CASE REPORT

Approach and presentation of quadrigeminal cistern arachnoid cyst in pediatrics: a case series

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ABSTRACT

Quadrigeminal cisternal arachnoid cysts (QACC) represent a rare group of central nervous system lesions. The clinical presentation varies depending on the size of the cyst and the degree of compression. A retrospective review of QACC cases surgically treated at the Instituto Nacional de Salud del Niño San Borja between 2017 and 2024 was conducted. Sociodemographic and clinical data were collected from patient medical records. During the study period, 10 patients diagnosed with QACC underwent surgery. Five presented with type I and five with type III QACC. Neuroendoscopy with cyst fenestration was performed in 80 % of cases. Seventy percent of patients required only one surgery, and 50 % remained free of cerebrospinal fluid shunting. A reduction in cyst size was observed in nine patients, while complete resolution of the lesion occurred in one patient after two surgeries. This case series suggests that neuroendoscopy with cyst fenestration improves the clinical outcomes of QACC and results in cyst size reduction or resolution.

Keywords: Arachnoid Cyst; Hydrocephalus; Craniotomy; Neuroendoscopy (Source: MeSH)

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Abordaje y presentación del quiste aracnoideo de la cisterna cuadrigeminal en pediatría: una serie de casos

RESUMEN

Los quistes aracnoideos de la cisterna cuadrigeminal (QACC) representan un grupo poco frecuente de lesiones del sistema nervioso central. El cuadro clínico depende de la extensión del quiste y de la severidad de la compresión. Se realizó una revisión retrospectiva de los casos con diagnóstico de QACC intervenidos quirúrgicamente en el Instituto Nacional de Salud del Niño San Borja en el periodo 2017-2024. Se recopilaron datos sociodemográficos y clínicos de los pacientes a partir de las historias clínicas. Durante el periodo de estudio, 10 pacientes diagnosticados con QACC fueron intervenidos quirúrgicamente. Cinco pacientes presentaron QACC tipo I y cinco tipo III. El 80 % de los casos recibió una neuroendoscopia con fenestración del quiste. El 70 % de pacientes requirió solo una cirugía. El 50 % de los pacientes quedaron libres de cualquier forma de derivación del líquido cefalorraquídeo. En nueve pacientes, se observó disminución del tamaño del quiste; mientras que en el caso restante, la resolución completa de la lesión luego de dos cirugías. En esta serie de casos, la aplicación de la neuroendoscopia con fenestración del quiste demostró una mejoría en el cuadro clínico de los QACC, así como una disminución del tamaño o resolución del quiste.

Palabras clave: Quistes Aracnoideos; Hidrocefalia; Craneotomía; Neuroendoscopia (Fuente: DeCS)

INTRODUCTION

Arachnoid cysts are congenital compartments filled with cerebrospinal fluid (CSF) located within the cisterns and surrounded by the arachnoid membrane, very similar to normal arachnoid membranes. Their content is clear and colorless, resembling ventricular CSF. Arachnoid cysts can arise anywhere in the central nervous system and are closely related to arachnoid cisterns (1). Quadrigeminal cistern arachnoid cysts (QACC) are usually

asymptomatic and may be incidentally diagnosed during cranial computed tomography (CT) or magnetic resonance imaging (MRI) (2,3). They account for 5% of arachnoid cysts in children and 10% in adults (4–6). QACC may be associated with other central nervous system malformations, such as holoprosencephaly, Chiari malformation, and encephaloceles (5,7).

A classification has been proposed for QACC comprising three types. Type I extends both supratentorial (at the trigone level) and infratentorial (in the supra cerebellar cistern); type II involves the supra cerebellar or supra-retro cerebellar regions; and type III extends laterally toward the ambient cistern and temporal lobe (5,7). The clinical presentation results from compression of the brainstem, cerebellum, and tectal plate, obstructing the aqueduct of Sylvius and subsequent hydrocephalus, which in turn leads to headache, somnolence, bulging fontanelle, vomiting, visual disturbances, psychomotor delay, and macrocephaly (2,4,8). The neuroimaging study of choice is brain magnetic resonance imaging (MRI), which shows a well-defined, extra-axial cyst with signal intensity similar to CSF in all sequences (2,4). Various treatment options for QACC include craniotomy, cyst excision or fenestration, cystocisternostomy, ventriculocisternostomy, and cystoperitoneal shunt. The approach may be neuroendoscopic; however, there is no consensus on the treatment of choice for this type of cyst (1,4,9). In this study, we describe 10 cases of QACC treated at the Instituto Nacional de Salud del Niño San Borja (INSN-SB) between January 2017 and April 2024.

CLINICAL CASES

A retrospective review was conducted of institutional cases diagnosed with symptomatic QACC between January 2017 and April 2024. Medical records were reviewed, and sociodemographic data were collected, along with information on admission, clinical course, CT and MRI findings, postoperative outcomes, and follow-up after discharge.

During the study period, 10 patients diagnosed with QACC underwent surgical intervention at INSN-SB. The mean age at diagnosis was 26 months, with 80% of cases being male. The observed signs and symptoms included macrocephaly, irritability, vomiting, and seizures. Seven cases were diagnosed with hydrocephalus, of which five had type I QACC and two had type III QACC. The remaining three cases, not associated with hydrocephalus, presented with type III QACC.

Regarding treatment, 80% of patients underwent neuroendoscopy with cyst fenestration. Seventy percent required only one surgical procedure. Fifty percent of patients remained free from any form of CSF shunting. As for the type of cyst diagnosed, five patients had type I QACC, and five had type III QACC. No cases of type II were identified (Table 1).

Table 1. Sociodemographic and clinical characteristics at diagnosis and during follow-up of the ten QACC cases included in the study

N°	Sex	Symptoms	Hydrocephalus	Age at diagnosis (months)	Complications	First surgery	Second surgery	Third surgery	Fourth surgery	Outcome	% reduction	Type	Follow-up (months)
1	M	Headache, seizures	Yes	39	None	Neuroendoscopy with fenestration + ETV	VPS			Resolved	100	I	52
2	M	Macrocephaly	Yes	12	None	Neuroendoscopy with fenestration				Reduced	35.3	I	0.1
3	M	Macrocephaly, seizures	No	12	None	Cystoperitoneal shunt				Reduced	40.7	III	37
4	F	Macrocephaly, irritability	Yes	1	Right occipital intracerebral hemorrhage	Neuroendoscopy with fenestration + EVD				Reduced	53.4	III	0.7
5	F	Headache, vomiting	Yes	187	None	Neuroendoscopy with fenestration	EVD	EVD	VPS	Reduced	39.2	I	21
6	M	Macrocephaly	Yes	0.8	None	Neuroendoscopy with fenestration				Reduced	36.2	I	0.9
7	M	Macrocephaly	Yes	0.8	None	Craniotomy	VPS			Reduced	34.1	I	22
8	M	Macrocephaly, irritability, vomiting	No	6	None	Neuroendoscopy with fenestration + CPS				Reduced	28.4	III	0.6
9	M	Macrocephaly	Yes	0.5	None	Neuroendoscopy with fenestration + ETV				Reduced	85.6	III	1
10	M	Cranial deformity	No	1	None	Neuroendoscopy with fenestration				Reduced	74.8	III	0.7

M: male; F: female; VPS: ventriculoperitoneal shunt; EVD: external ventricular drain; CPS: cystoperitoneal shunt; ETV: endoscopic third ventriculostomy; QACC: quadrigeminal arachnoid cistern cyst.

The mean postoperative follow-up period was 13.59 months. One patient underwent craniotomy plus cyst fenestration and later developed hydrocephalus requiring a ventriculoperitoneal shunt. In nine patients, a reduction in cyst size was observed, while in one patient, complete resolution of the lesion was noted on CT after two surgeries. Only one case presented with postoperative occipital intracerebral hemorrhage, which resolved spontaneously, leaving a cerebral infarction area as a sequela.

DISCUSSION

QACCs are more frequent in males, with macrocephaly being the predominant sign. Due to their proximity to the brainstem and cerebellum, QACCs cause compression and narrowing of the aqueduct of Sylvius, leading to hydrocephalus and symptoms related to brainstem and cerebellar dysfunction (2,4,8,10). No single surgical treatment is considered the gold standard; both neuroendoscopy and microsurgery are widely reported in the literature (4,11). Consequently, QACCs are considered conditions that may require more than one surgical intervention (1). The surgical approach is individualized for each patient, utilizing either neuroendoscopy or microsurgery, with the primary objective of treating hydrocephalus, which is common in most cases. Hydrocephalus is managed by fenestrating the cyst via the neuroendoscopic technique, namely endoscopic fenestration and third ventriculostomy. These procedures can be performed concurrently during the same surgical session (1,6,10).

In our case series, neuroendoscopy with cyst fenestration emerged as the most commonly performed procedure, leading to clinical improvement and a reduction in cyst size or complete resolution. The choice of treatment was based on the clinical history, physical examination findings, and their correlation with radiological imaging. Neuroendoscopy using a rigid endoscope was considered the first-line approach, with an effort made to avoid the placement of a ventriculoperitoneal shunt. We advocate for neuroendoscopy as the preferred treatment option, as it avoids the need for permanent implants such as CSF shunts, which are often associated with a higher rate of complications and a poorer prognosis. This approach not only ensures patient comfort but also helps prevent complications that may arise from craniotomy. The use of this technique led to satisfactory clinical outcomes in the patients.

Neuroendoscopic cyst fenestration is a minimally invasive procedure that, at INSN-SB, is the first-line surgical approach for patients diagnosed with QACC. This procedure is associated with a low rate of complications, most of which are minor. Furthermore, it contributes to a better prognosis by eliminating the need for CSF shunting. Neuroendoscopic cyst fenestration can be combined with other hydrocephalus treatments, achieving satisfactory outcomes for the patient.

Author contributions

The author confirms responsibility for the conceptualization and design, data collection, analysis, interpretation, and preparation of the final manuscript.

Conflicts of interest

The authors declare no conflicts of interest related to the content of this manuscript.

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Ethical considerations

This study adhered to the fundamental ethical principles outlined in the Declaration of Helsinki, including non-maleficence and confidentiality. All collected data were strictly confidential and used solely for research purposes. Authorization for medical record review and informed consent were obtained prior to data collection. The study was approved by the Institutional Research Ethics Committee of the Instituto Nacional de Salud del Niño San Borja (CIEI INSNSB).

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