familiar diseases with multiple cysts [7,8].

There is much controversy regarding the best approach of QACs, and several techniques have been described for its treatment, such as the (1) microscopic approach with wall fenestration or excision, (2) shunting, and (3) endoscopic approach of the lateral ventricle with lateral ventriculocystostomy or third cystostomy or concurrently with endoscopic third ventriculostomy (ETV) [9-13].

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The primary aim of this study was to examine the current evidence regarding the most effective approach for QACs and revisit the anatomy of the quadrigeminal cistern. The secondary aim was to propose an evidence-based algorithm that guides the treatment approach for QACs.

MATERIALS AND METHODS

Following the PRISMA guidelines [14], to enhance the quality of the systematic review, a search considering the articles published in PubMed and Web of Science in the last 20 years regarding QACs was undertaken using the following constructs: "Quadrigeminal arachnoid cyst" AND "hydrocephalus," "Quadrigeminal arachnoid cyst" AND "natural history."

All case series with at least three cases were considered for the analysis, as well as prospective and retrospective studies. Studies on adults were also included. Case reports, commentaries, surgical technical notes, revision papers, studies in animals, and those written in non-English language were excluded.

A qualitative analysis was undertaken on selected studies regarding surgical techniques and their effectiveness. Complications were also compiled.

RESULTS

The search initially identified 286 articles, and after the application of the exclusion and inclusion criteria and elimination of duplicates, nine studies were considered for the analysis.

On the Web of Science, the search using [Quadrigeminal arachnoid cyst] found 123 articles, but no articles were found for [Quadrigeminal arachnoid cyst] AND [Hydrocephalus] or [Quadrigeminal arachnoid cyst] AND [natural history].

On PubMed, the search for [Quadrigeminal arachnoid cyst] resulted in 105 articles. The construct [Quadrigeminal arachnoid cysts] AND [Hydrocephalus] resulted in 57 papers, and [Quadrigeminal arachnoid cysts] AND [natural history] resulted in one article.

The flow chart of the selection process and studies analyzed are represented in Figure 1 and Table 1, respectively.

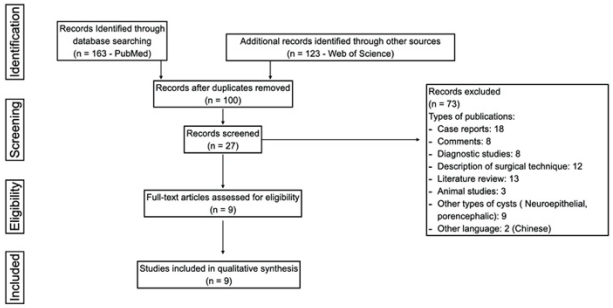

Figure 1- Flow chart of the selection of articles included in the study

Table 1 - Nine studies on the treatment of quadrigeminal arachnoid cysts

Author/year	Design	Sample	Treatment	Conclusions
Tamburrini et al., 2007[23]	Retrospective	11	Endoscopic fenestration	100% effectiveness by VC
Ersahin et al., 2008[19]	Retrospective	17	Endoscopic fenestration Shunt	Failure of 87.5% in <6 months and 100% success >6 months (p = 0.005)
Cinalli et al., 2010[12]	Retrospective	14	Endoscopic fenestration	90% successful in VC and enhanced by ETV
Sengul et al., 2012[24]	Retrospective	3	Endoscopic fenestration plus ETV	Effective in all cases
El-Ghandour, 2013[21]	Prospective	18	Endoscopic	Clinical improvement in 83.3% of the cases
Yu et al.,2015[4]	Retrospective	8	Endoscopic	Clinical improvement in 100% of the cases and permanent in 62.5% of cases with endoscopic fenestration. ETV did not play an important role
Gui et al., 2016[20]	Retrospective	28	Endoscopic	Clinical improvement and cyst reduction in 89.3% of cases

DISCUSSION

Anatomical relationship

In order to provide the foundations of microsurgical as well as endoscopic approach of quadrigeminal arachnoid cysts, a detailed description of the anatomy of quadrigeminal cistern based on the classical and recent works was offered. Conceptually, the quadrigeminal or colicular cistern occupies a strategic place in the brain and establishes an important relationship with neural and

vascular structures in the pineal region [4]. Moreover, this cistern is placed on the posterior incisural space, which is posterior to the upper portion of the brainstem surrounded by the tentorial edge [15]. It is composed of the posterior arachnoid complex, which is formed by the posterior peri mesencephalic membrane and the cerebellar precentral membrane [16]. Interestingly, Zhang et al.[16] pointed out the similarity between this arachnoid complex and the Liliequist membrane and noted the distribution of these two membranes in the posterior incisural space in an inverted shape in comparison with the Liliequist membrane, which is located in the anterior incisural space. Superiorly, the quadrigeminal cistern is limited by the splenium of the corpus callosum, vein of Galen, and falx tentorial junction. Anteriorly, it is limited by the quadrigeminal plate that is formed by the superior and inferior colliculi of the midbrain and the pineal gland. Inferiorly, it is limited by the culmen of the cerebellum and laterally by the pulvinar of the thalamus.

The superior cerebellar artery, which irrigates the tentorial surface of the cerebellum, is near this cistern [17]. Moreover, given their relation to the pulvinar of the thalamus and the atrial segment of the choroidal fissure, QACs frequently expand laterally toward the lateral ventricles through the choroidal fissure and are apparent on the lateral ventricle, thinning the ependyma and becoming available in the endoscopic fenestration [18] (Figure 2). These cysts could grow inferiorly and compress the vermis of the cerebellum or predominantly anteriorly to the tectal plate and could lead to aqueductal obstruction (Figures 2 and 3). Therefore, the surgical approach to this cistern should be performed carefully to avoid damaging great vessels such as the vein of Galen, internal cerebral veins, and Rosenthal veins, which converge to the straight sinus in the falx tentorial joint (Figure 2).

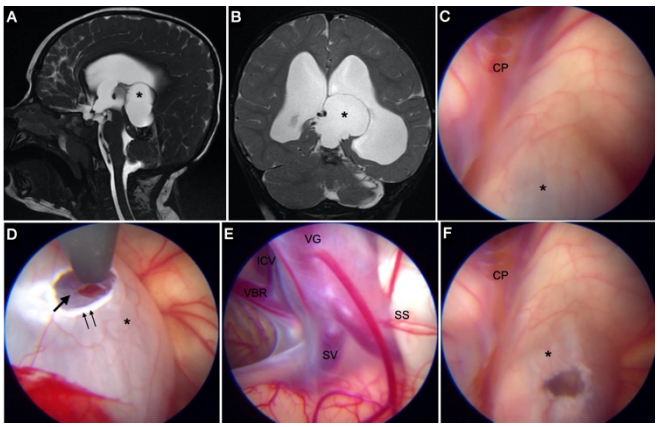

Figure 2- Quadrigeminal arachnoid cyst compressing the cerebellum (triple arrows) and the tectal plate of the midbrain (thick white arrow) with no hydrocephalus in an eight-year-old girl who presented with headache.

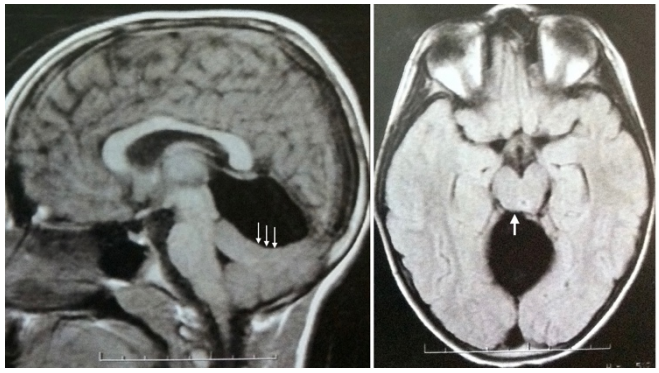

Figure 3- Quadrigeminal arachnoid cyst (*) in a boy aged 1 year and 8 months who underwent surgery via an endoscopic approach. The enhanced T2 sagittal magnetic resonance (MR) image showing the compressed cerebellum, tectal plate, and upper extension (A). Enhanced T2 coronal MR image showing the lateral extension to the left trigone (B). The endoscopic approach was achieved in a left frontal burr hole assessing the left lateral ventricle, and after ETV, a fenestration of the outer (double arrows) and inner (thick black arrow) walls of the cyst was undertaken, and the quadrigeminal cistern was observed in E and the resulting ostomy in F. CP, choroid plexus; VC, vein of Galen; ICV, internal cerebral vein; VBR, basal vein of Rosenthal; SV, superior vermian vein; SS, straight sinus

Rationale of treatment

With progression, QACs usually compress the tectal plate and infrequently generate focal symptoms, such as eye gaze abnormalities with diplopia, nystagmus and even Parinaud syndrome. Psychomotor retardation, hemiparesia, and symptoms due to hydrocephalus secondary to the obstruction of the aqueduct of Sylvius were also reported [19,20]. Essentially, hydrocephalus is the most common presentation observed on the majority of studies [20-23], and signs of intracranial hypertension such as headache, drowsiness, bulging fontanel, visual impairment, and macrocrania have been frequently reported [20]. Ideally, the goal of surgery is to restore the pathway of the CSF by performing fenestrations on the cyst wall that communicate to the normal subarachnoid space, ventricle, and other cisterns [19,24]. Alternatively, a diverting shunt could be inserted into the cyst to reduce the internal pressure and consequently the compression of the surroundings, even with an augmented risk of failure [21]. Even if a cystoperitoneal shunt has been considered an easy procedure, the rate of dysfunction could be as high as 40% which is linked to obstructions and migrations of the hardware. Additionally, there is the risk that the catheter may not perforate the cyst wall adequately; thus, this technique is avoided and the endoscopic approach or craniotomy is suggested [25]. Although some studies have considered patients who have undergone shunts, which were unavailable in other modalities, assuming shunt dependence, other studies have described the success of the endoscopic approach in cases of failed cystoperitoneal shunts [12,21].

There are some controversies regarding the surgical indication for large cysts in children aged <6 months with no

symptoms. Some authors advocated the endoscopic approach, expecting cyst reduction instead of close surveillance, and ulterior surgery in large cysts or symptomatic cases. However, the clinical presentation is preponderant to the cyst size, which is a criterion for surgery [20].

Another treatment option is suboccipital craniotomy via supracerebellar infratentorial approach to fenestrate QACs to the third ventricle and the subarachnoid space of the surrounding cistern. However, this treatment modality presents some risks, such as being more invasive, bleeding from damaged deep vessels, infections, and even recurrence [9,26].

Nevertheless, the ventricular endoscopic approach has been considered ideal for the management of fluid-filled cavities and will be most likely the technique to be indicated when QACs turned out to be associated with hydrocephalus [20,21].

Silva et al.[1] retrospectively analyzed 12 patients, with one adult, reported an incidence of 83% of hydrocephalus, and performed the ventricular endoscopic approach using the ETV concurrently in three cases. In addition, they advocated the avoidance of shunt whenever possible and have indicated it in selected cases. Nevertheless, they reported 33% of recurrence and pointed out younger age as the main risk factor. Indeed, such a high failure rate in children aged <6 months could be understandable in the light of new conceptions of minor CSF pathways according to Di Rocco and Shizuo Oi[27] who hypothesized the inexistence of conventional absorption by Pachionni bodies in young children with a predominance of other sources such as the gliolymphatic system, choroid plexus, and neurons, which could be less functional in some cases. Therefore, young children may have a mixed mechanism of hydrocephalus, and fenestration is not enough. Considering evidence of hydrocephalus management, endoscopic choroid plexus coagulation could be an option in young children, as it is expected to increase the success rate when combined ETV [28,29].

Several authors advocated performing ETV concurrently with ventriculocystostomy assuming that the fenestration of the cyst wall to the ventricle is not enough to warrant CSF restoration and aqueductal stenosis will remain even after cyst reduction. Therein, ETV could warrant CSF circulation [21]. However, no reliable predictable factors can indicate cyst fenestration alone in both procedures. To our knowledge, ETV is feasible in the majority of cases and should be performed with caution mainly in patients with myelomeningocele, which is a common anatomical distortion on the floor of the third ventricle [30]. In addition, Gui et al.[20] recommended performing ETV before cyst fenestration because bleeding from the cyst wall could prevent a safe ETV.

In another case series, El-Ghandour et al.[21] analyzed prospectively 18 children with symptomatic QACs who underwent surgery via a totally endoscopic approach and described five cases in which the patients have undergone a prior shunt into the cyst with no clinical improvement or reduction of cyst size. They have undertaken lateral ventriculocystostomy in 10 patients, third cystostomy in 4 patients, and ETV in 14 patients, and in four patients, the cysts additionally communicate to the cistern. Furthermore, they stated that the endoscopic approach is a less invasive, safe, and fast (mean of 43 min) procedure, with a few minor complications. However, the indication of ETV varies among studies, and in another case series, Cinalli et al. reported ETV in 6 of 14 patients who have undergone surgery via an endoscopic approach to treat QACs [12].

Similarly, Ersahin et al. [19] described their experience with 17 children with QACs, and the most common presentations were macrocrania and psychomotor retardation. They used the endoscopic approach in five patients who had received prior shunting. They reported one case of recurrence after 3 years, which was successfully treated by a new endoscopic approach. Interestingly, they pointed out lateral extension to the lateral ventricles in the majority of cases, and the endoscopic fenestration was facilitated through the lateral ventricle. Moreover, during the procedure, aqueductoplasty with stenting was performed on two patients.

To analyze the effectiveness of the endoscopic approach, Gui et al. [20] performed a retrospective analysis of a mixed population of 28 patients, which was predominantly composed of children (n = 25) who underwent primary endoscopic surgery for QACs, excluding those who had a history of surgery. They reported that the procedure was effective in 25 of 28 cases, and one endoscopic reoperation was performed 5 months later due to closed fenestration, and they inserted a cystoperitoneal shunt in two cases and found its effectiveness even in children aged <6 months.

Therefore, the endoscopic approach was considered the first technique of choice in the management QACs on the basis of the recurrence rate of patients who have received shunts for the first time. Technically, the degree of cyst fenestration depends on the direction of enlargement. In lateral enlargement, it is more feasible to assess via the trigone of the lateral ventricle, or in the case of midline expansion, the burr hole could be placed more anteriorly and the cyst fenestration could be performed on the third ventricle. An algorithm was proposed based on the best evidence available to handle QACs (Figure 4).

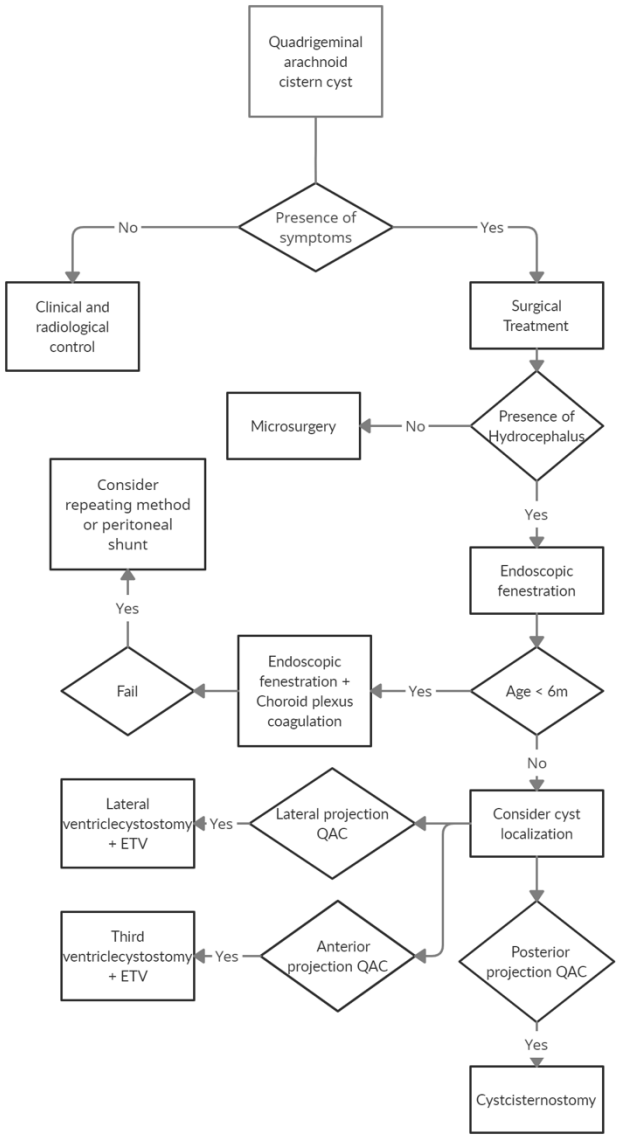

Figure 4- Management algorithm of quadrigeminal arachnoid cysts

Complications

Subdural hygroma was the most common complication reported, which varies from 16.7% of cases in a prospective series21 to 14.3% in a large retrospective study20.

Bleeding during endoscopic fenestration was described in one case in a prospective case series, in which it occurred on the deep wall of the cyst and was easily managed by irrigation. In addition, the authors did not remove the cyst wall in any situation21.

Overall, the endoscopic approach was indicated by the vast majority of studies as a safe and effective treatment of QACs and presented a low complication rate.

Limitations

Given the rarity of QACs, drawing any solid conclusions regarding its management is difficult, and studies are restricted to small case series and retrospective works. However, our evidence of the benefit of the endoscopic approach was facilitated by consolidated works on the field of hydrocephalus, and we found a similar relationship in failed cases in young patients and concerns with shunts.

CONCLUSIONS

Neurosurgeons should consider avoiding shunt placement on QACs, and in children aged <6 months, choroid plexus coagulation could be combined with ETV and ventriculocystostomy concurrently to increase the effectiveness.

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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