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Interhemispheric arachnoid cysts: case series.

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Objective: Interhemispheric arachnoid cysts (IHACs) are a rare congenital malformation arising from the duplication of the arachnoid membrane of the cerebral meninges associated with malformations of the corpus callosum. The scope of this study is to describe a series of cases of IHACs treated via neuroendoscopy in a reference center.
Method: description of a case series of children diagnosed with IHAC undergoing neuroendoscopic treatment (cystoventriculostomy) between 2015 and 2022, describing the main clinical manifestations, the surgical indication, and the neuroendoscopic technique performed.
Results: Four children diagnosed with IHACs, with a predominance of males (3:1), were included in the study. Considering the Mori and Barkovich classification systems, magnetic resonance imaging of the brain showed multiseptate extra-axial interhemispheric cysts without communication with the ventricular system in all cases. Macrocrania and bulging fontanel were the clinical manifestations that indicated urgent hospitalization for the proposed neurosurgical treatment.
Conclusion: Signs of increased intracranial pressure were observed in all cases of IHACs in the first months of life. Endoscopic cystoventriculostomy was performed in all cases with satisfactory results, showing that this is an option to be evaluated for the treatment of IHACs classified as multiseptate extra-axial and without communication with the ventricular system or brain cisterns, according to Mori and Barkovich classification systems.

Keywords: arachnoid cysts, cysts of the central nervous system, neuroendoscopy, neurosurgery, congenital malformations.

INTRODUCTION

Interhemispheric arachnoid cysts (IHACs) are in general of primary or congenital origin, formed from the arachnoid membrane of the cerebral meninges and involved by it, filled with cerebrospinal fluid (CSF) and commonly associated with midline brain malformations, especially of the corpus callosum [1-5]. Most often these cysts can be diagnosed in the fetal phase during adequate obstetric (prenatal) care [6].

When treatment is required, neurosurgical options can range from cyst-peritoneal shunt (CPS), microsurgery for opening the cyst membranes and communicating it with the ventricular system or brain cisterns, or neuroendoscopic treatment [3,4,5,7,8,9,10,11]. Due to the low incidence, there are few reports or case series of IHAC cases in the medical literature, hindering a consensus regarding management or the elaboration of protocols for the most appropriate timing for treatment and neurosurgical option [3,4,5,8,9,11,12,13]. The scope of this study is to describe a case series of children diagnosed with IHACs submitted to neuroendoscopy.

METHODS

Study approved by the research ethics committee (CAAE 67351323.0.0000.0048). Data were obtained through consecutive review of medical records of children up to 15 years of age who underwent neuroendoscopy for the treatment of cerebral arachnoid cysts between 2015 and 2022. Patients > 15 years of age, diagnosed with brain cysts other than IHACs, submitted to non-endoscopic treatments, or with less than 01 year of postoperative follow-up were excluded from the final sample.

The following data were collected from the included medical records:

- Sex; age at diagnosis; pre-surgical cranial circumference, aspect of the fontanels, and neurological examination; classification of IHAC based on magnetic resonance image (MRI) of the brain, according to Mori [1] and Barkovich [2] classification systems; reason for surgical indication as described in the medical records and surgical technique performed.

- Follow-up: The minimum postoperative follow-up time for defining and detecting possible post-surgical

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complications was 01 year, and at least 02 imaging exams were performed during this period in all patients included in the study: transfontanellar ultrasound [TFUS], cranial tomography [CT scan] or MRI.

Surgical technique

Patients underwent neuroendoscopy (Karl Storz - OI HandyPro 120 rigid pediatric neuroendoscope) performed under general anesthesia. A single burr hole in the skull was used for access, made in the frontal or parietal region depending on the proximity and planned trajectory for communication between the IHAC and the ventricular system (endoscopic cystoventriculostomy - ECV) or between the cyst and the brain cisterns (endoscopic cystocisternostomy - ECC). After opening the bone plane, the dura mater was coagulated and opened carefully to avoid postoperative CSF leakage or bleeding during the procedure. Neuronavigation for planning the surgical access was used in two cases. After the introduction of the neuroendoscope into the cyst, the decision to establish the communication between the cyst and the ventricular system or the brain cisterns was based on the observation of the thinnest and most transparent site of the arachnoid membrane, to avoid damage to the adjacent brain or bleeding. We generally tried to make at least 3 communications between the cyst and the ventricular system and /or the brain cisterns. These fenestrations were made using endoscopic monopolar cautery, usually without the use of the coagulation function. After perforation of the cyst membrane, the stoma was enlarged by the use of a number 3 Fogarty catheter. Coagulation was used exclusively in cases of bleeding. The technique used was based on previously standardized neuroendoscopic surgery protocols, including the use of planning with neuronavigation [4,5,8].

CASE SERIES

Four children were diagnosed with IHAC and underwent brain endoscopy during the proposed period. There was a predominance of males (3:1) whose initial diagnosis was suspected during the fetal phase (prenatal diagnosis). In all cases, the birth occurred at term, without the need to anticipate the delivery. At birth, all children had macrocrania (cranial perimeter with standard deviation > +2). In preoperative MRI, in all cases, IHACs were classified as extra-axial without ventricular communication (Mori classification) and type 2 IHAC according to Barkovich classification (multiseptate without communication with the ventricular system or brain cisterns). All children underwent emergency brain neuroendoscopy up to the second month of life due to signs of increased intracranial pressure. The initial treatment was CPS in one of the children and ECV was the first surgical option in the others. The child whose initial treatment was CPS underwent ECV at the age of 4, after removal of the CPS. The general characteristics of the series are described in Table 1.

Case 1

Boy with prenatal diagnosis (2nd gestational trimester) of IHAC. He was subjected to CPS at 2 months of age due to increased cranial perimeter and bulging fontanels. At 4 years of age, an increase in the frontal cystic cavity (IHAC with multiple septa) was identified in the imaging exams and ECV was performed (Figure 1). At that time, the decision was to maintain the CPS. At 11 years of age, a surgical revision of the CPS was performed in view of the shortening of the peritoneal catheter due to the growth of the child and a new neuroendoscopic approach was proposed. In this surgical review, we identified the calcification and obstruction of the peritoneal catheter, without CSF drainage. The neuroendoscopic access made it possible to check the patency of previously created stomas and establish new communications between the cyst and the ventricular system. In agreement with family members, we opted for the removal of the CPS. There were no surgical or post-surgical complications. The child is being regularly followed-up without need for further neurosurgical interventions and imaging tests identified the patent communication of the ECV.

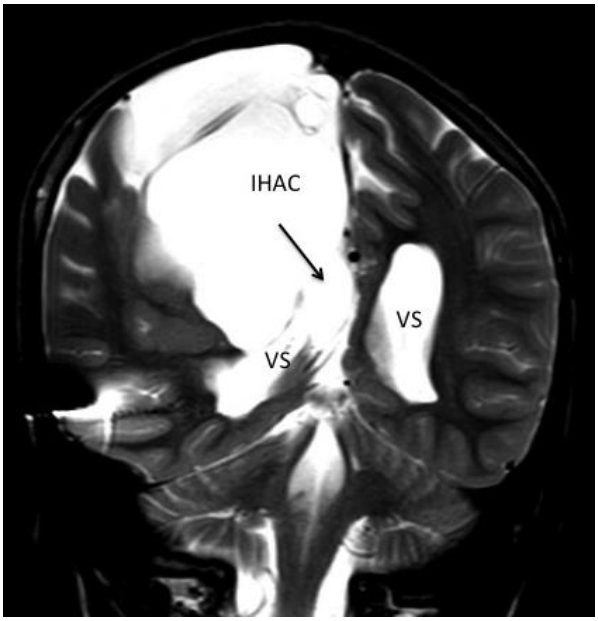

Figure 1- Post-surgical magnetic resonance image (endoscopic cystoventriculostomy stoma: black arrow) showing an extra-axial interhemispheric arachnoid cyst (IHAC) with multiple septa. VS: ventricular system.

Case 2

Male infant with prenatal diagnosis (12th gestational week) of malformation of the corpus callosum associated with bulky midline cerebral cyst. At birth, he had a cranial perimeter of 39 cm, with no other changes on neurological examination. MRI of the brain showed a voluminous extra-axial IHAC in the right sagittal plane with malformation of the corpus callosum, without communication with the ventricular system (Figure 2). Urgent hospitalization was

Table 1- Clinical and radiological features (brain magnetic resonance image) of the four cases of congenital interhemispheric arachnoid cysts (IHACs) identified and treated through neuroendoscopy (endoscopic cystoventriculostomy) (Salvador da Bahia, Brazil, 2022).

Case	Sex	Clinical condition	Predominant location	Septate	Hydrocephalus	Agenesis of the corpus callosum	Mori [a]	Barkovich[b]
1	Boy	Macrocrania and bulging fontanels	R	Yes	Yes	Partial	EA	2
2	Boy	Macrocrania and bulging fontanels	R	Yes	Yes	Complete	EA	2
3	Boy	Macrocrania and bulging fontanels	R	Yes	Yes	Partial	EA	2
4	Girl	Macrocrania and bulging fontanels	R	Yes	Yes	Complete	EA	2

R - right; EA - extra-axial; a: Classification proposed by Mori (1992); b: Classification proposed by Barkovich et al. (2001).

necessary due to increased cranial perimeter (43.5 cm; standard deviation +4), bulging fontanels, and irritability. The patient was urgently submitted to neuronavigation-guided ECV at 45 days of age. During the follow-up, despite the macrocrania, there was a reduction in the growth rate of the cranial perimeter, with no signs or symptoms of intracranial hypertension. MRI showed the patent communication of the ECV.

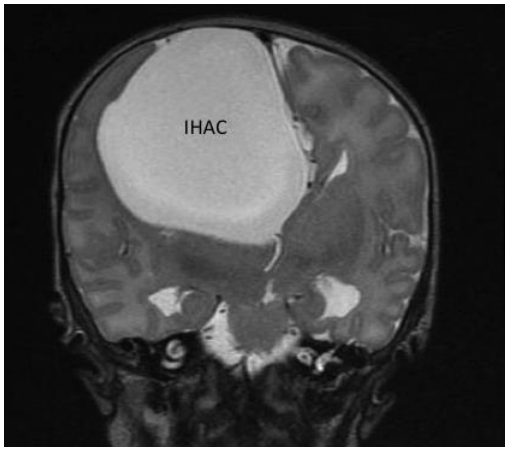

Figure 2- Magnetic resonance image of the brain showing an interhemispheric extra-axial arachnoid cysts (IHAC) without communication with the ventricular system or brain cisterns, according to Mori and Barkovich classification.

Case 3

Male infant with prenatal diagnosis (16th gestational week) of IHAC. MRI identified a large cyst in the posterior middle third of the brain, in the right para-median region, septate, associated with an increase in the ventricular system. At birth he had a cranial perimeter of 39 cm, with no other changes on neurological examination. At 2 months of age, he evolved with bulging fontanels, irritability, disjunction of sutures, and increased cranial perimeter (42.3 cm; standard deviation + 3). He was hospitalized and urgently submitted to neuronavigation-guided ECV. There were no surgical complications. The child was regularly monitored after surgery without neurological complications, without signs of intracranial hypertension, and with reduced growth rate of the cranial perimeter despite macrocrania. Follow-up MRI showed the patency of the communications of the ECV (Figure 3).

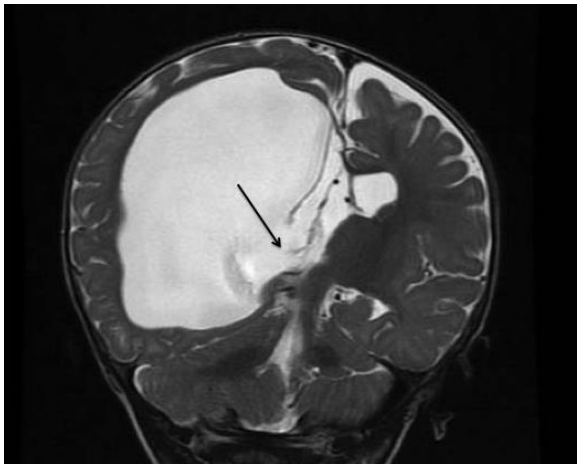

Figure 3- Post-surgical magnetic resonance image of thebrain showing neuroendoscopic communication (black arrow) between the cyst walls and the ventricular system.

Case 4

Female infant with fetal diagnosis of congenital malformation of the central nervous system including bulky IHAC. She was born at term, with macrocrania associated with other congenital malformations (polydactyly, cleft palate, micrognathia). In the first month of life, she evolved with bulging fontanels and increased cranial perimeter (39 cm; standard deviation of +3), requiring urgent hospitalization. Pre-surgical MRI showed agenesis of the corpus callosum, polymicrogyria, pachygyria, multiseptate IHAC with greater extension to the right, and enlargement of the ventricular system. The child was urgently submitted to ECV (Figure 4). There were no surgical or postoperative complications. The child was followed-up without neurological complications or signs of intracranial hypertension and with reduced growth rate of the cranial perimeter. Imaging tests for postoperative follow-up showed the patency of the ECV.

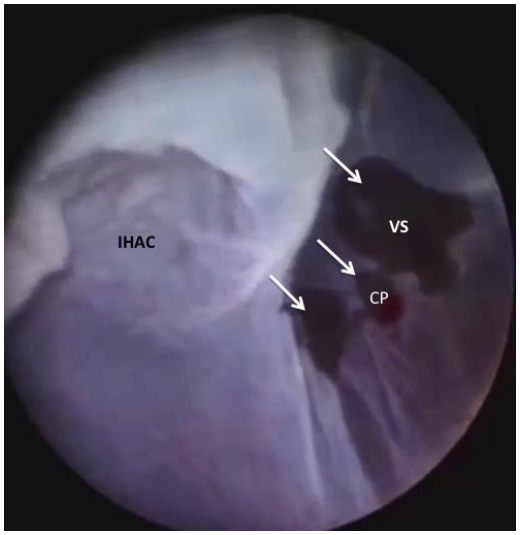

Figure 4- Communications (stomas: white arrows) performed during endoscopic cystoventriculostomy. IHAC: interhemispheric extra-axial arachnoid cyst. VS: ventricular system. CP: choroid plexus.

DISCUSSION

Congenital arachnoid cysts occur in approximately 2.6% of children [9,13] and IHAC are considered rare, corresponding to approximately 10% of arachnoid cysts, and this precludes the creation of standardized management and treatment protocols [4,5,7,8]. The higher incidence in boys, a finding of the current study, has been described in the literature [5,8]. Regular follow-up and prenatal examinations are essential for early diagnosis in order to provide greater comfort and adequate guidance and explanations to parents, as in other congenital malformations of the central nervous system [6], giving the pediatric neurosurgeons the opportunity to plan and define the best treatment strategies, given the lack of standardized protocols. In the current study, the diagnosis made still in the fetal phase, during prenatal consultations, resulted from adequate and regular obstetric follow-up, described in the four medical records analyzed in this study. In these cases, the pediatric neurosurgeon usually recommends periodic obstetric ultrasounds and fetal MRI for a better definition and possible classification of the IHAC and investigation of other possible malformations of the central nervous system. Always seeking consensus with the opinion and follow-up of the obstetrician, the possibility of term delivery must be considered so as to avoid complications related to prematurity [14]. In none of the cases reported in this series was there a need to anticipate the delivery.

Regarding the clinical manifestations, increased cranial perimeter and bulging fontanels are often described in cases of intracranial arachnoid cysts with indication for neurosurgical treatment, either due to the increase of the cyst and/or obstructive hydrocephalus [7,8,13]. In the present study, all children were born with macrocrania and evolved in the first months of life with signs of increased intracranial pressure due to increase of the cyst and obstructive hydrocephalus. Mori (1992) [1] and Barkovich et al. (2001) [2] elaborated and proposed criteria for distinguishing and defining IHACs, thus assisting in the planning of the best management [5,15]. According to Mori classification, IHACs can be divided into two major groups: intra-axial and extra-axial. Considering the interhemispheric location, the aspects evaluated include malformations of the corpus callosum and the aspect concerning the pattern of the ventricular system (increased or not, with hypertensive hydrocephalus or ventricular dilatation ex-vacuo) [1]. In this classification, the main aspects for an adequate indication of a neuroendoscopic approach are the extra-axial interhemispheric location, absence of communication of the cyst with the ventricular system or the subarachnoid space/brain cisterns, and absence of hydrocephalus ex vacuo [5,15]. The Barkovich classification system, in turn, consists of 4 subcategories in-group 2 (multiseptate cysts without ventricular communication) [2]. Other research groups have found that IHACs considered for neuroendoscopic treatment should be included in these categories [5,15]. These criteria

were identified in the four children in the present case series and the development of hydrocephalus was possibly a determining factor for the indication of urgent surgery.

There is no consensus in the literature on the most appropriate moment (age) for the neurosurgical approach in the case of children with IHAC, but the consensus regarding the surgical indication in cases of increased intracranial pressure is indisputable [7,8]. Regarding the neuroendoscopic technique, we consider it important to perform the largest possible number of septa openings and communications between cavities because these septa prevent the circulation of CSF between the cystic cavities and the ventricular system or brain cisterns. We emphasize that attention and care in surgical preparation and planning are essential to reduce complications related to neuroendoscopic surgery, including planning of the stoma site. Deciding whether IHAC communication with brain cisterns or with the ventricular system is more appropriate seems to depend on the proximity of the cyst to these structures [3,5,15]. In the 04 cases described here, ECV was chosen due to the proximity between the cyst and the ventricular system, a factor that facilitates the communication between these two spaces. This option has also been described as preferable among endoscopic techniques in previous studies [5].

The results presented here regarding surgical complications corroborate the literature on the subject, which describes low rates of surgical or postoperative complications. These figures make neuroendoscopic technique to be considered safe and adequate for the treatment of certain congenital brain cysts, including IHACs [3,5,8]. In the postoperative follow-up of the four children, despite the macrocrania, there was a reduction in the growth rate of the cranial perimeter, also described by other authors in previous studies [5]. In the imaging tests performed during the follow-up period proposed for the present study, there was stabilization of growth or even reduction of the IHAC or the ventricular pattern in all children of this series.

CONCLUSION

There is no consensus on the best surgical strategy for IHACs due to their low incidence. In children who evolve with increased cyst volume and signs of elevated intracranial pressure, associated or not with hydrocephalus, the indication for surgery is indisputable. Endoscopic cystoventriculostomy was performed in all cases with satisfactory results, showing that this is an option to be considered for the treatment of IHACs classified as multiseptate extra-axial and without communication with ventricular system or brain cisterns, according to Mori and Barkovich classifications.

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, number: 67351323.0.0000.0048

Consent to participate

The patients gave consent to use their information and images for research purposes. *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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CONTRIBUTIONS

-José Roberto Tude Melo: Conceptualization, Data curation, Formal Analysis, Funding acquisition, Investigation, Methodology, Project administration, Resources, Supervision, Validation, Visualization, Writing – original draft, Writing – review & editing

-Leonardo Conrado Silva Lima: Conceptualization, Data curation, Funding acquisition, Resources, Writing – review & editing

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